



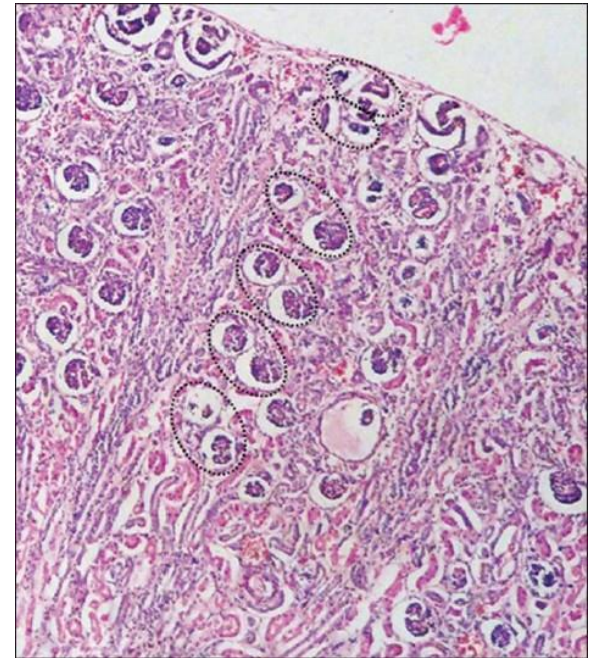
Histologie Rénale Normale (pratique) et lésions élémentaires glomérulaires.

DIU de Néphropédiatrie

**Pr L. DANIEL,
Décembre 2018**

Rein Foetal ...

- Immature à la naissance (élongation des tubules et volume glomérulaire).
- 9 à 11 générations de néphrons de maturation centrifuge (8 s à 36 s).
- Existence d'une glomérulosclérose physiologique (2 à 3%).



Rein 19 S de gestation avec 6 générations de glomérules

artères:

1. interlobaire

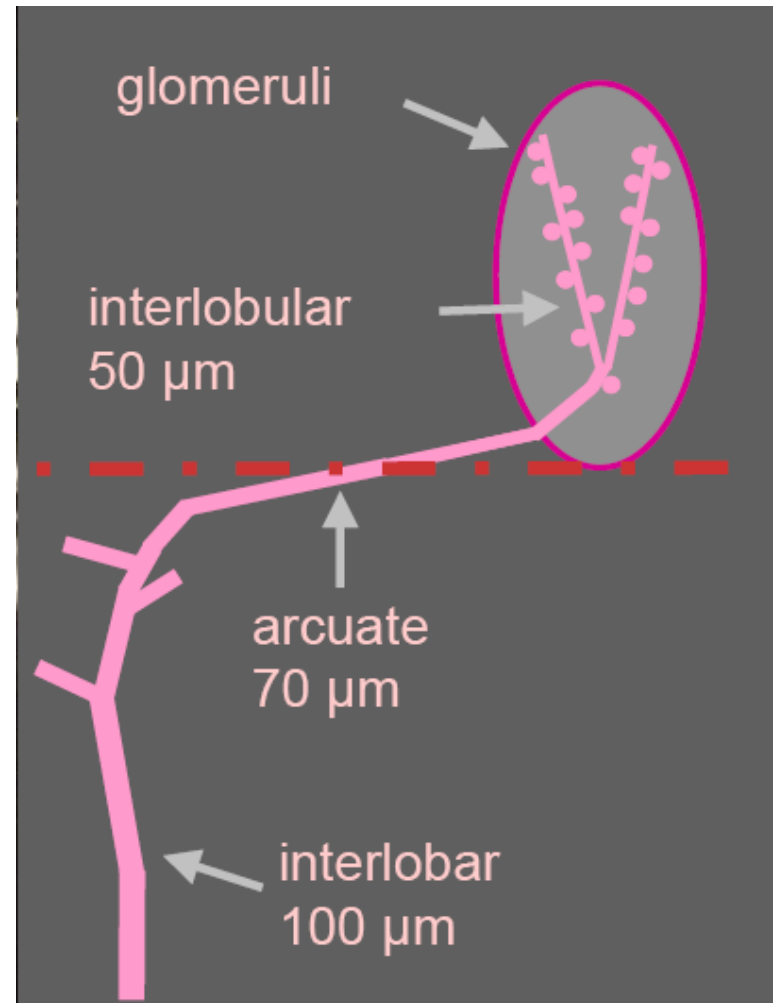
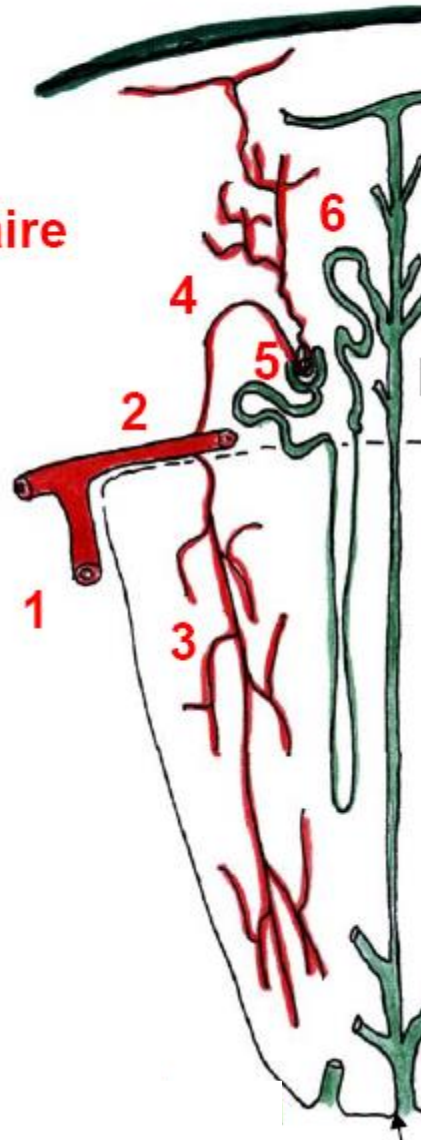
2. arquée

3. droite

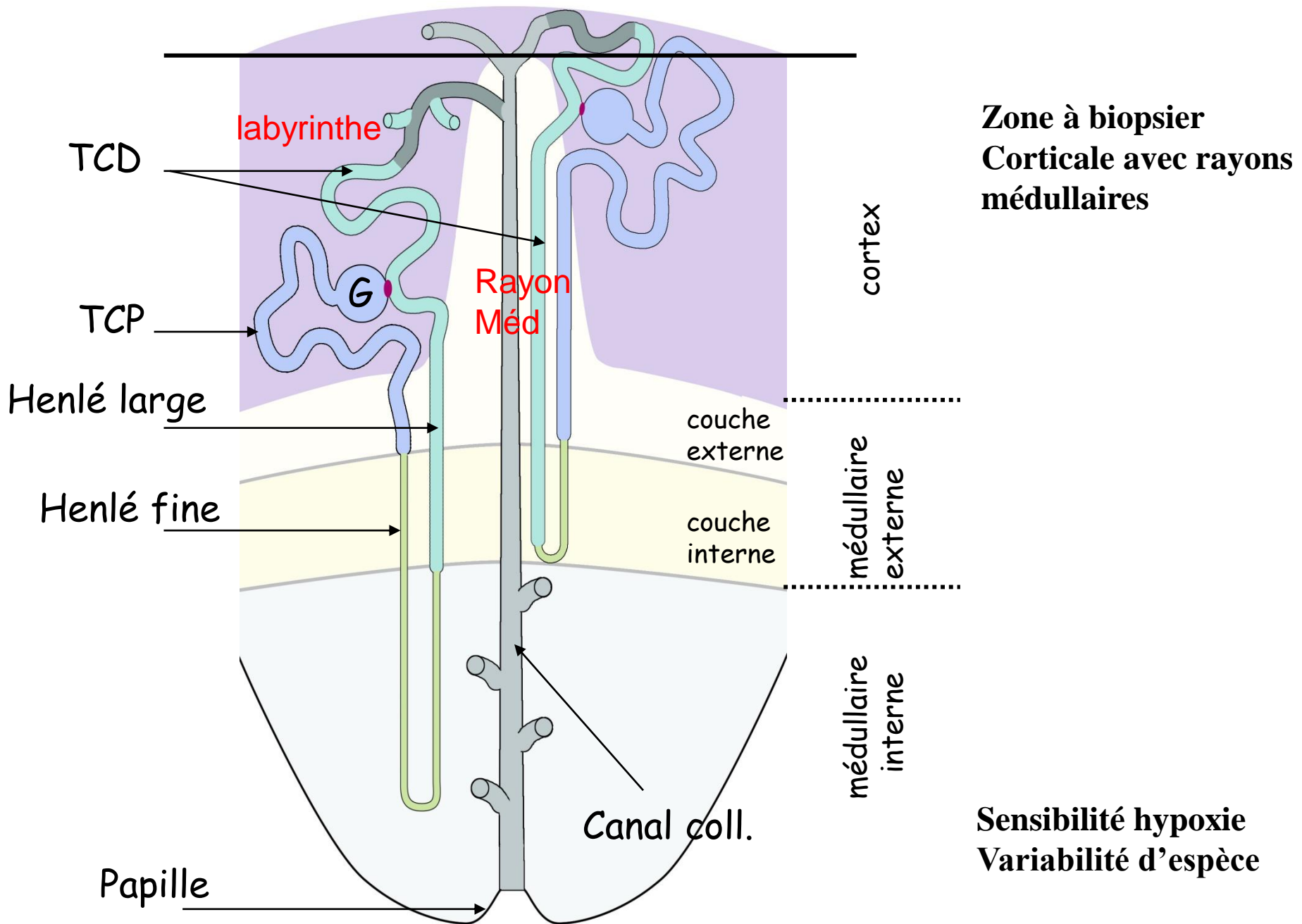
4. interlobulaire

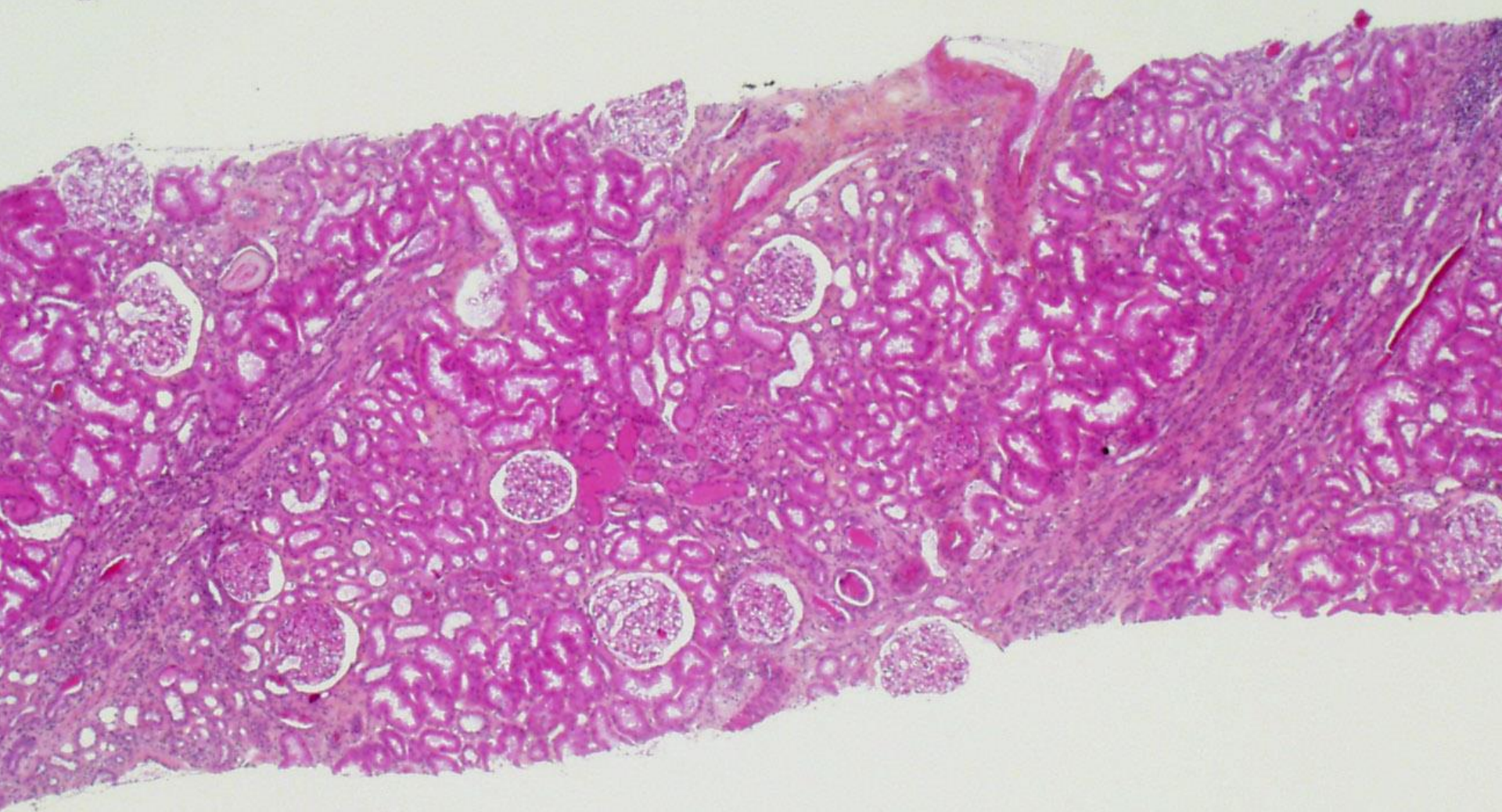
5. afférente

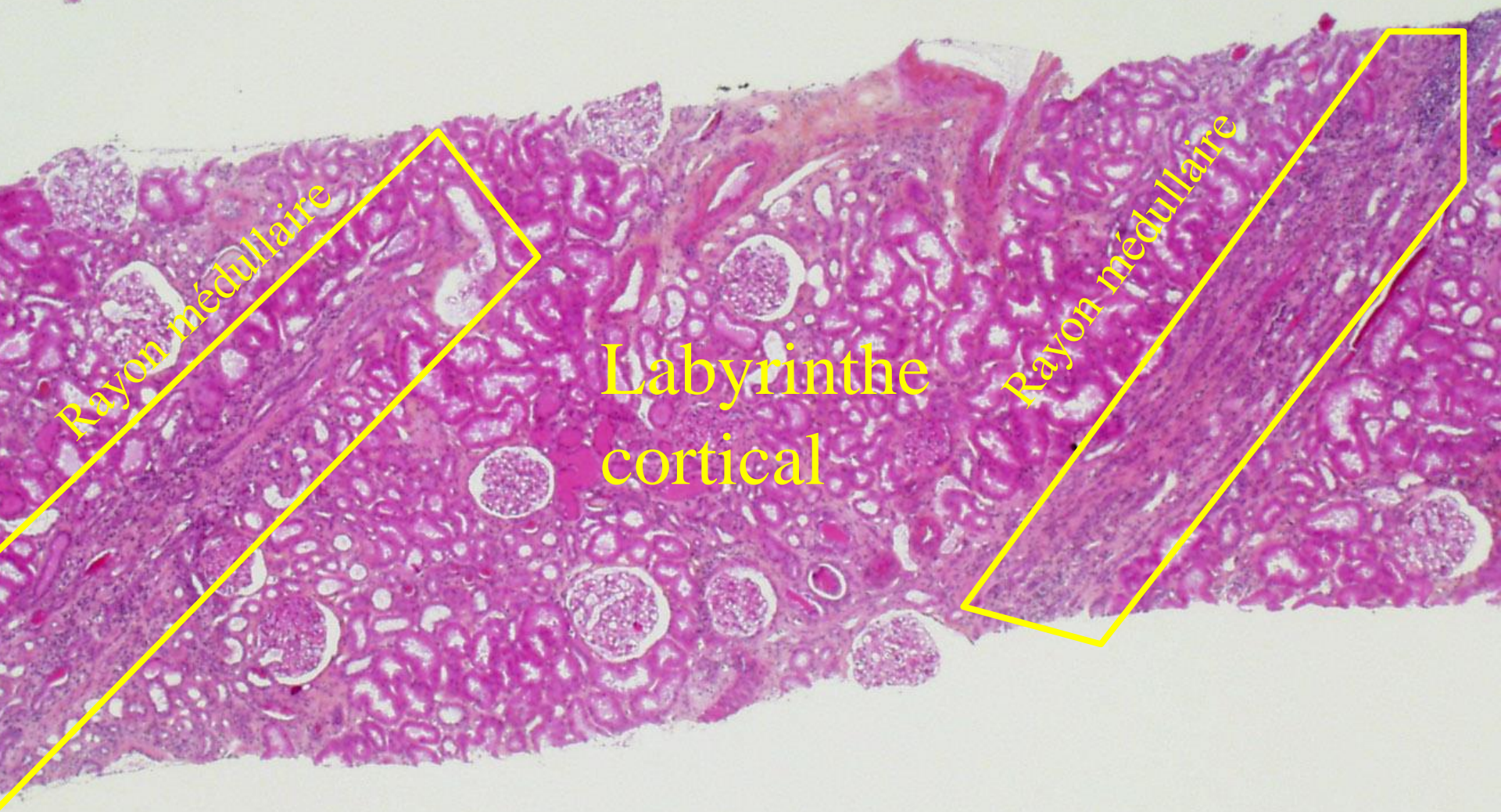
6. efférente



TCP: 12-24 mm, unistratifié (4-5 cell); TCD: 5mm, unistratifié (5-8 cell.).



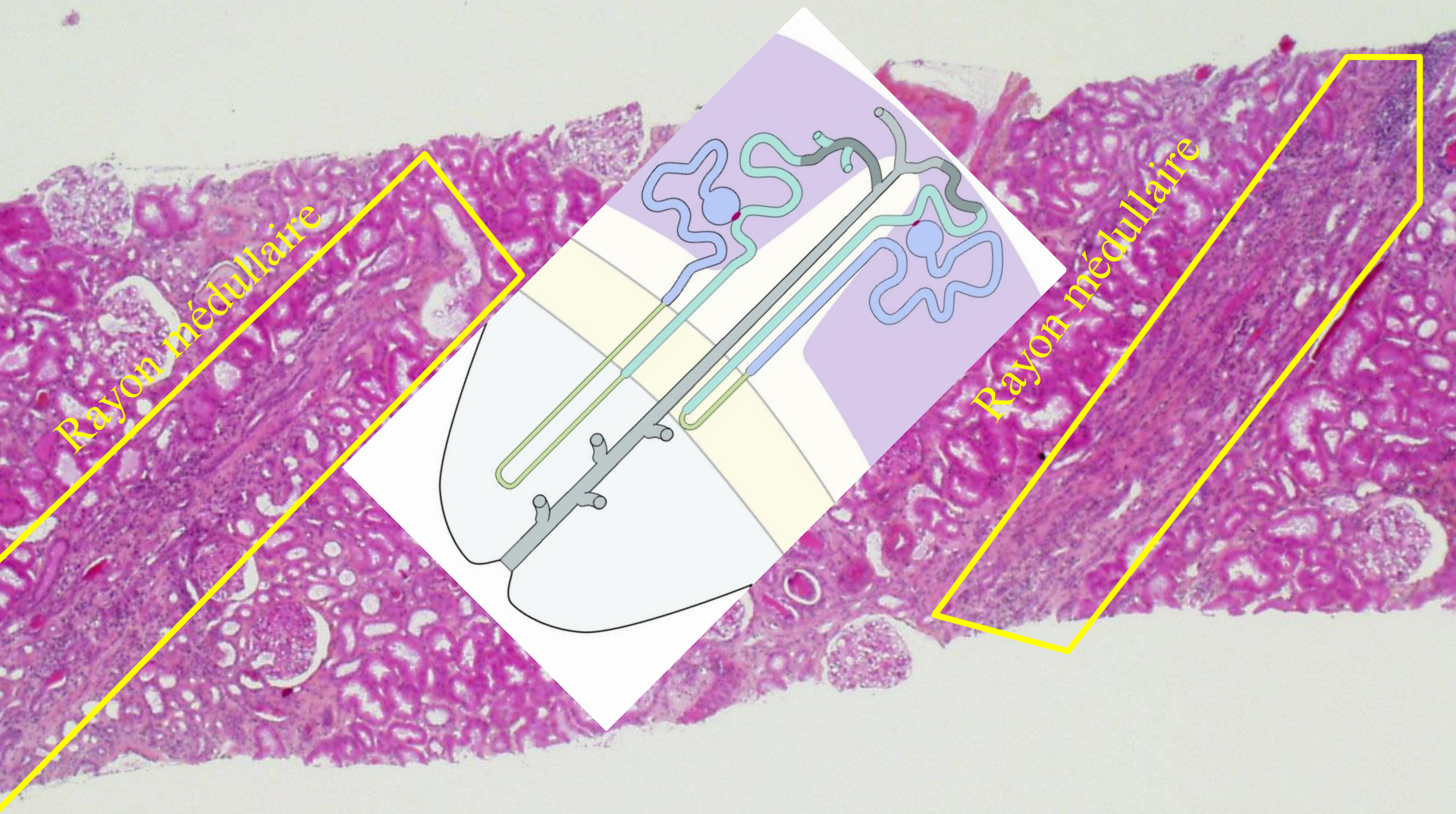




Rayon médullaire

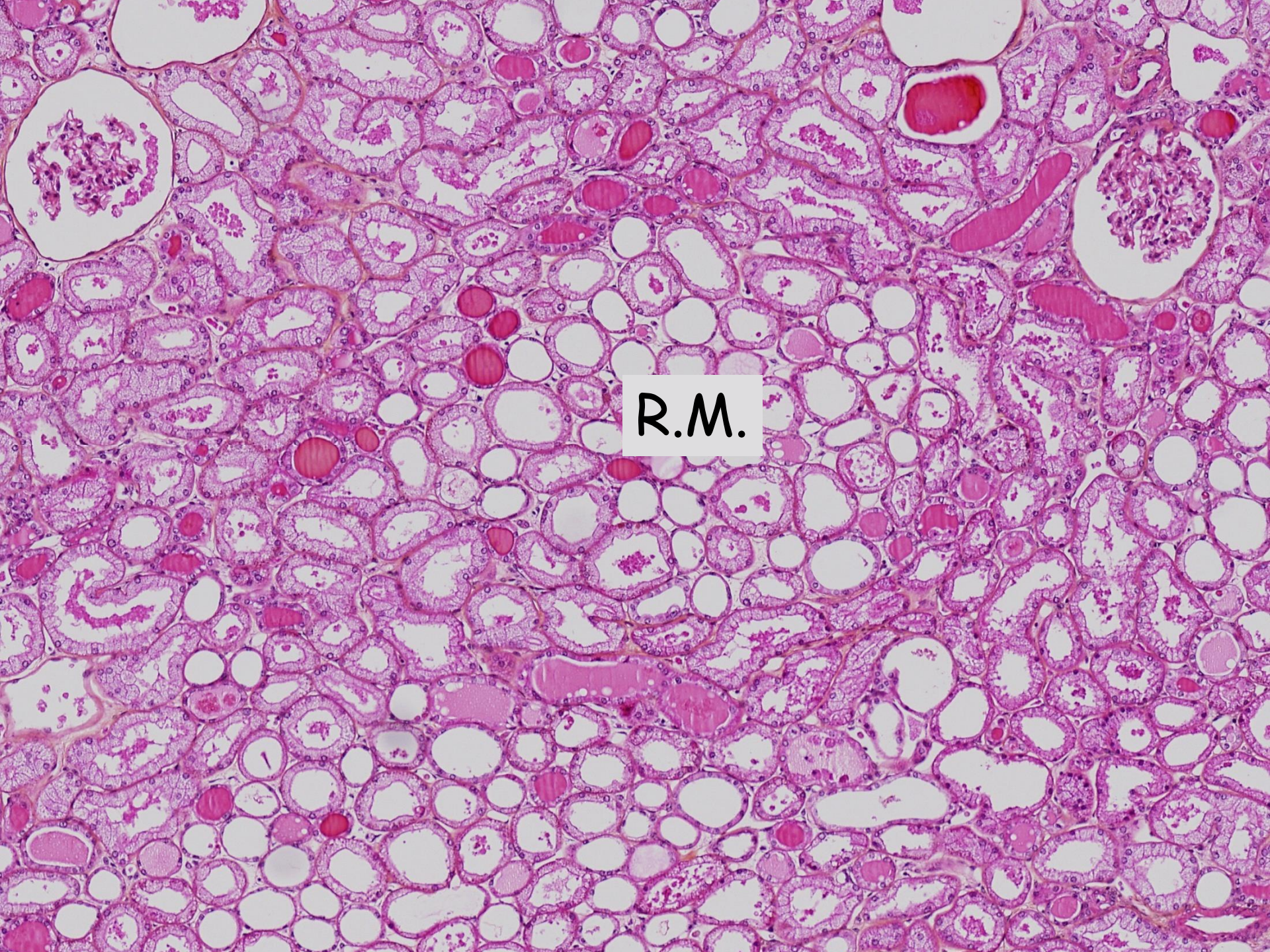
Labyrinthe
cortical

Rayon médullaire

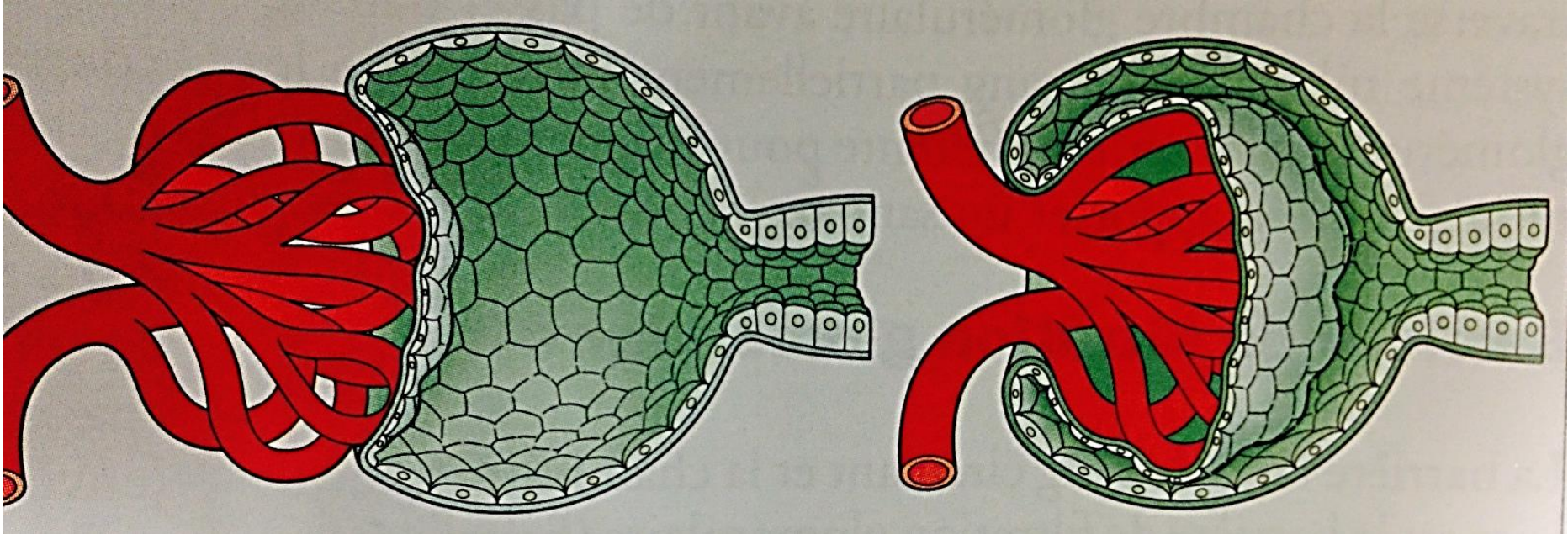


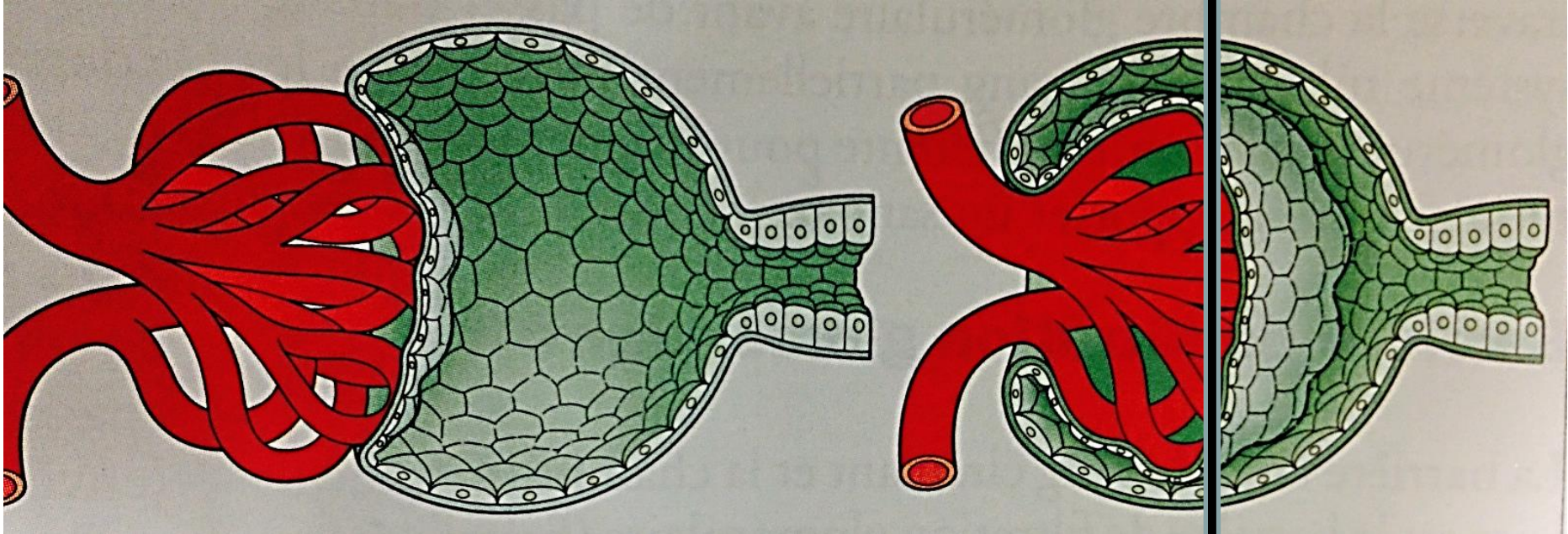
Rayon médullaire

Rayon médullaire

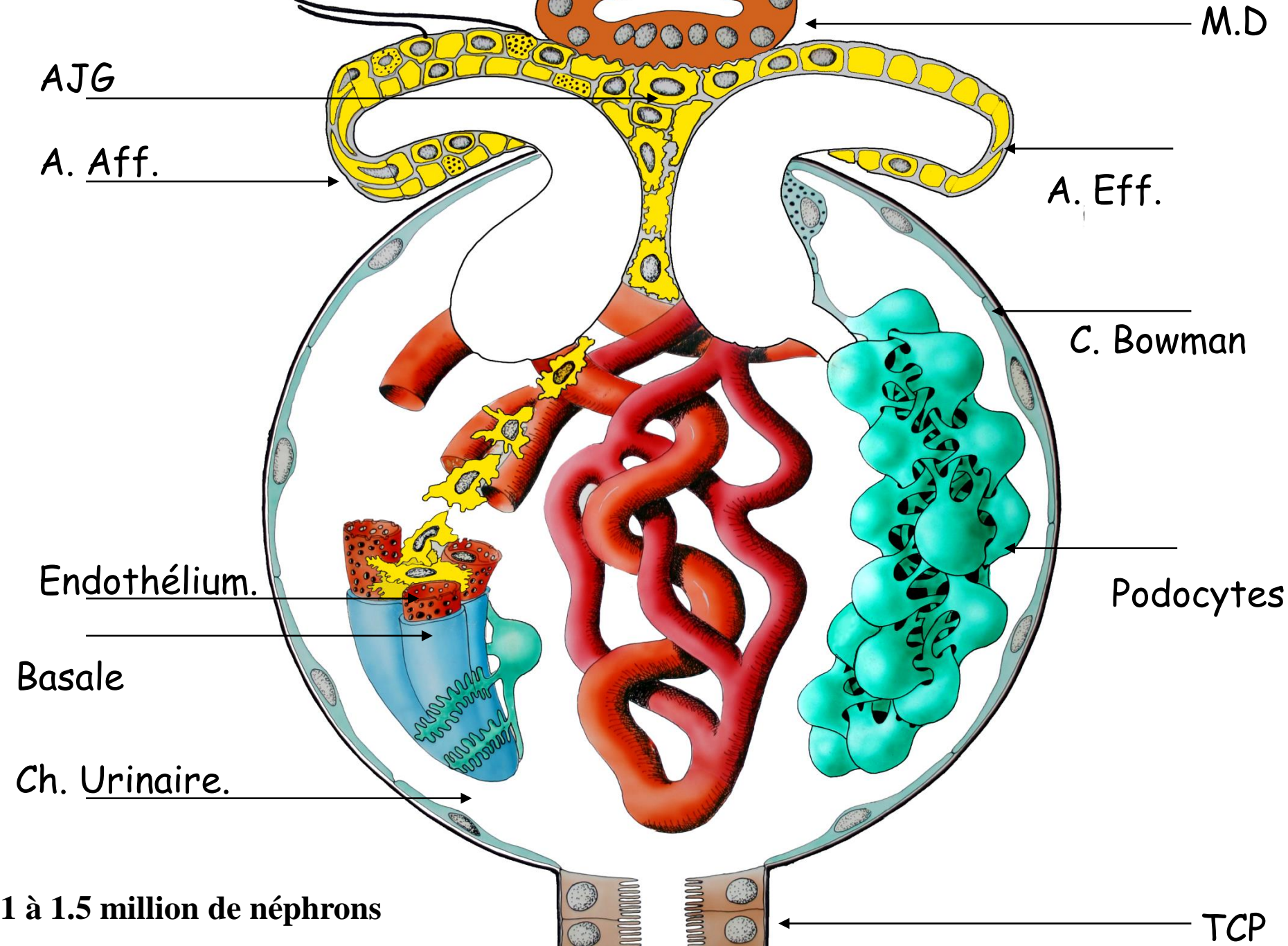


R.M.

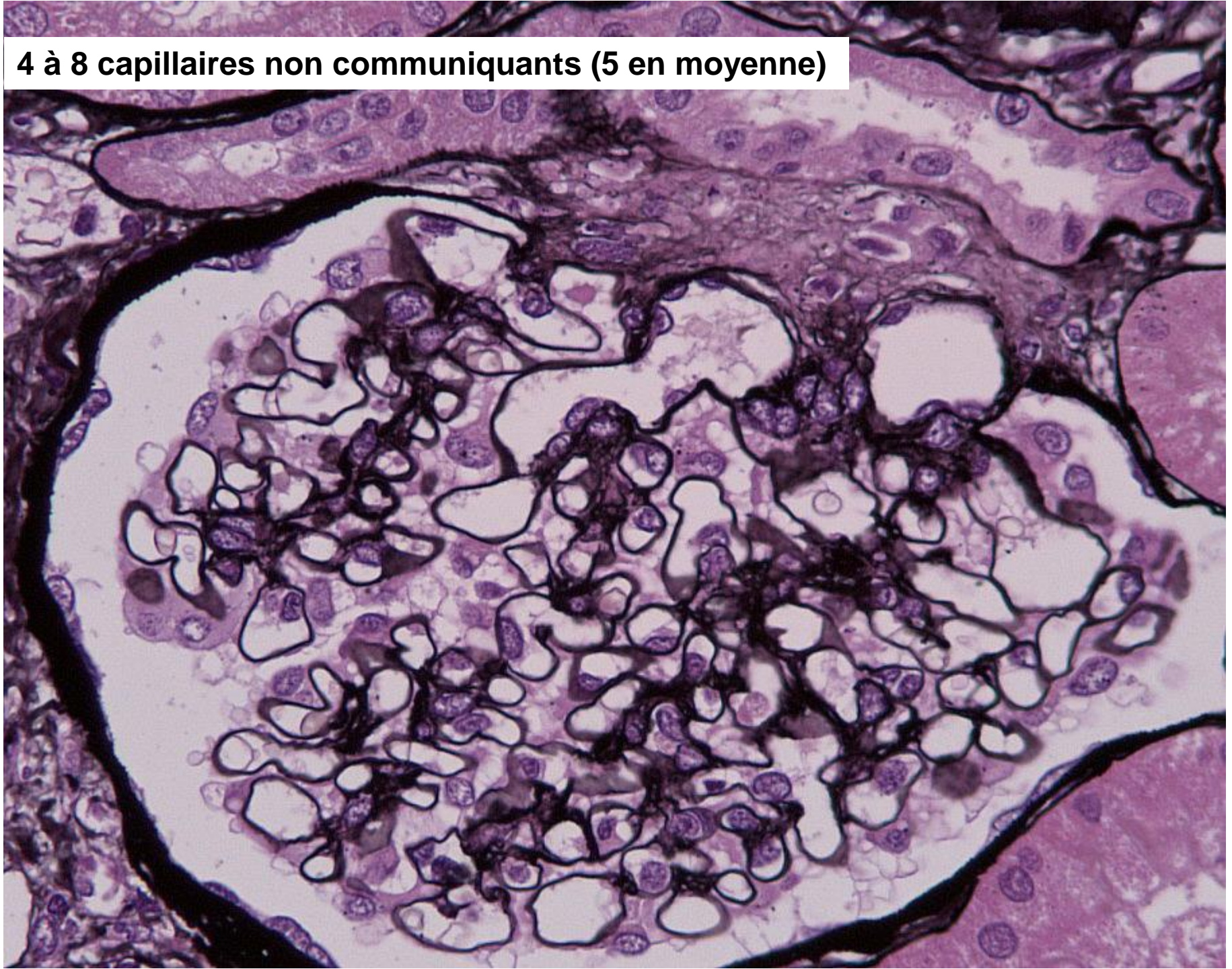


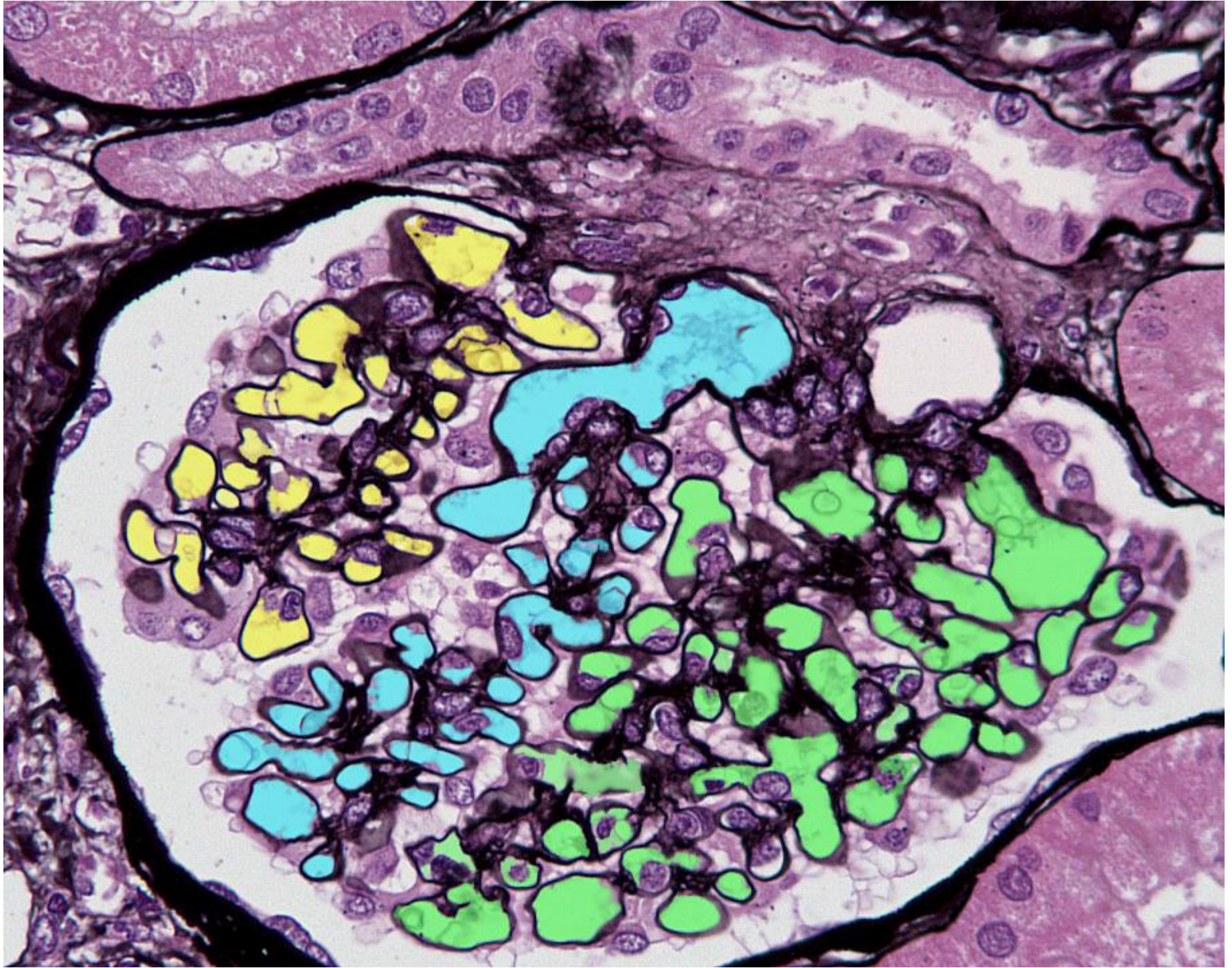


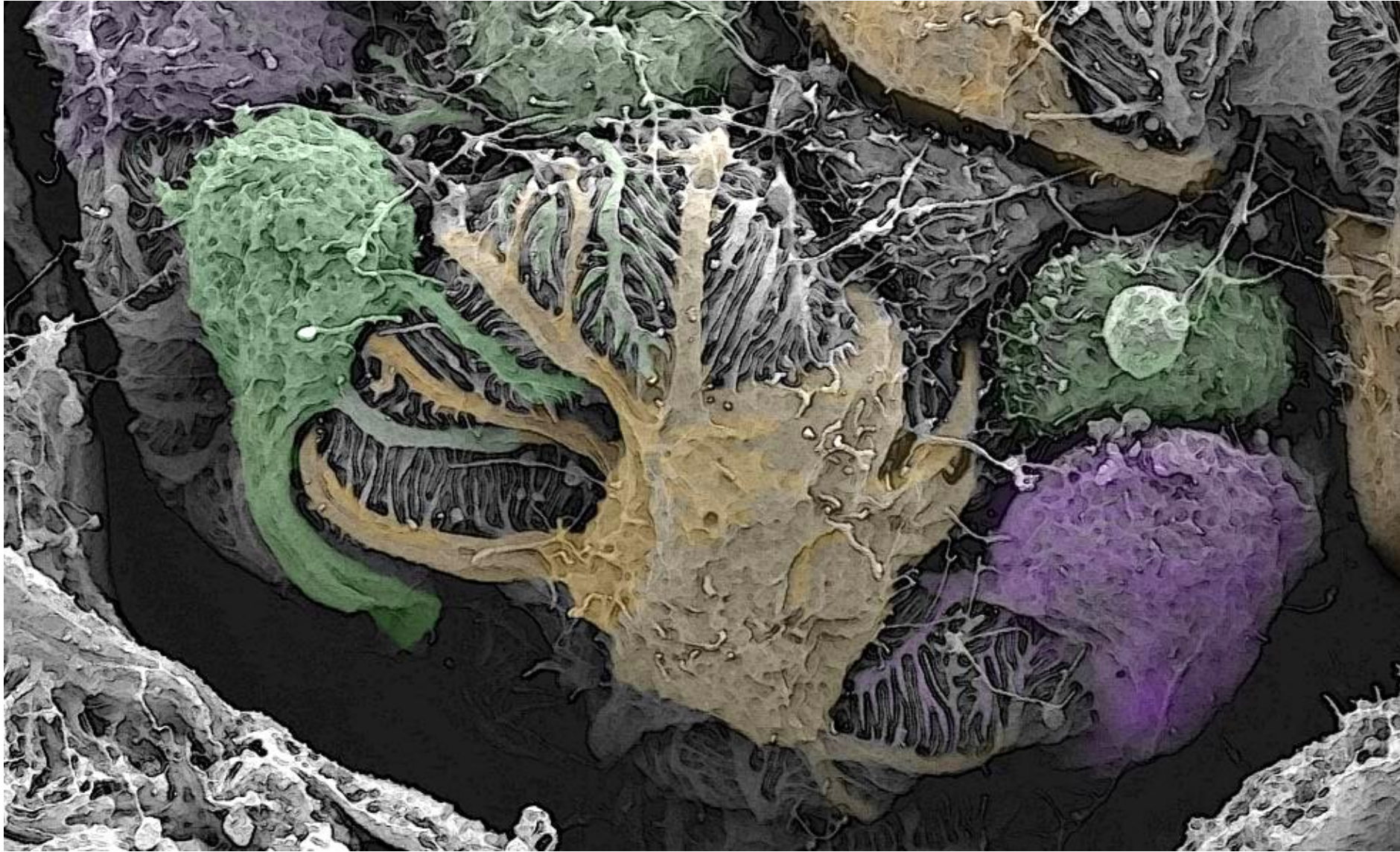
Malpighi, 1660



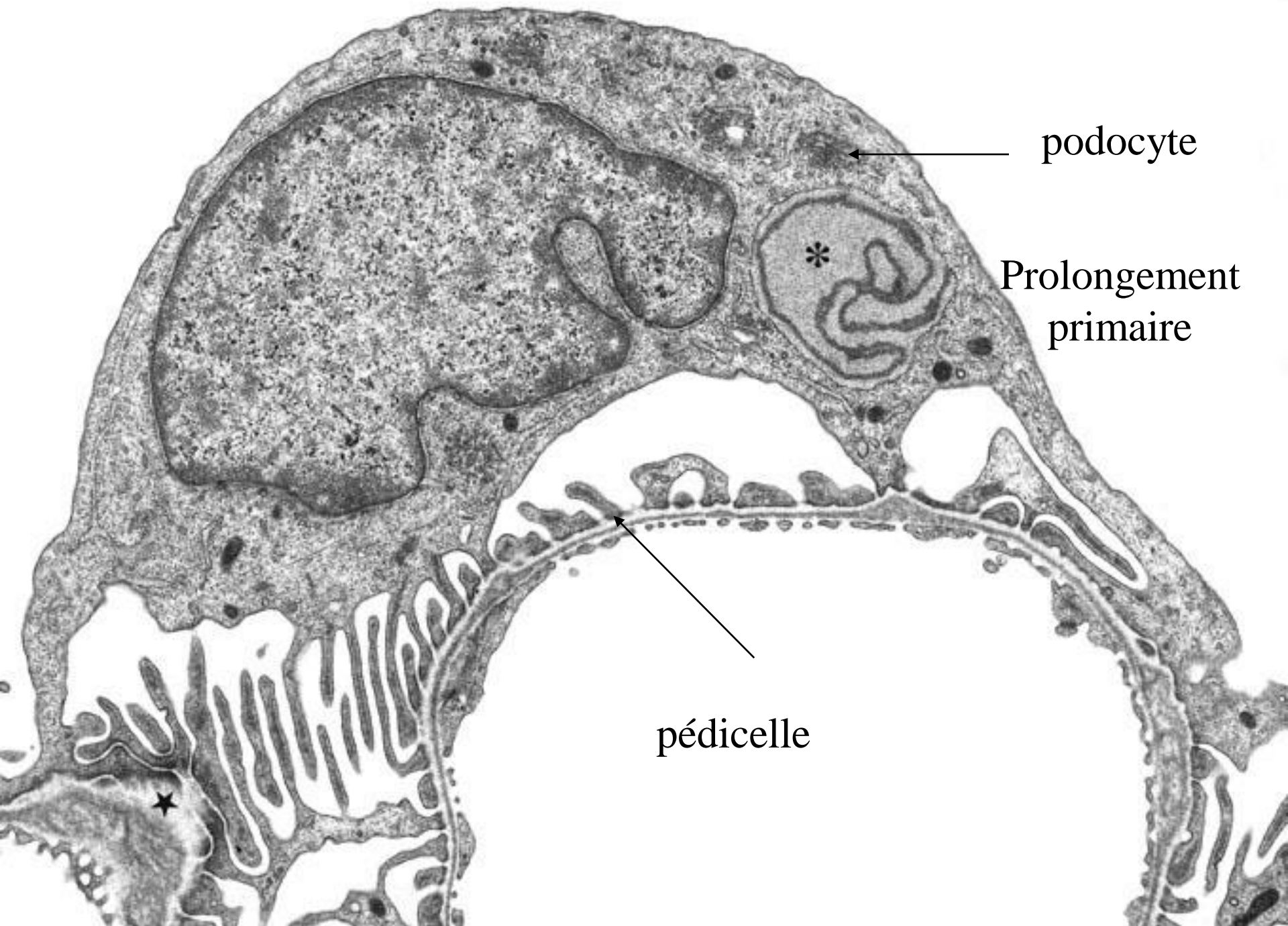
4 à 8 capillaires non communiquants (5 en moyenne)







Surface filtrante de 2 à 3 m²



podocyte

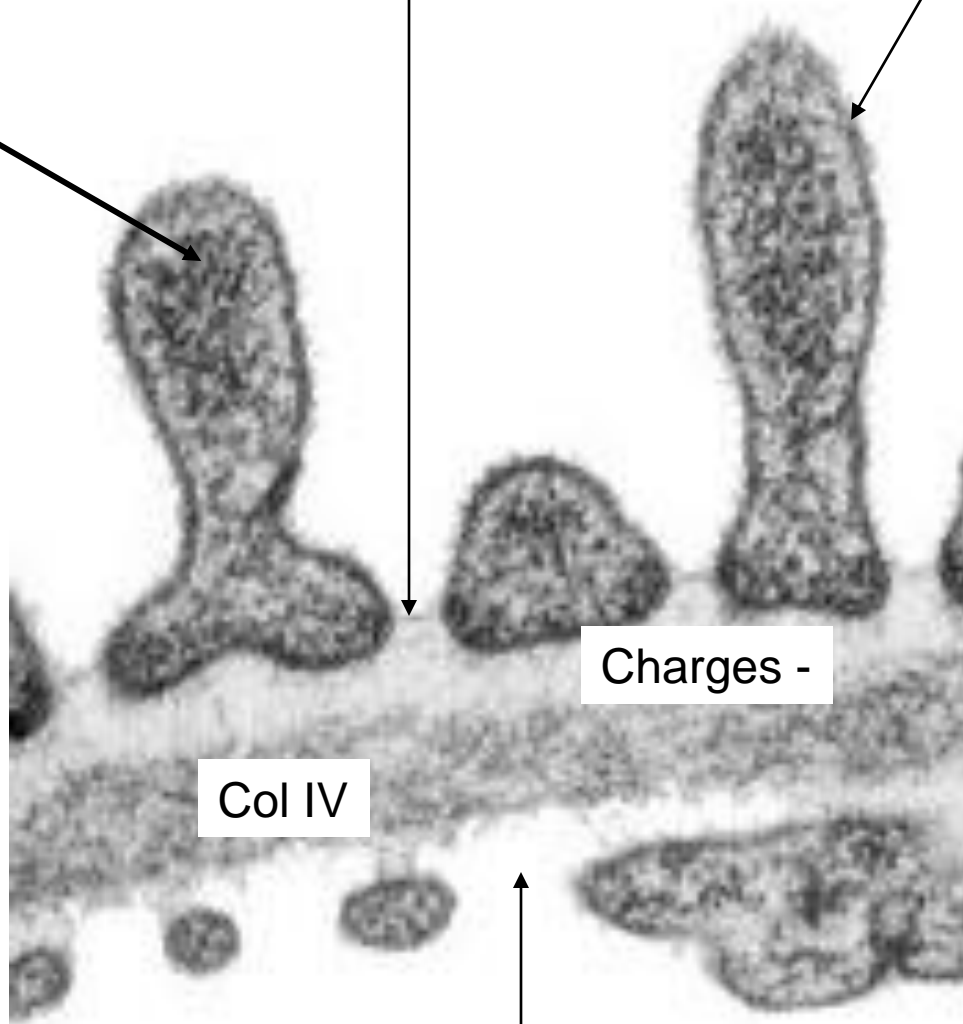
Prolongement
primaire

pédicelle

Diaphragme de fente

glycocalix

filaments



Charges -

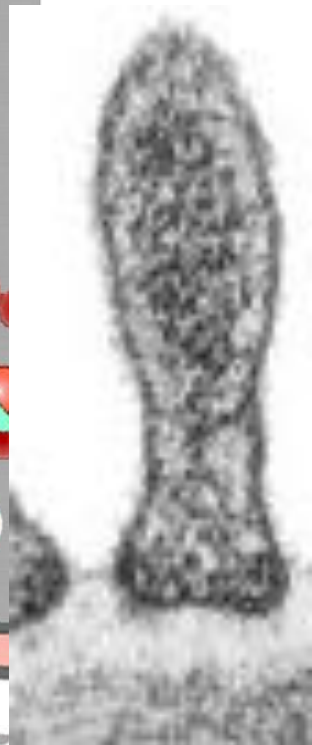
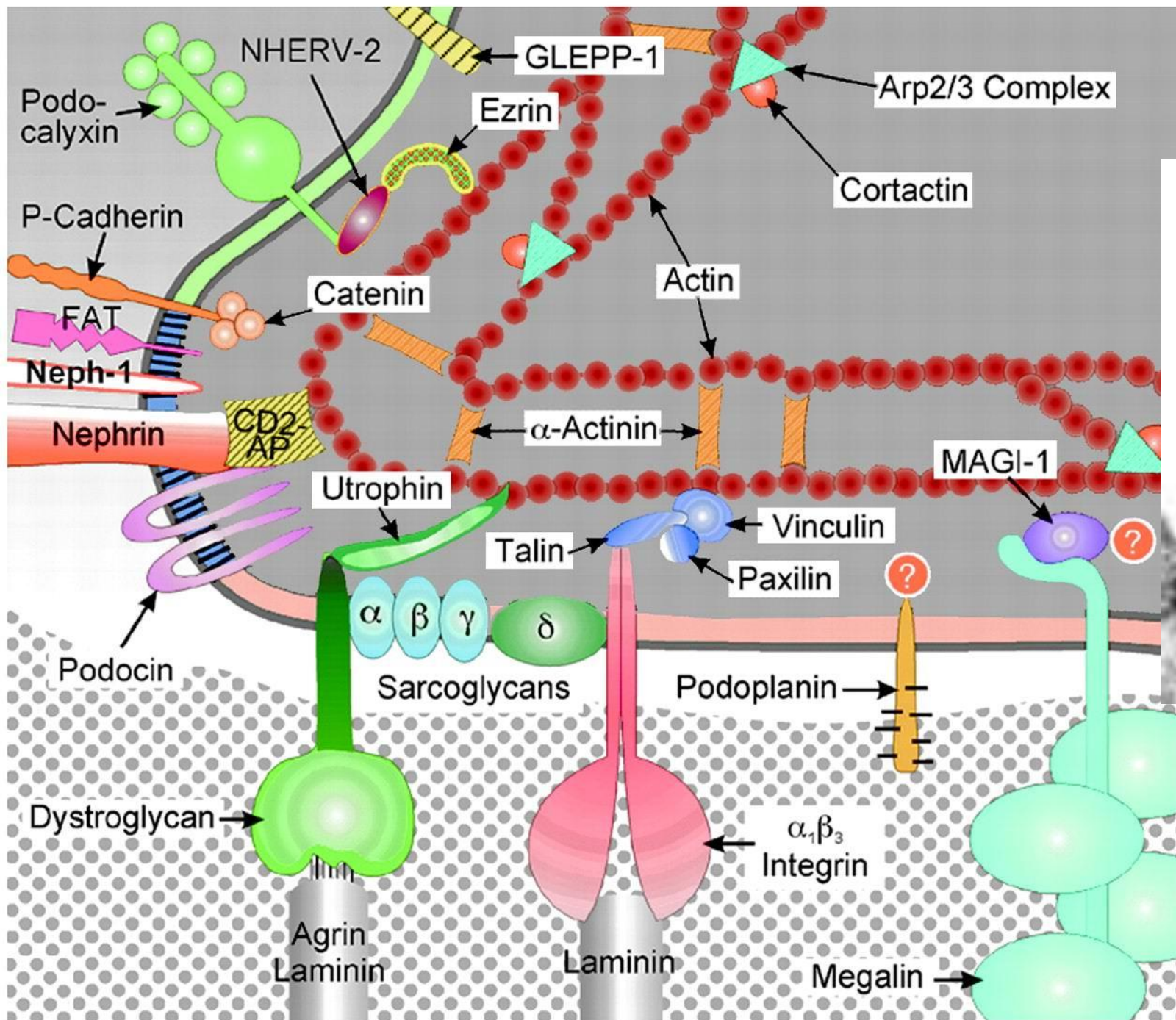
Col IV

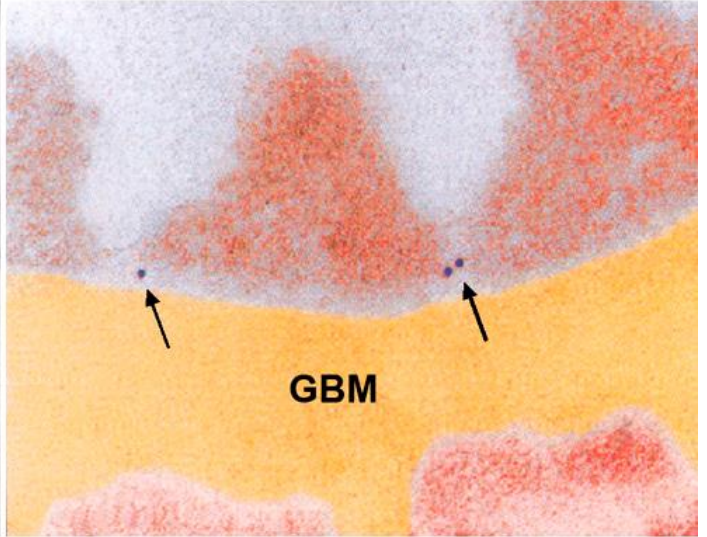
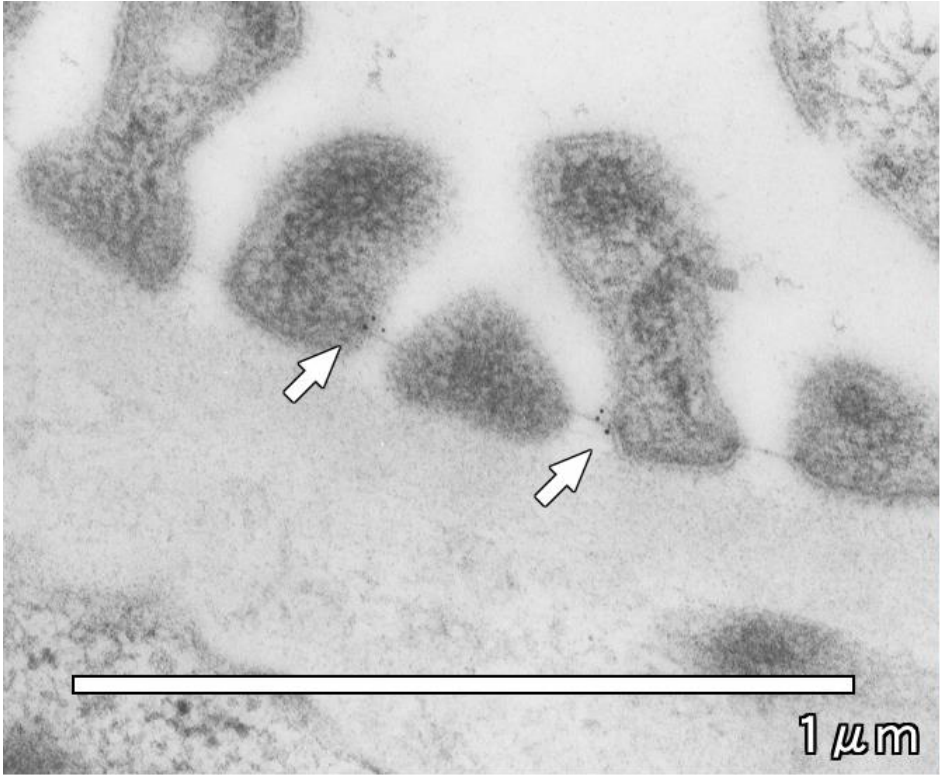
rara externa
lamina densa
rara interna

(visibles enfants et rongeurs).....

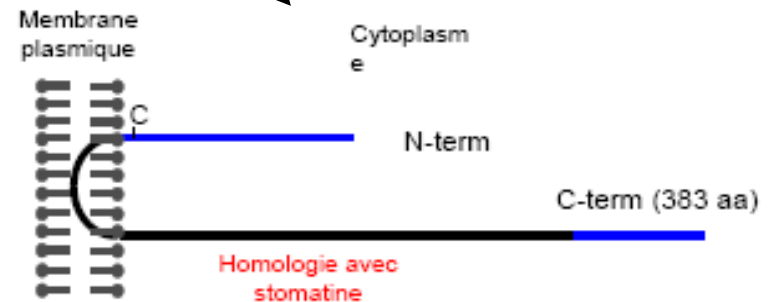
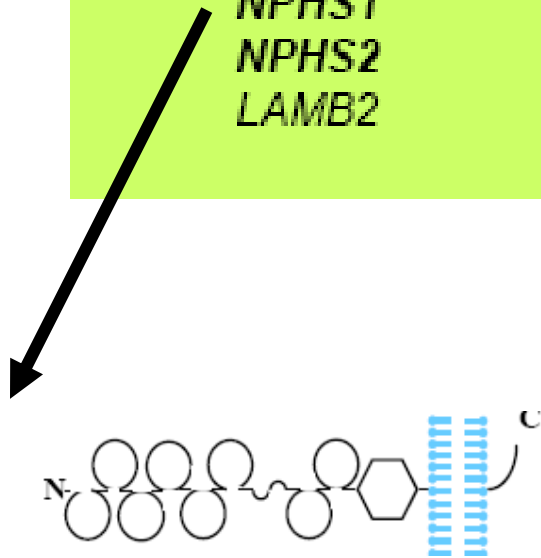
> 200 nm (F)
> 300 nm (H)
(basalopathie)

Fenêtre de l'endothélium





| SN cong | SNCR <15 ans | SMD | SNCR adulte |
|---|---|---|---------------------------------|
| <p><i>NPHS1</i> <i>NPHS2</i> <i>LAMB2</i></p> | <p><i>NPHS2</i> <i>NPHS1 - WT1</i> <i>NPHS3</i></p> | <p><i>NPHS3</i> <i>WT1</i> <i>LAMB2</i></p> | <p>R229Q (<i>NPHS2</i>)</p> |

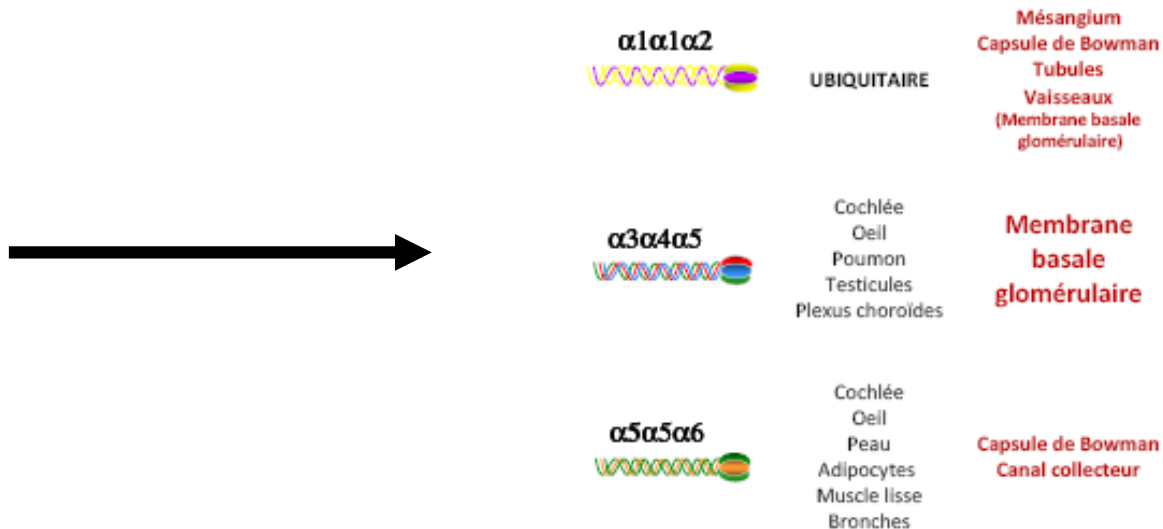


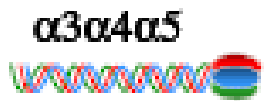
- Syndrome néphrotique autosomique récessif.
- 1/10 000 naissances en Finlande.
- Protéinurie massive à la naissance (à début prénatal), gros placenta oedémateux.
- Evolution spontanée sévère : mort dans les premières années d'IRT ou de complications intercurrentes.
- > Première démonstration, en 1998, du rôle fondamental du podocyte, et spécifiquement du diaphragme de fente, dans le maintien de la barrière de filtration glomérulaire.

- 57 mutations décrites dans les huit exons
- Mutation la plus fréquente : R138Q (32% des allèles mutés)
- Mutations du gène *NPHS2* dans 42% cas familiaux et 10% cas sporadiques
- Début en moyenne vers 4 ans (toujours <13 ans)
- Présence de mutations du gène *NPHS2* chez 37,5% des enfants ayant un SN avant l'âge de 1 an.

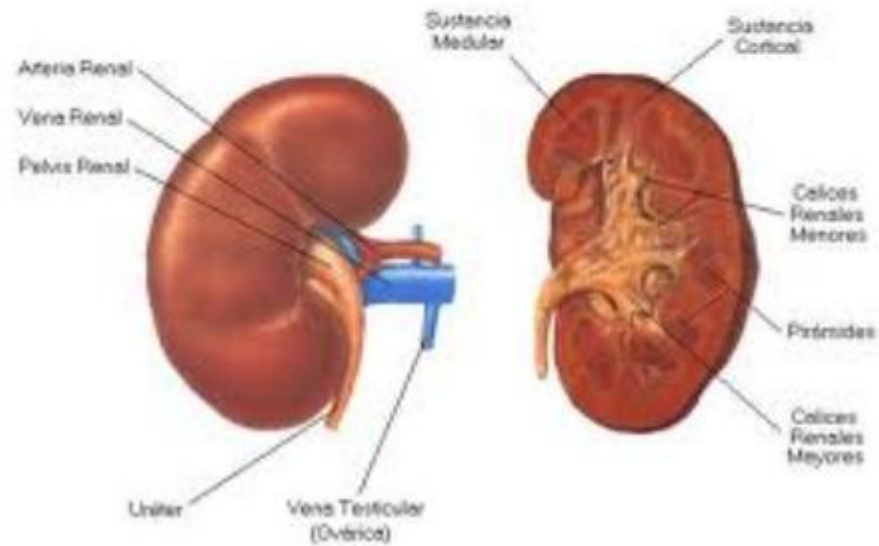
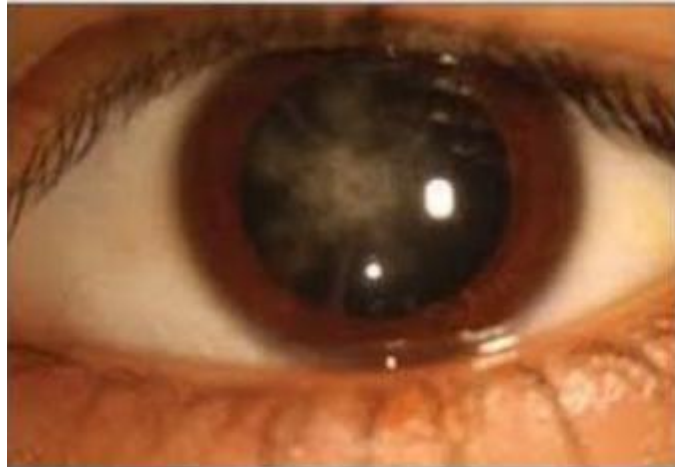
Constituants des membranes basales du glomérule et du mésangium

| | MBG | Capsule de Bowmann | Mésangium |
|--------------------|-----|--------------------|-----------|
| Coll IV | + | + | + |
| Laminine-1 | | + | + |
| Laminine-2 | | | + |
| Laminine-10 | | + | + |
| Laminine-11 | + | | |
| Agrine | + | | |
| Perlecan | | + | + |
| Bamacan | | + | + |
| Entactine/nidogène | | | + |

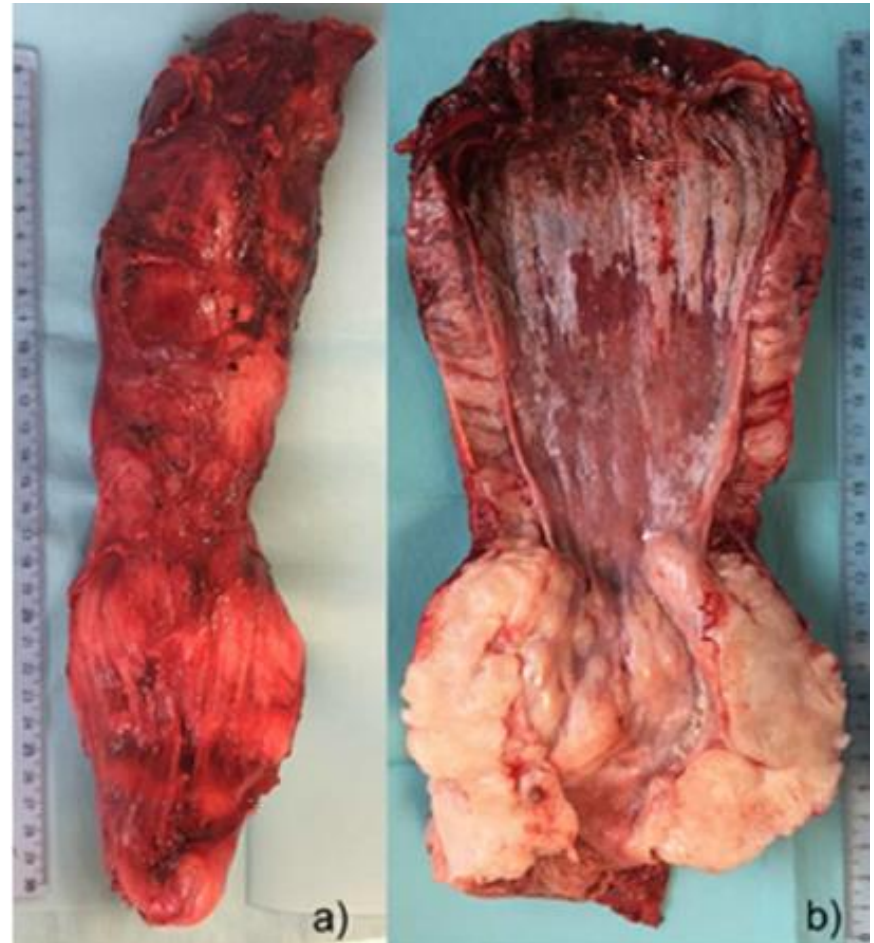
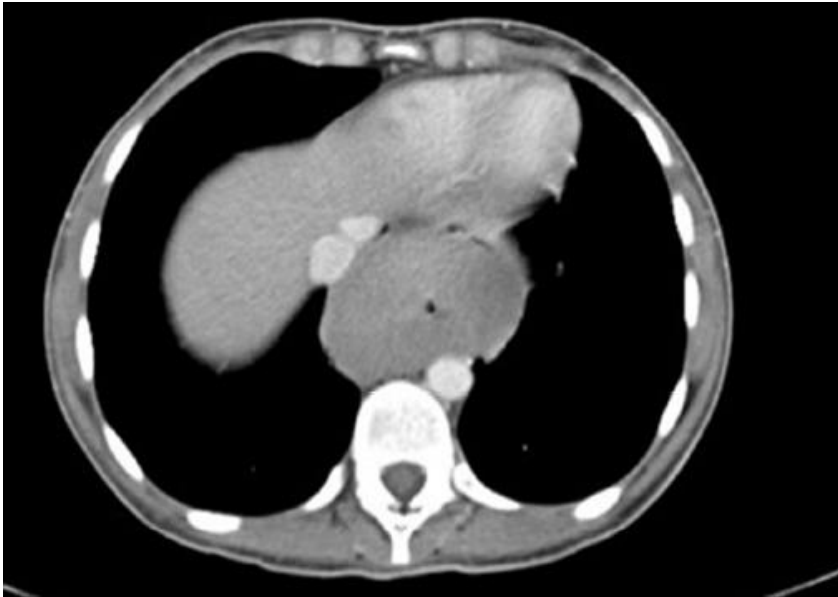




Cochlée
Oeil
Poumon
Testicules
Plexus choroïdes



Délétion dans le chromosome X qui affecte COL4A5 et COL4A6





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E-mail: smri@shigei.or.jp

Engl. Version 3.2

MONOCLONAL ANTIBODIES PRODUCED
BY THE RAT LYMPH NODE METHOD

Research use only

Fluorochrome-conjugated MoAbs for Alport's syndrome

FITC-Anti Collagen IV $\alpha 5(\text{IV})$ Chain, Human (Mono)
+ Texas Red-Anti Collagen IV $\alpha 2(\text{IV})$ Chain, Human (Mono)

Code: **CFT- 45325**

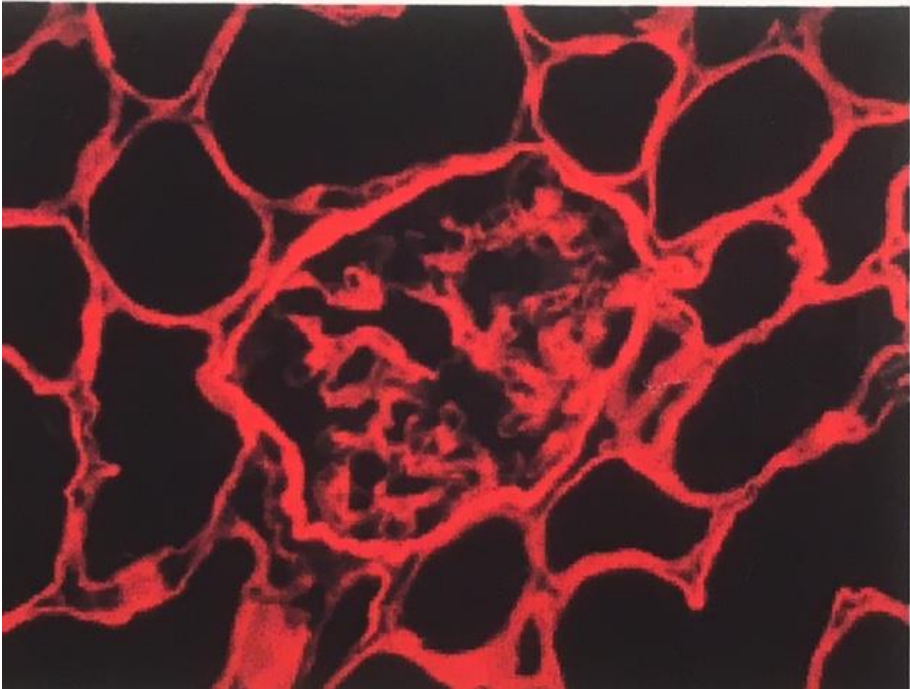
Volume: **1.0 ml**

Price: **JPY55,000**

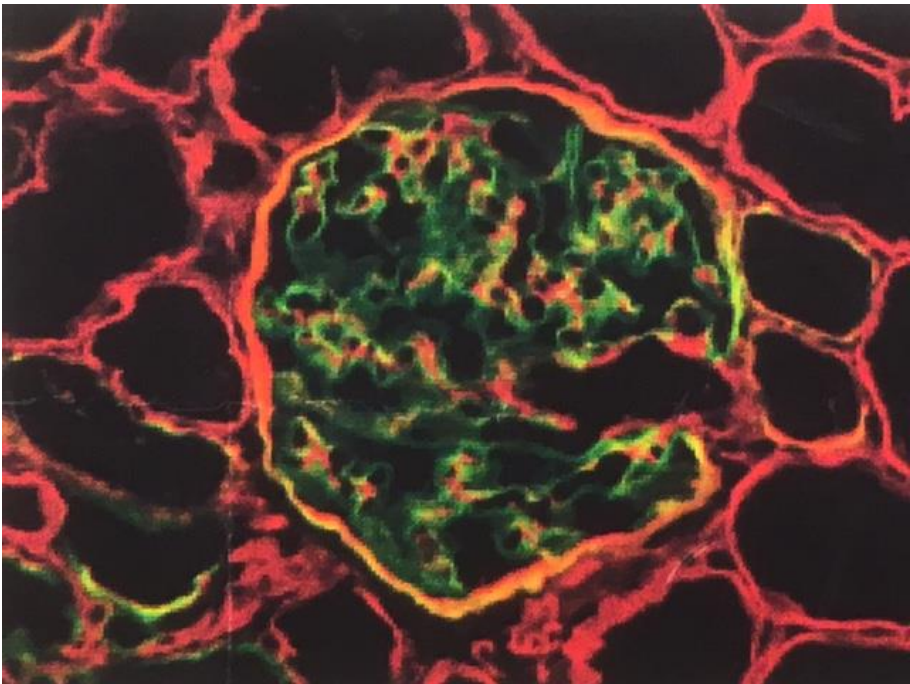
This product can be used for staining of human renal and skin biopsy sections very easily and rapidly because two monoclonal antibodies are conjugated with two different fluorochromes. FITC-conjugated-anti $\alpha 5(\text{IV})$ demonstrates presence or absence of the $\alpha 5(\text{IV})$ chain, and Texas-Red-anti $\alpha 2(\text{IV})$ reveals the renal basement membrane structure.

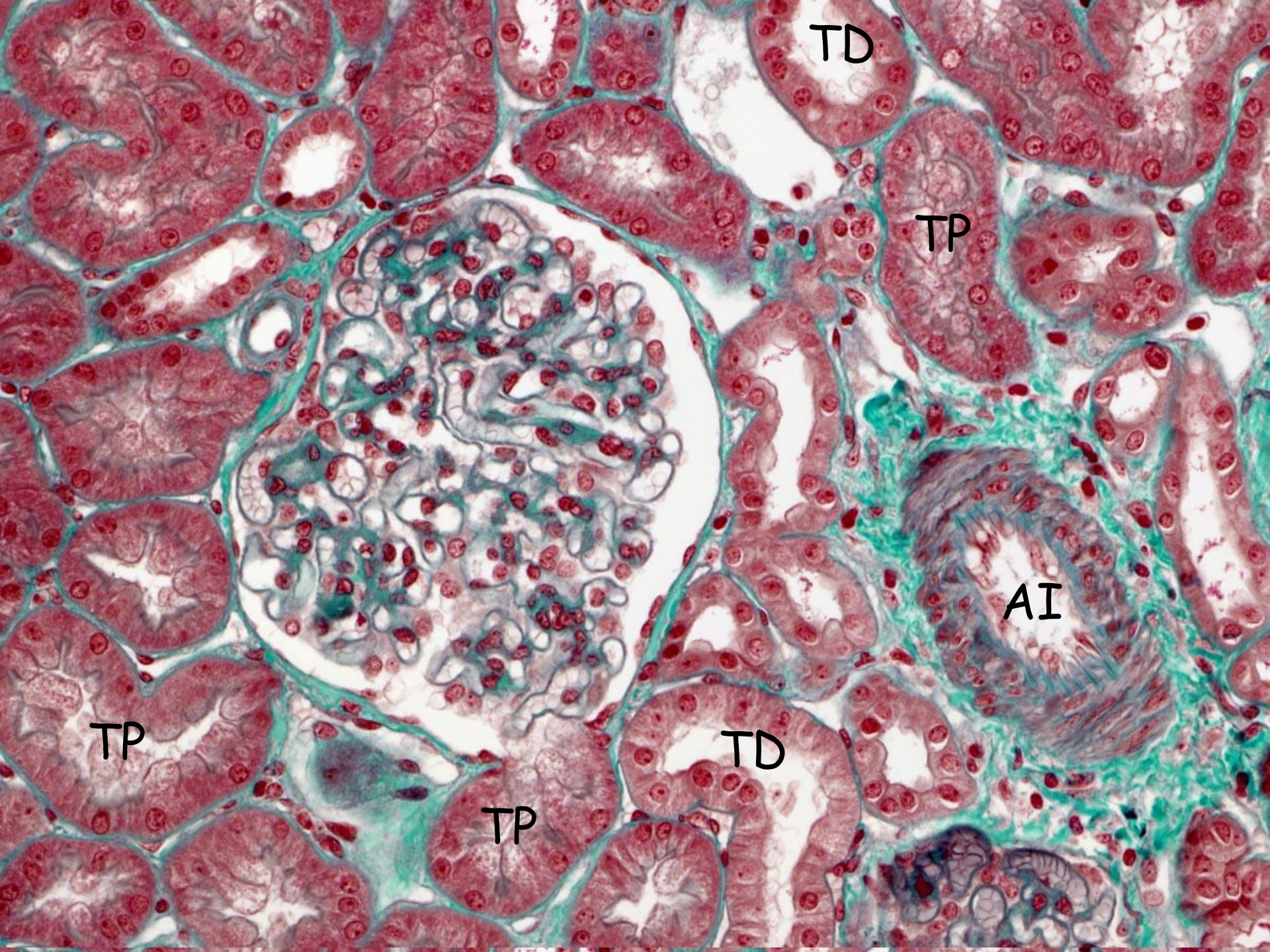
Alport's syndrome, an inherited disease, shows absence or reduction of the $\alpha 5(\text{IV})$ chain in the glomerular, tubular and Bowman's capsular basement

Alpha 2



Alpha 5





TD

TP

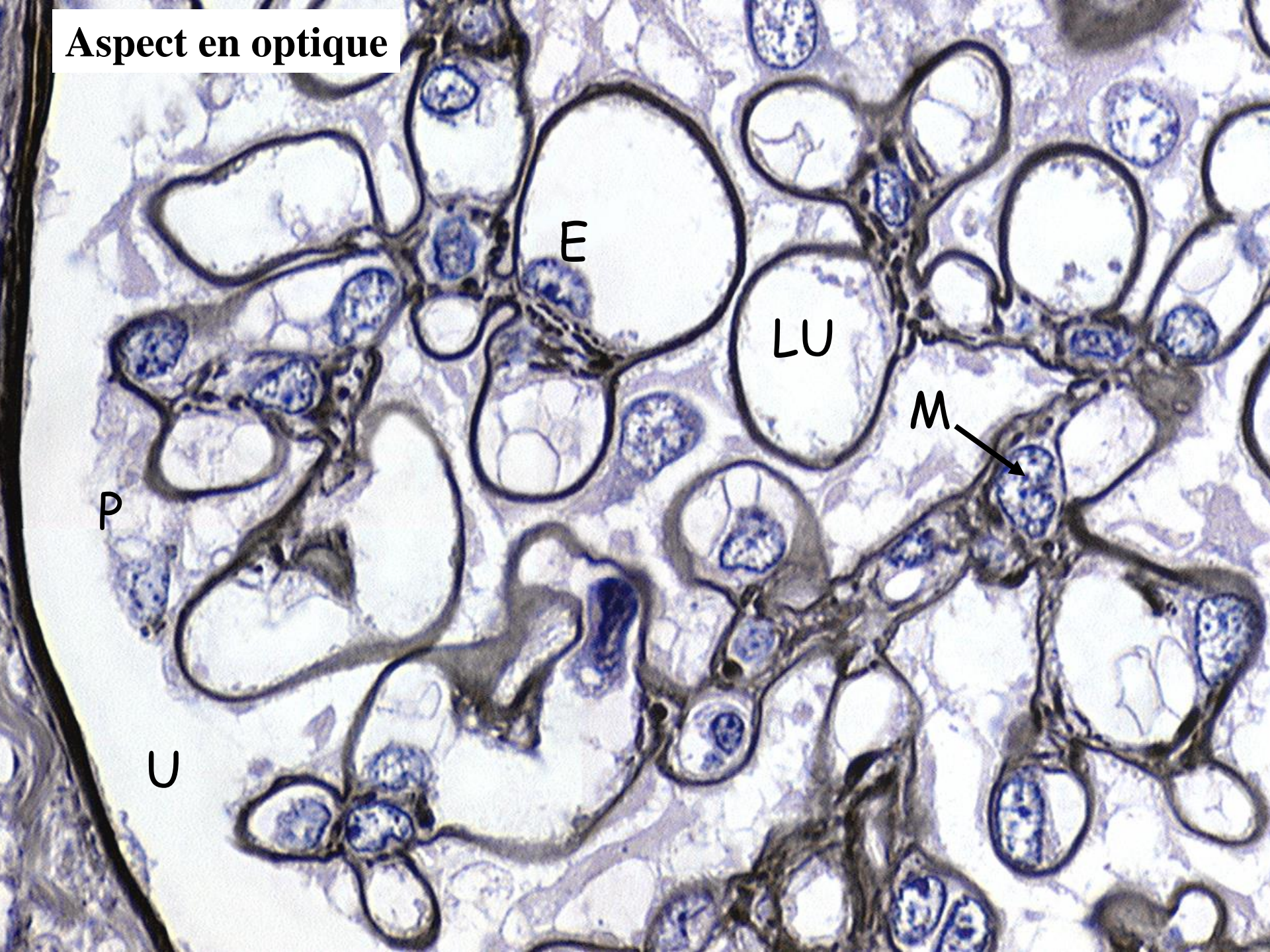
AI

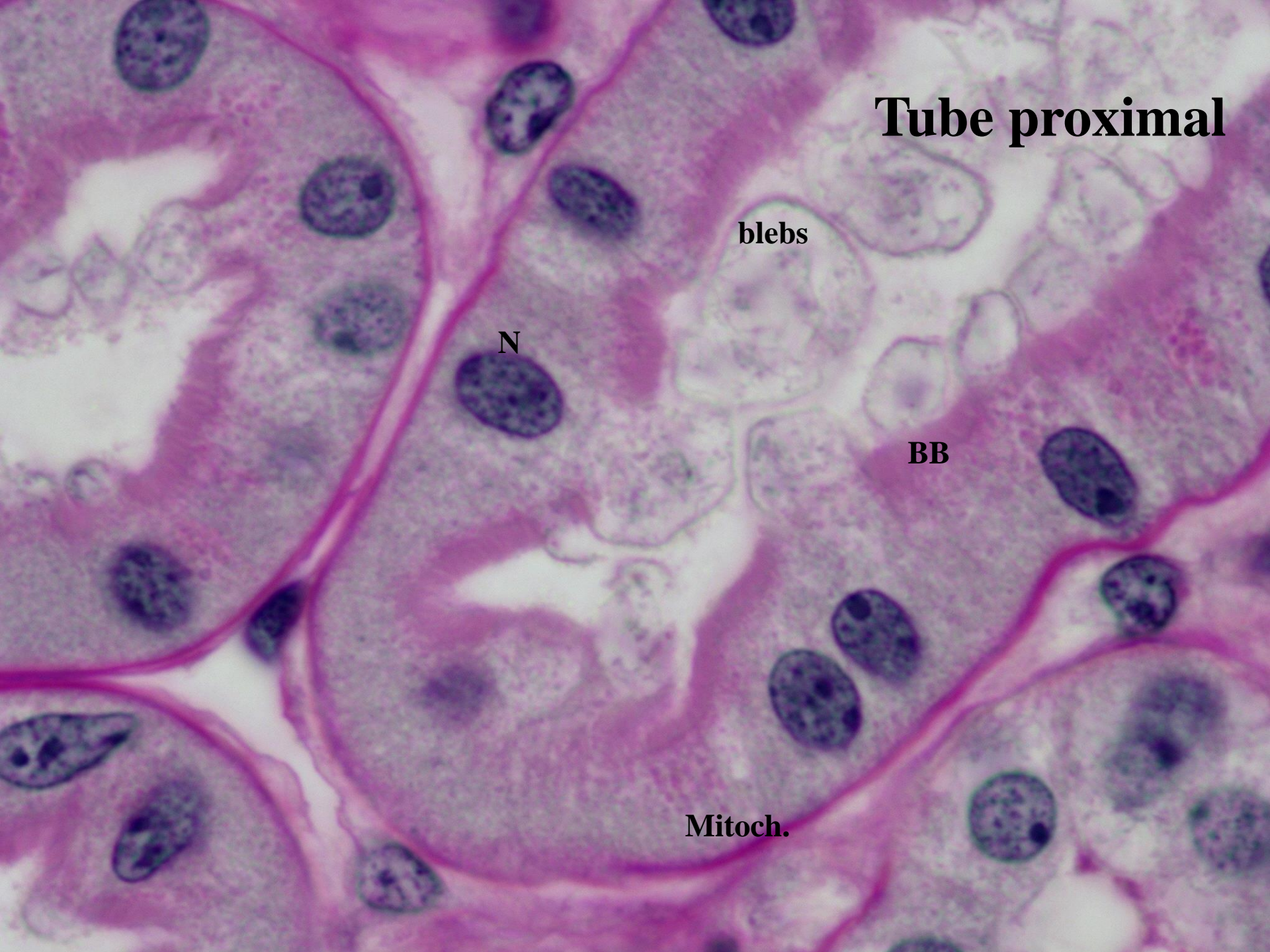
TP

TD

TP

Aspect en optique





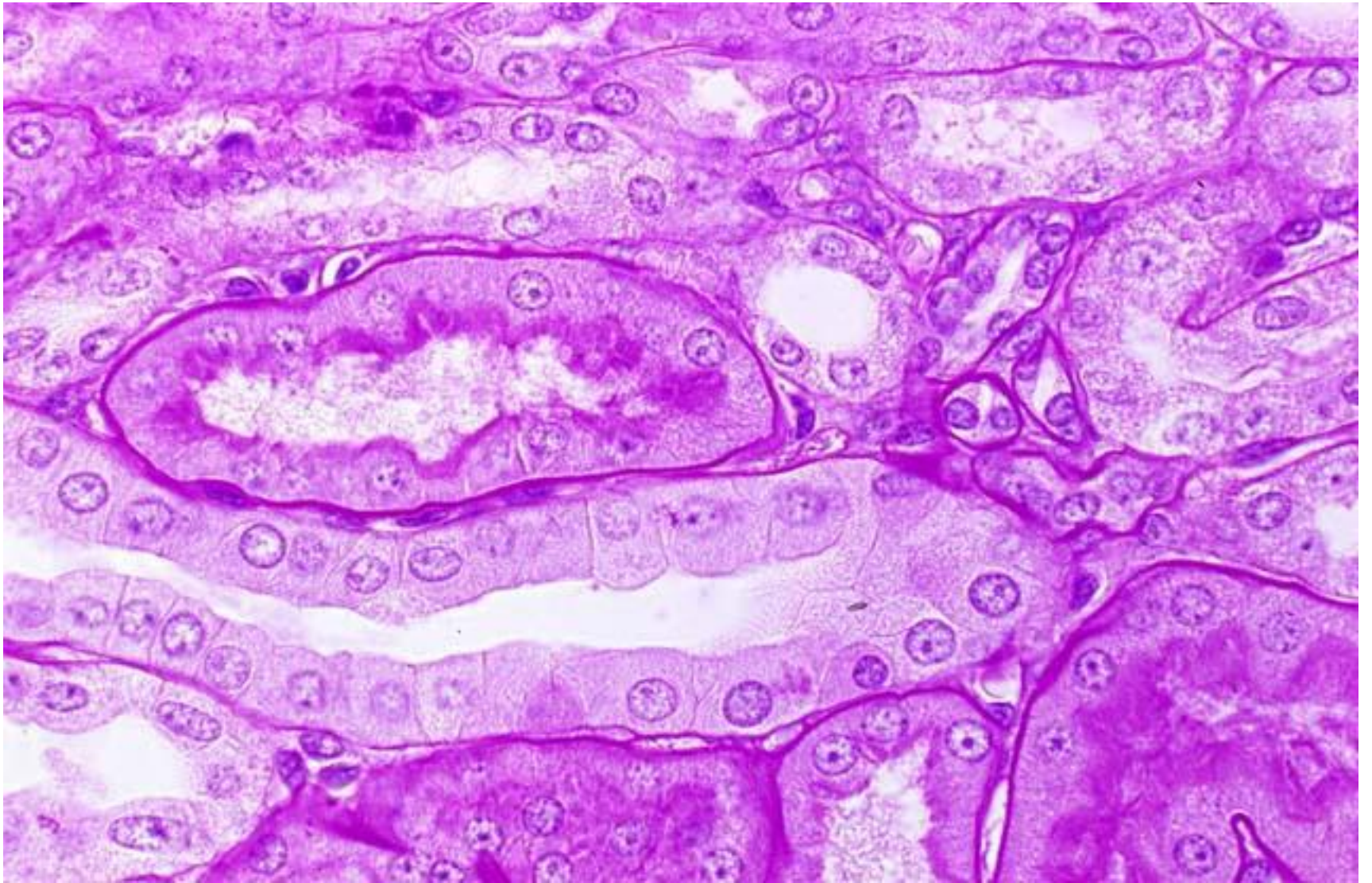
Tube proximal

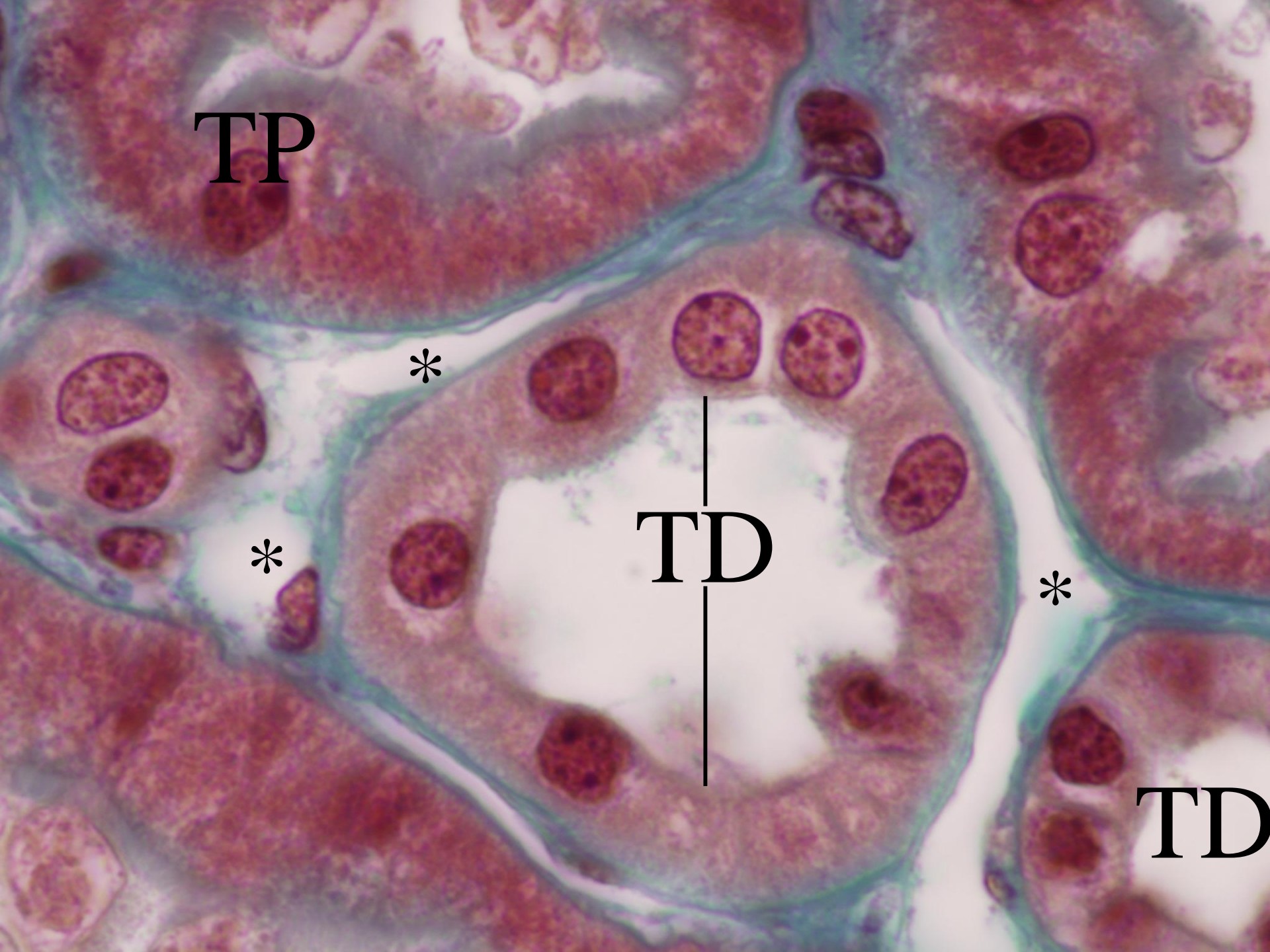
blebs

N

BB

Mitoch.





TP

*

TD

*

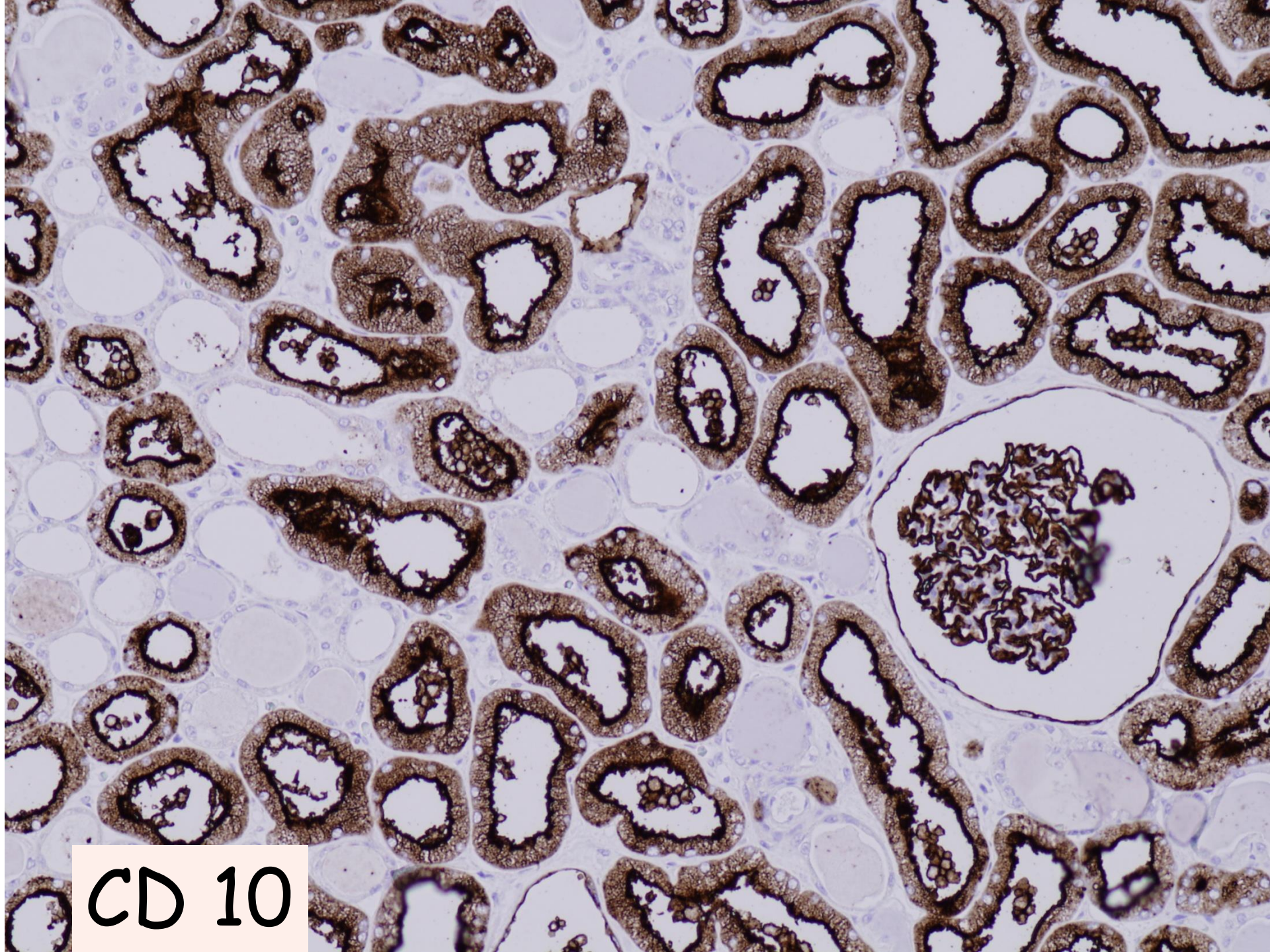
*

TD

Tableau 3 : principaux antigènes des cellules tubulaires

| Antigène | Tube proximal | | Anse de Henlé | Tube distal | | | Canal collecteur |
|-----------|--------------------|----------------|---------------|----------------|--------------|--------------------|------------------|
| | Portion contournée | Portion droite | | Portion droite | Macula densa | Portion contournée | |
| CK AE1AE3 | - | - | + | + | - | - | + |
| CK7 ← | - | - | + | + | - | + | +/- |
| CK8 | + | + | + | + | + | + | + |
| CK 34bE12 | - | - | - | - | - | - | + |
| EMA | - | - | + | + | + | + | +/- |
| E- cad | - | - | + | + | + | + | + |
| CD 10 ← | + | + | - | - | - | - | - |
| CD138 | + | - | - | - | - | - | - |

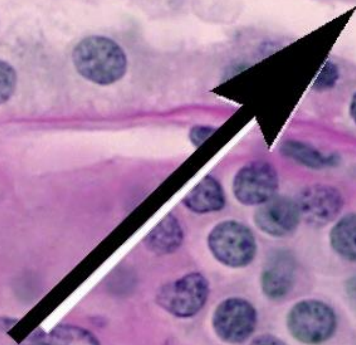
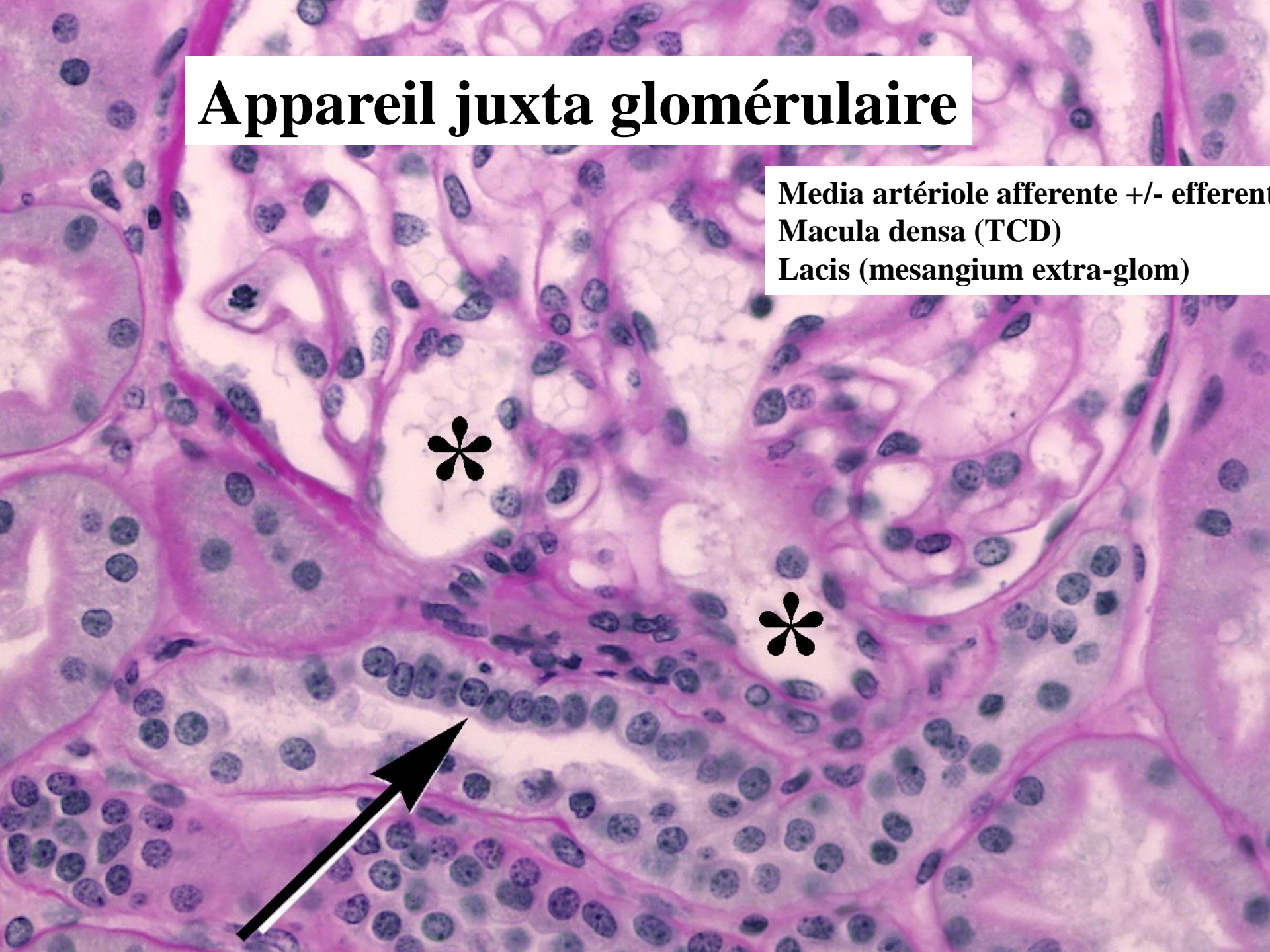
Lésions souvent non spécifiques: Atrophie, inflammation, dystrophie,.....
Marquages de peu d'utilité en pathologie non tumorale.....

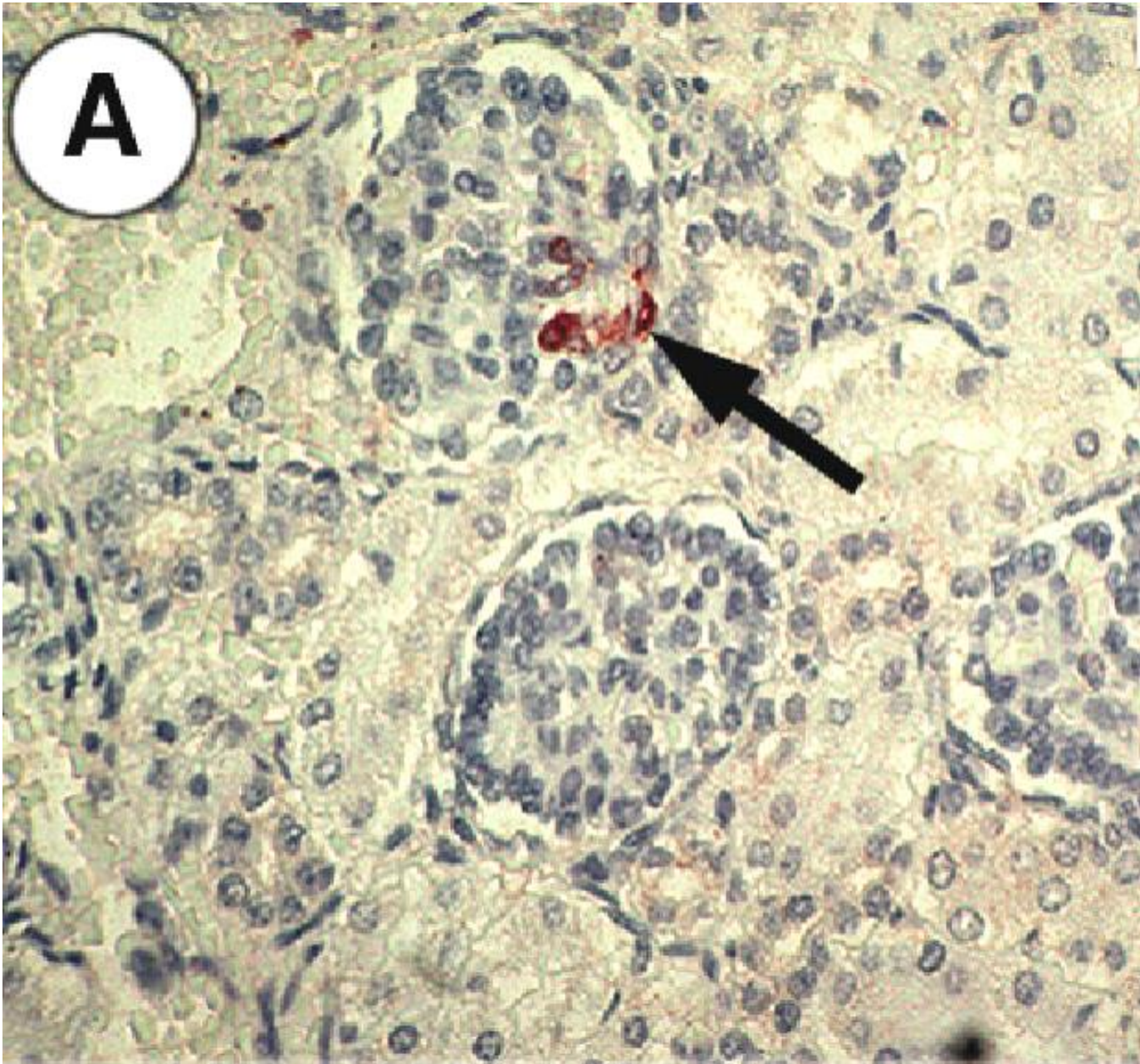


CD 10

Appareil juxta glomérulaire

Media artériole afferente +/- efferente
Macula densa (TCD)
Lacis (mesangium extra-glom)

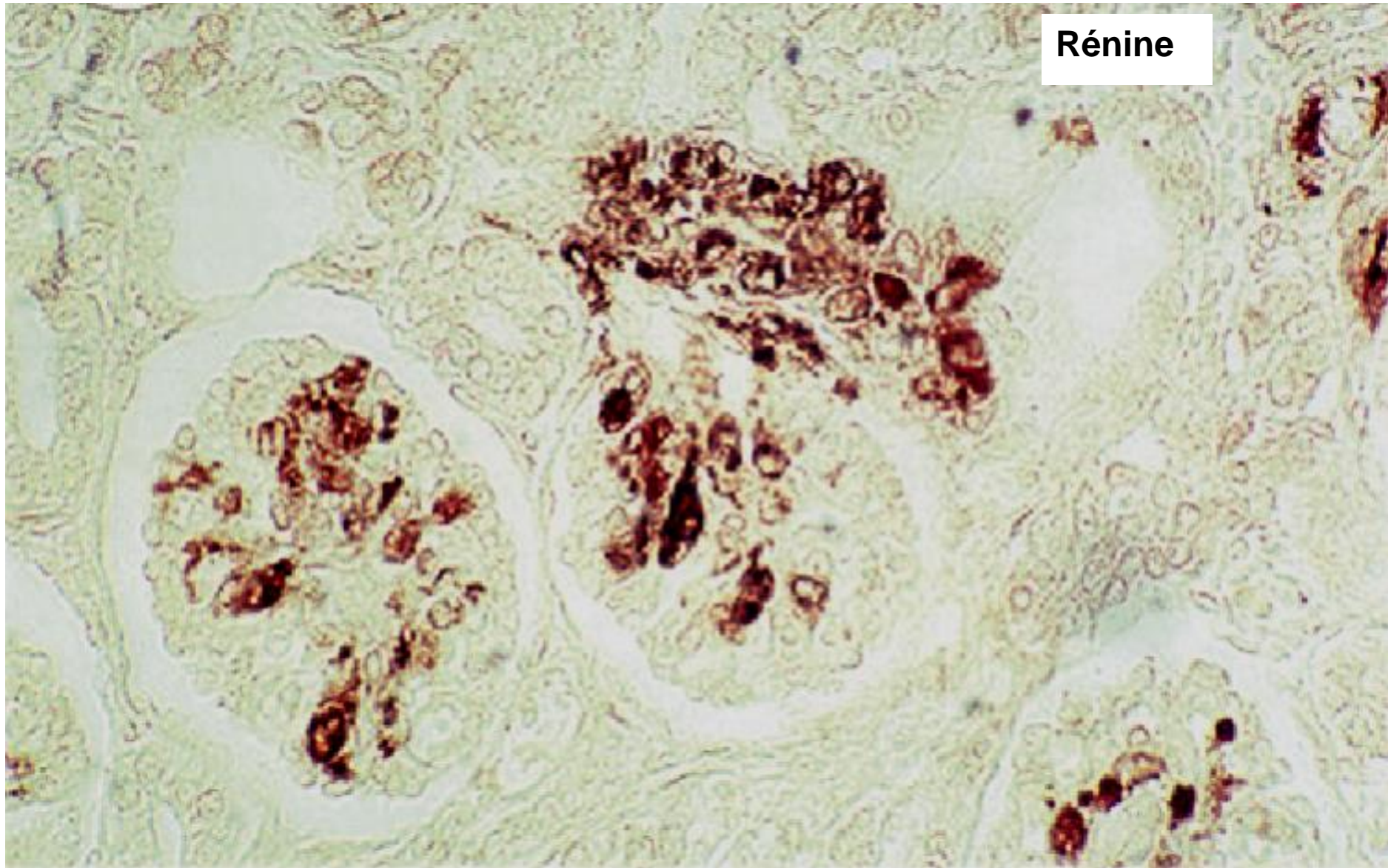




Rénine

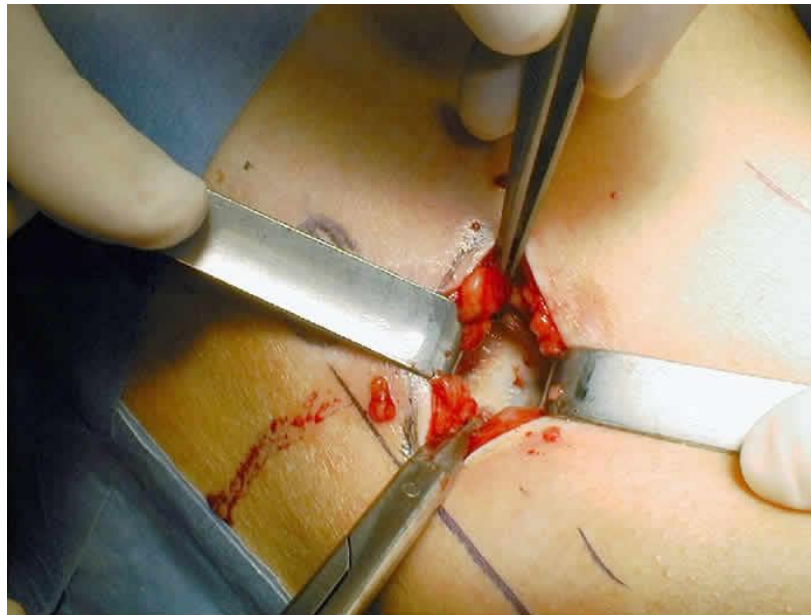
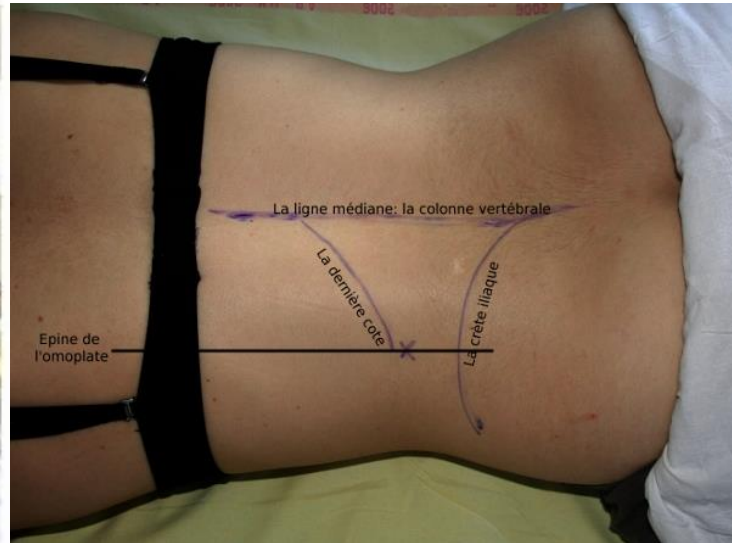
Normal

Rénine



Cas de dysgénésie tubulaire

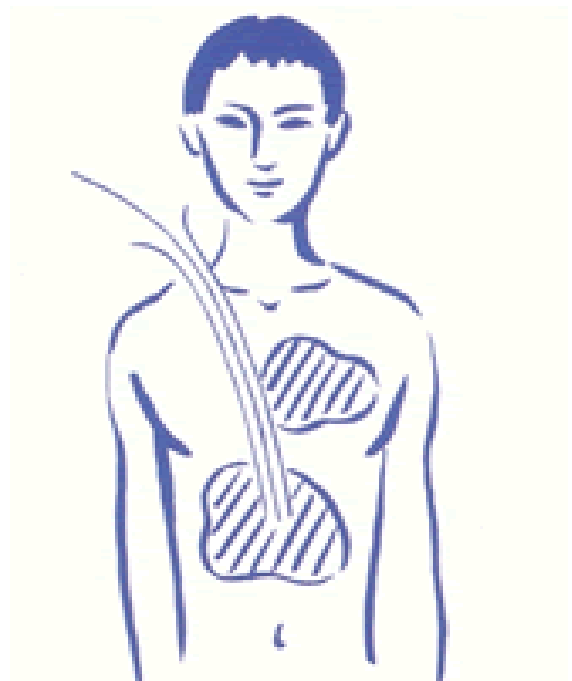
La Pratique de la biopsie rénale.....



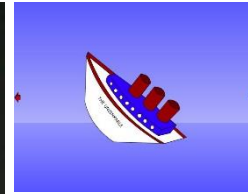
- **Diamètre (gauge).**
- **Nombre de tir.**
- **Efficacité.**
- **Transport.**
- **Causes de prb.....**



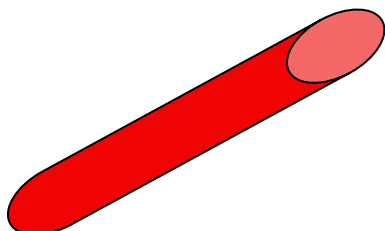
a.







**Biopsie souvent
collée dans le
bouchon.....
Si le fragment
flotte ce n'est pas
du rein....**



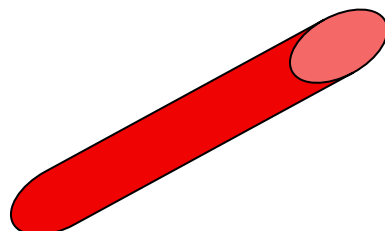
Formol (ou AFA)



**24 h (standard)
12 h (cours)
6 h (Xpress)**

Trop imprécis !

HES, Jones



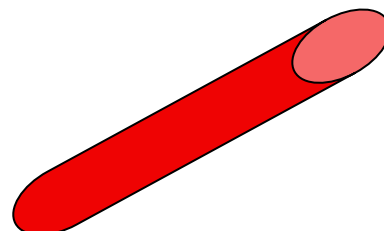
Glutaraldéhyde



**72 h
144 h (6 jours)**

Trop long !

**Trichrome, Jones
+/- ME**



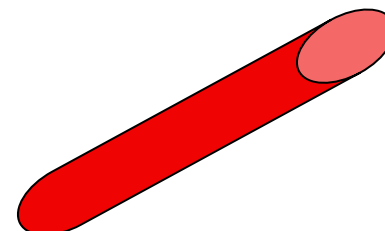
Liquide de Michel



IF

pas de morphologie !

7 anticorps



**Frais dans compresse
Imbibée de sérum phy**

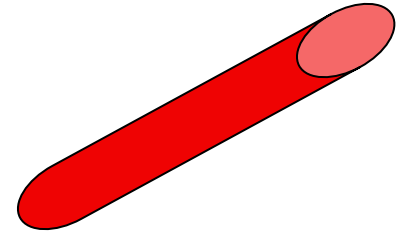
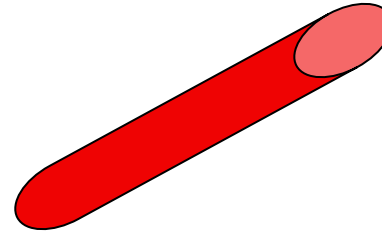
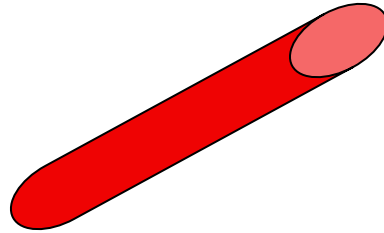
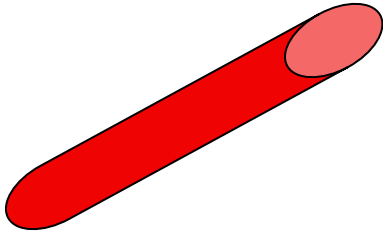
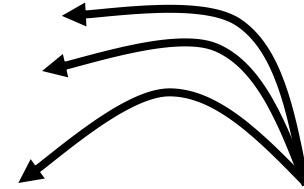


**Au choix
(IF)**

ça sèche !

Au choix...

Bref le néphropathologiste n'est par essence jamais content.....



Formol ou MF

Glutaraldéhyde

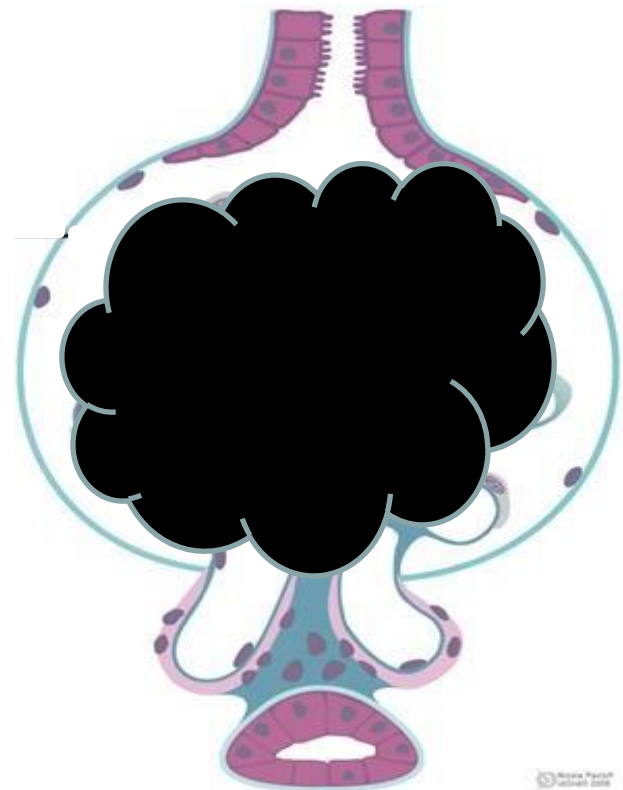
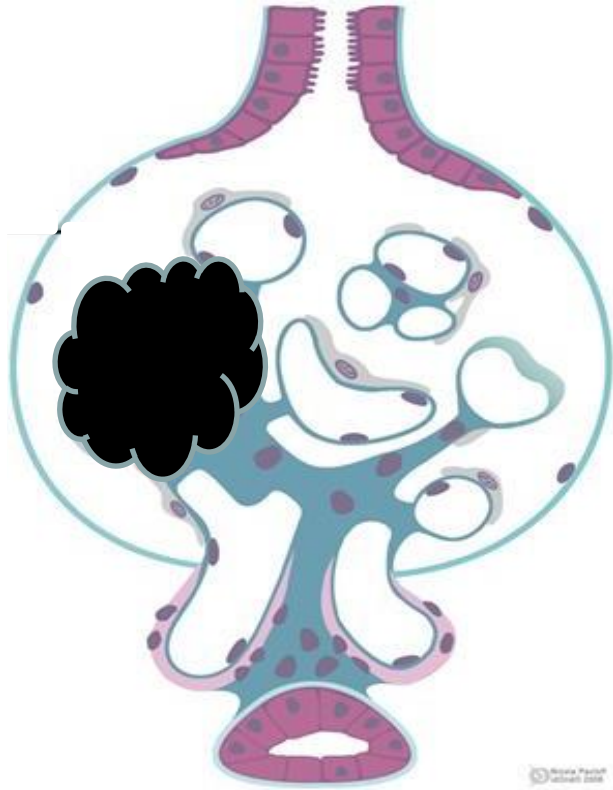
Liquide de Michel

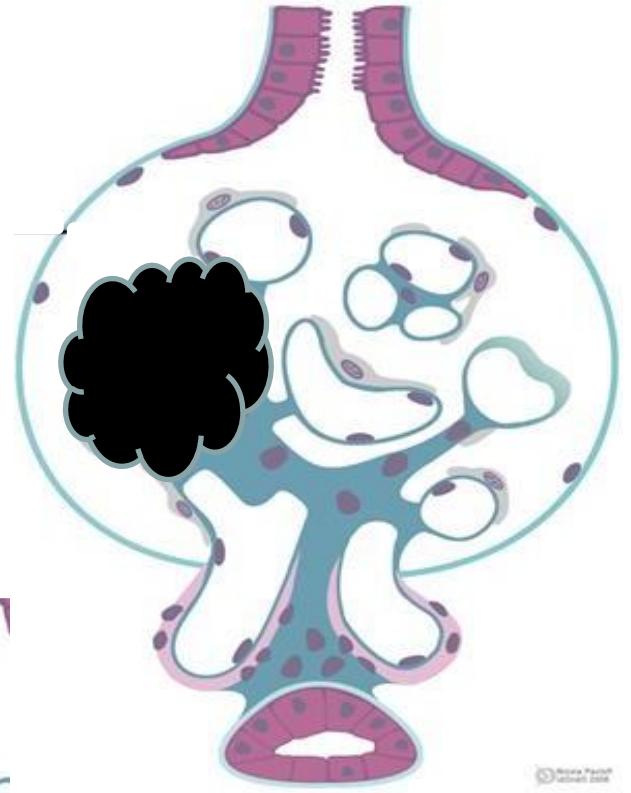
**Frais dans compresse
Imbibée de sérum phy**



Lésions élémentaires glomérulaires

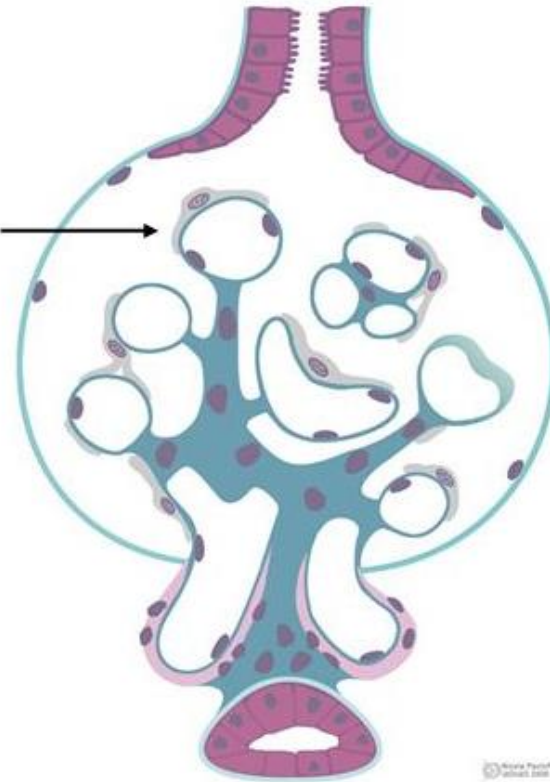
- Le bilan des lésions donne un type de glomérulopathie.**
 - Formes primaires versus secondaires.**
 - lésions segmentaires/globales**
 - Lésions focales/diffuses**



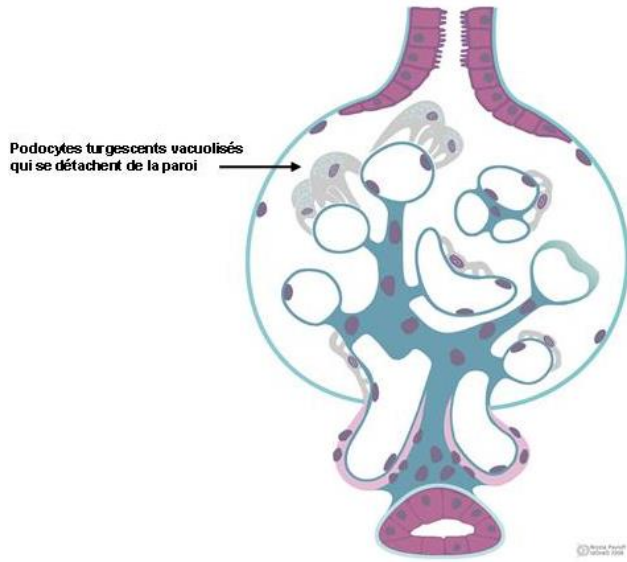


Glomérulopathie de type Lésions Glomérulaires Minimales

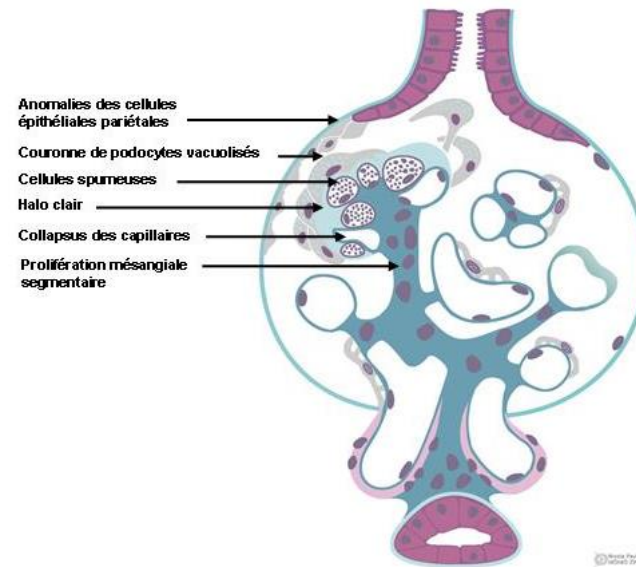
Étalement des pédicelles
des podocytes
(visible en microscopie
électronique)



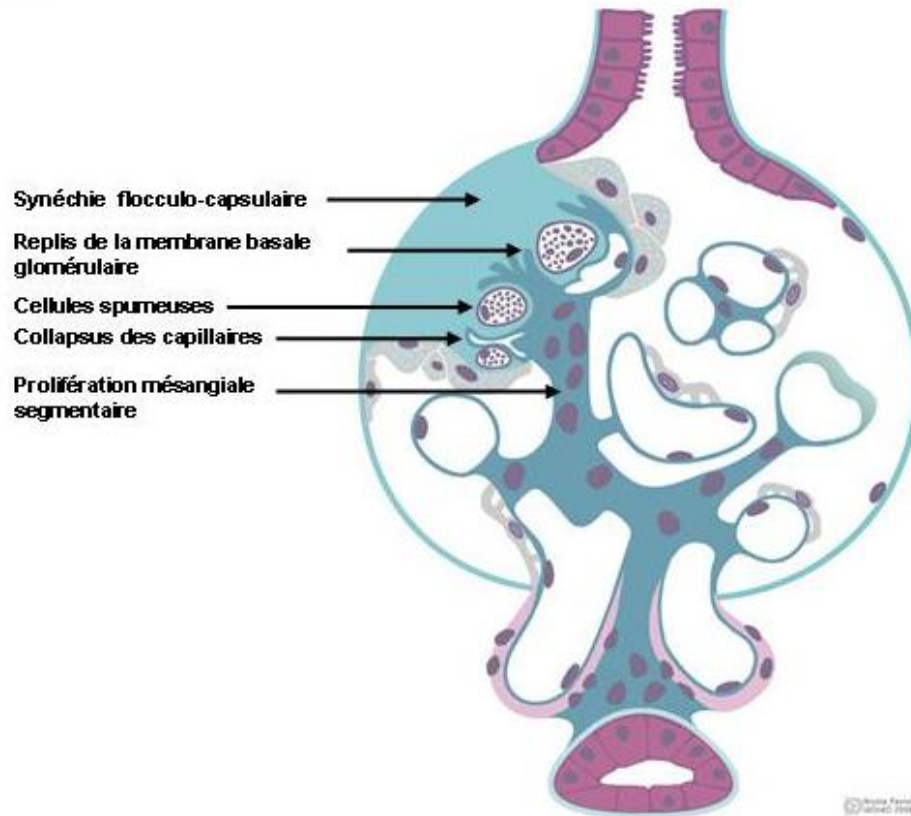
Glomérulopathie de type Hyalinose Segmentaire et Focale : 1^{er} stade



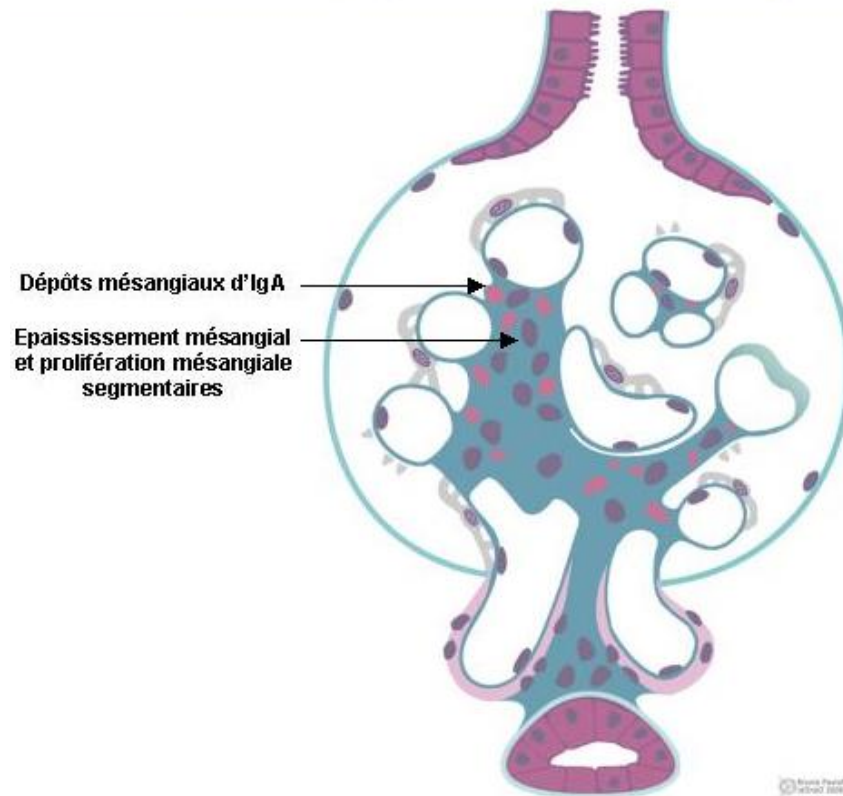
Glomérulopathie de type Hyalinose Segmentaire et Focale : 2^e stade



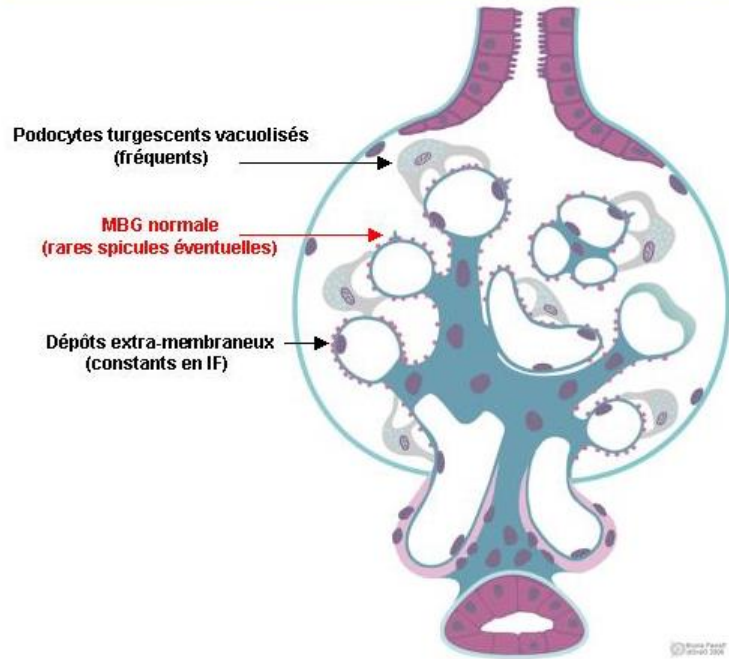
Glomérulopathie de type Hyalinose Segmentaire et Focale : 3e stade



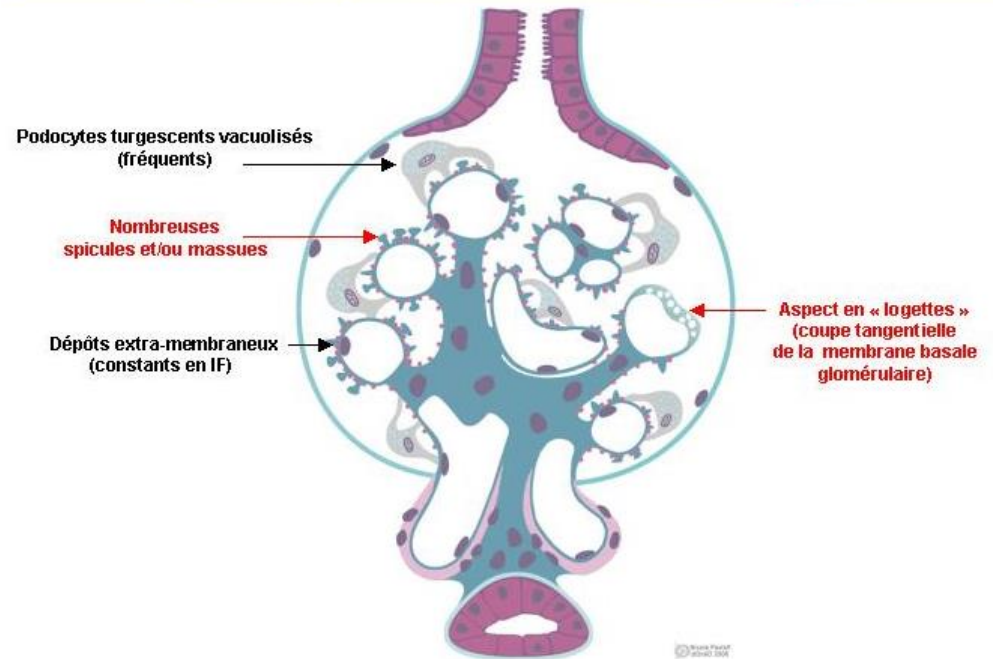
Glomérulopathie à dépôts mésangiaux d'IgA



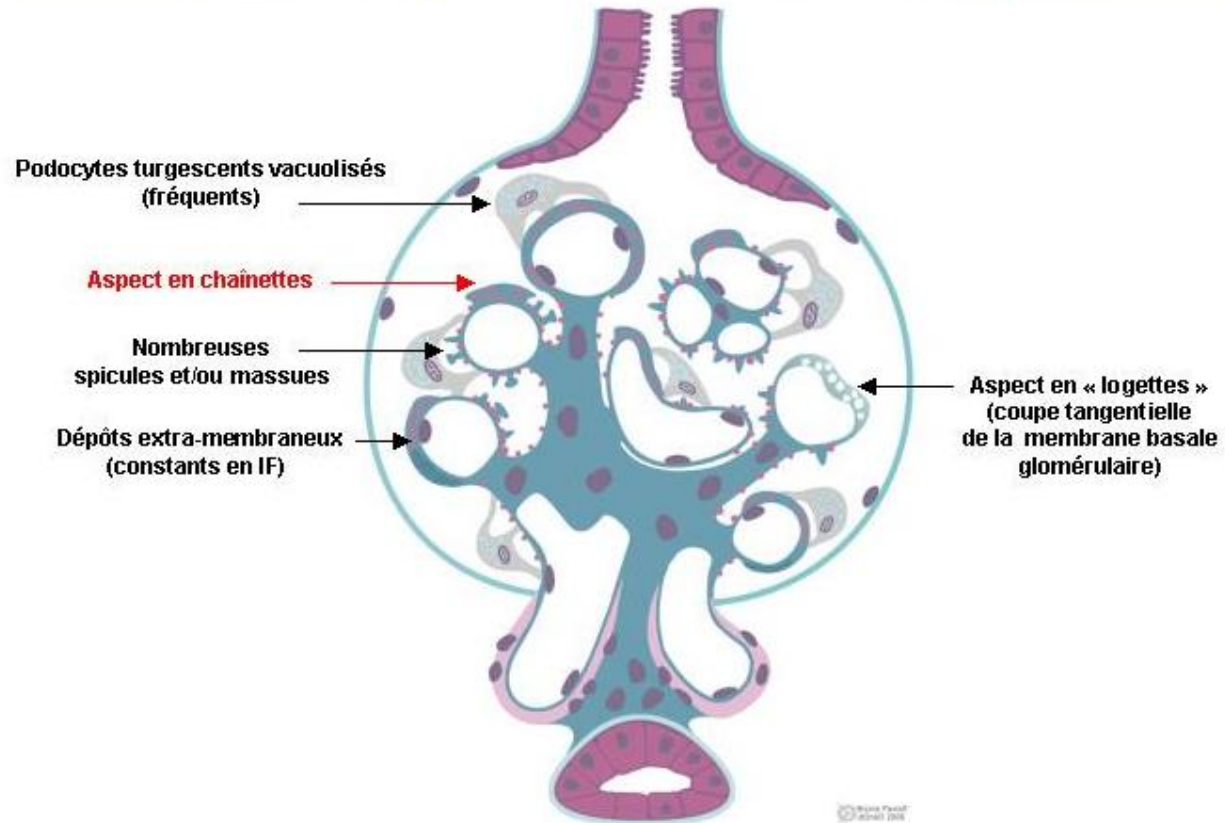
Glomérulonéphrite extra-membraneuse de **type I**



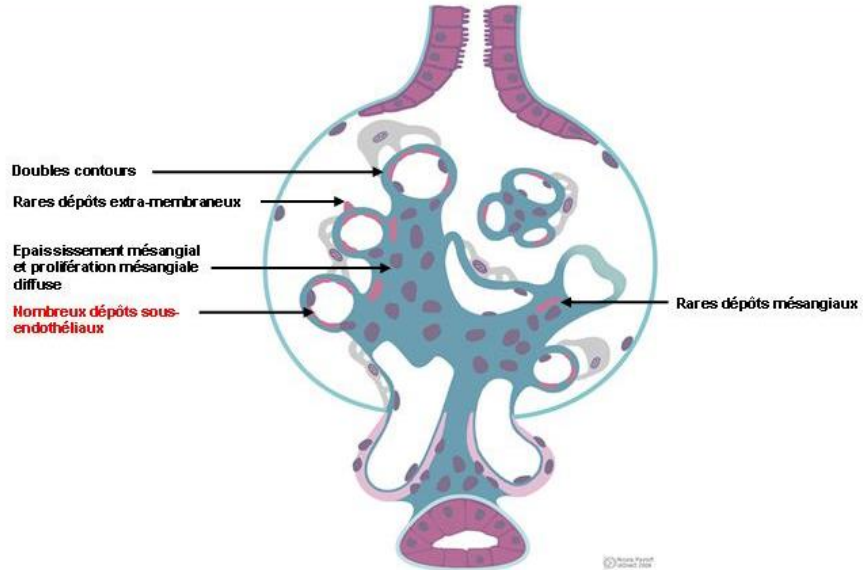
Glomérulonéphrite extra-membraneuse de **type II**



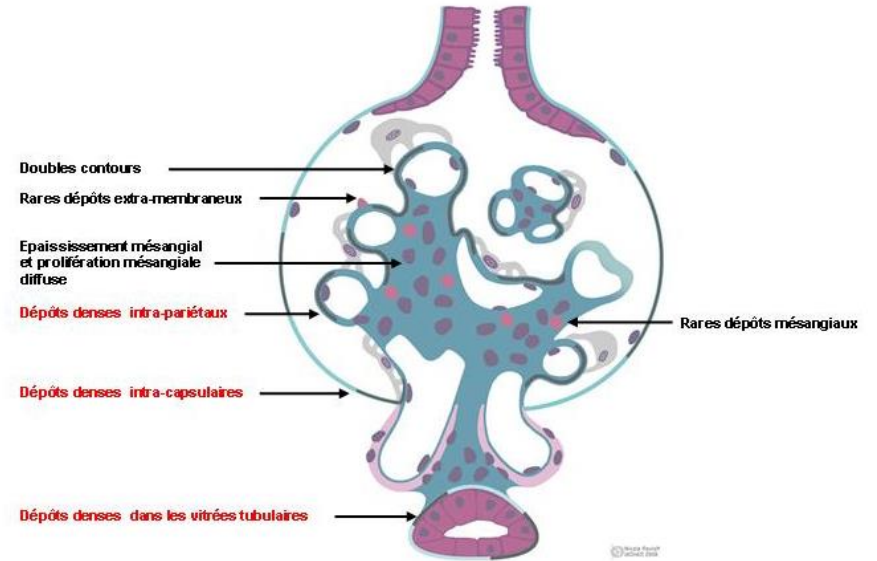
Glomérulonéphrite extra-membraneuse de **type III**



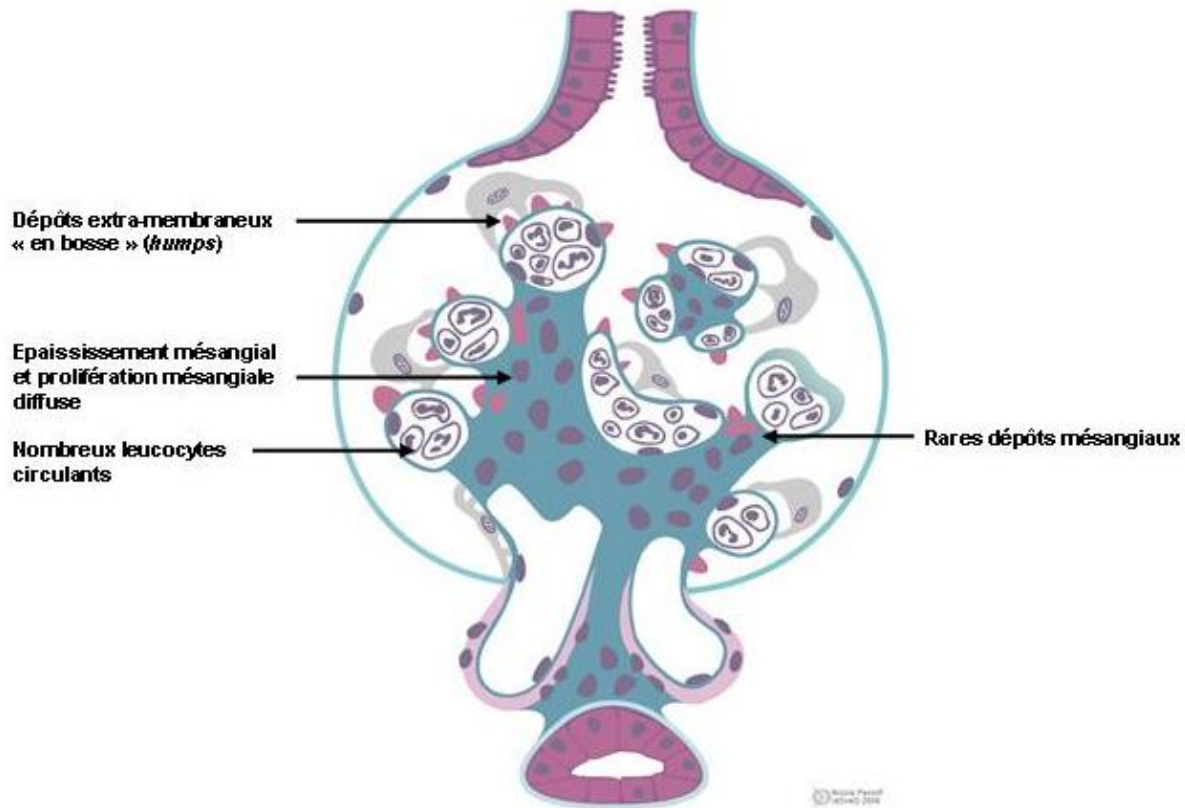
Glomérulonéphrite Membranoproliférative de **type I**



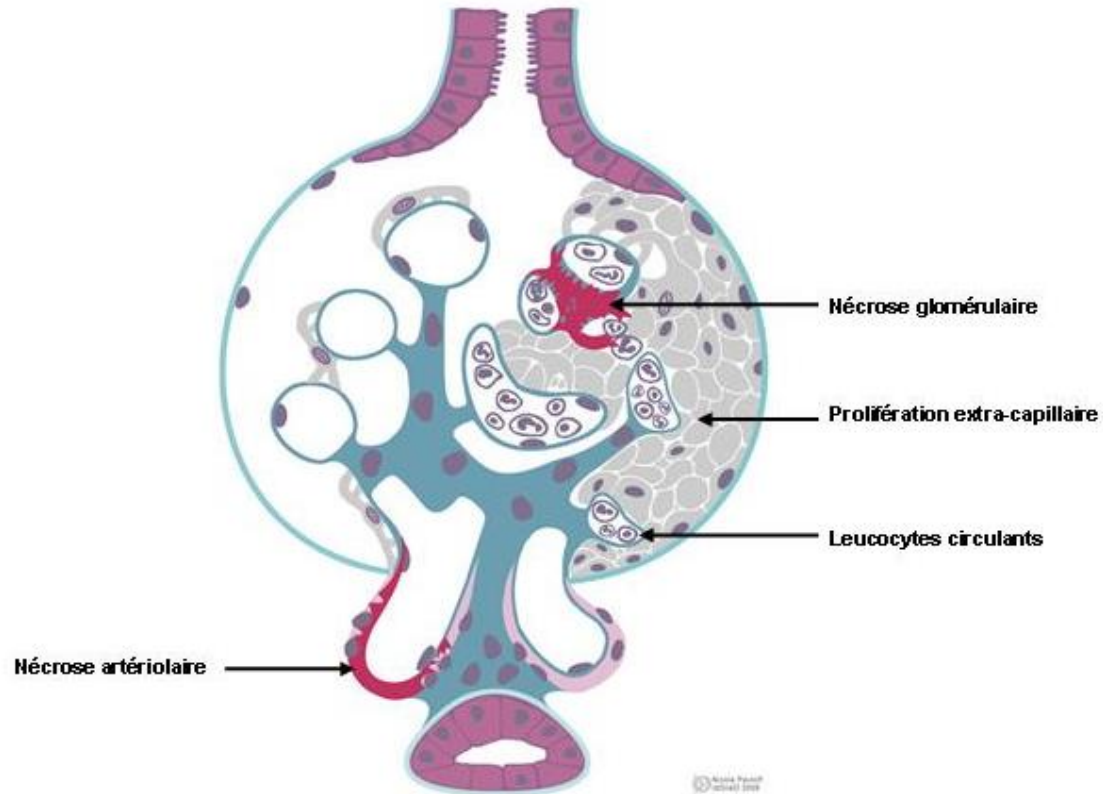
Glomérulonéphrite Membranoproliférative de **type II**



Glomérulonéphrite aiguë post-infectieuse



Glomérulonéphrite Nécrisante à Croissants



Glomérulonéphrite lupique de classe III ou IV

Lésions chroniques

Synéchie flocculo-capsulaire

Sclérose glomérulaire

Croissant fibreux cicatriciel

Lésions actives

Nécrose glomérulaire

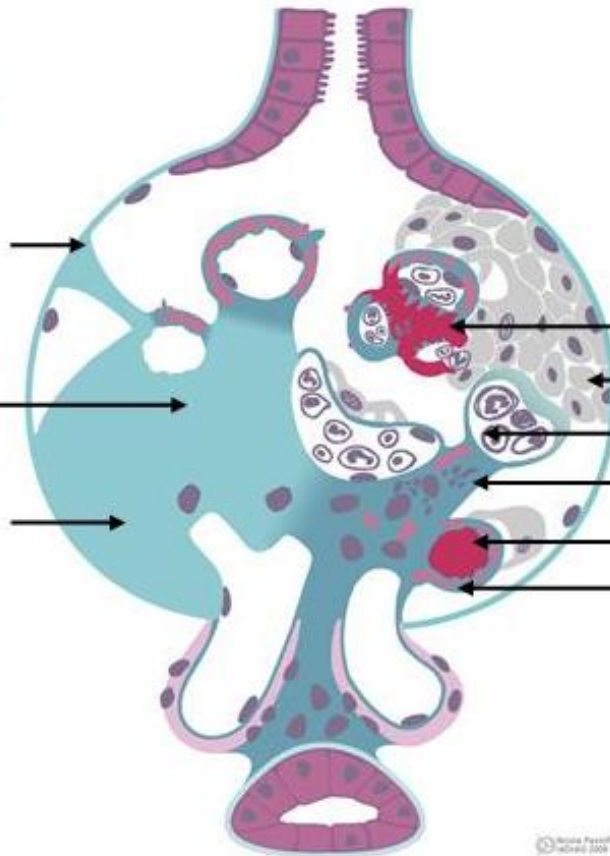
Prolifération extra-capillaire

Leucocytes circulants

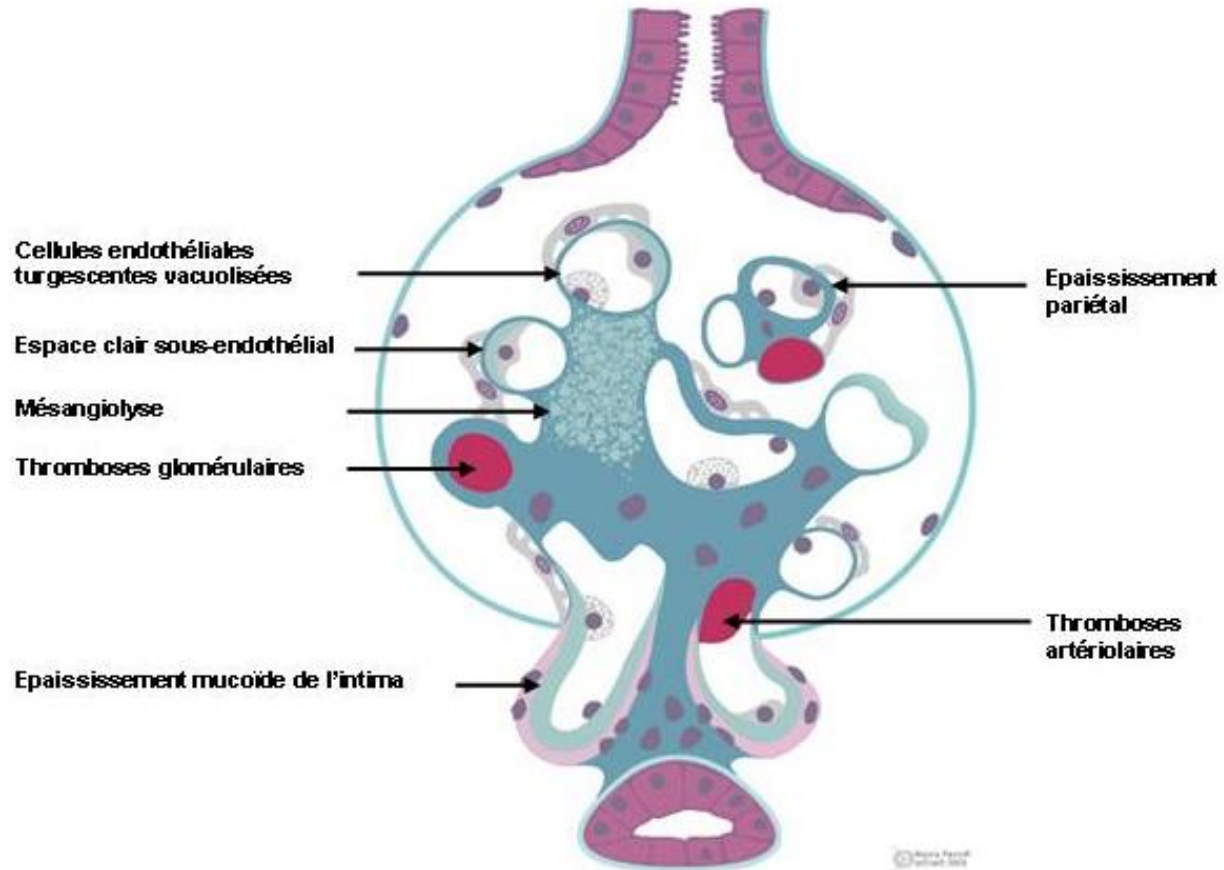
Karyorrhexis

Aggrégat immun (thrombus)

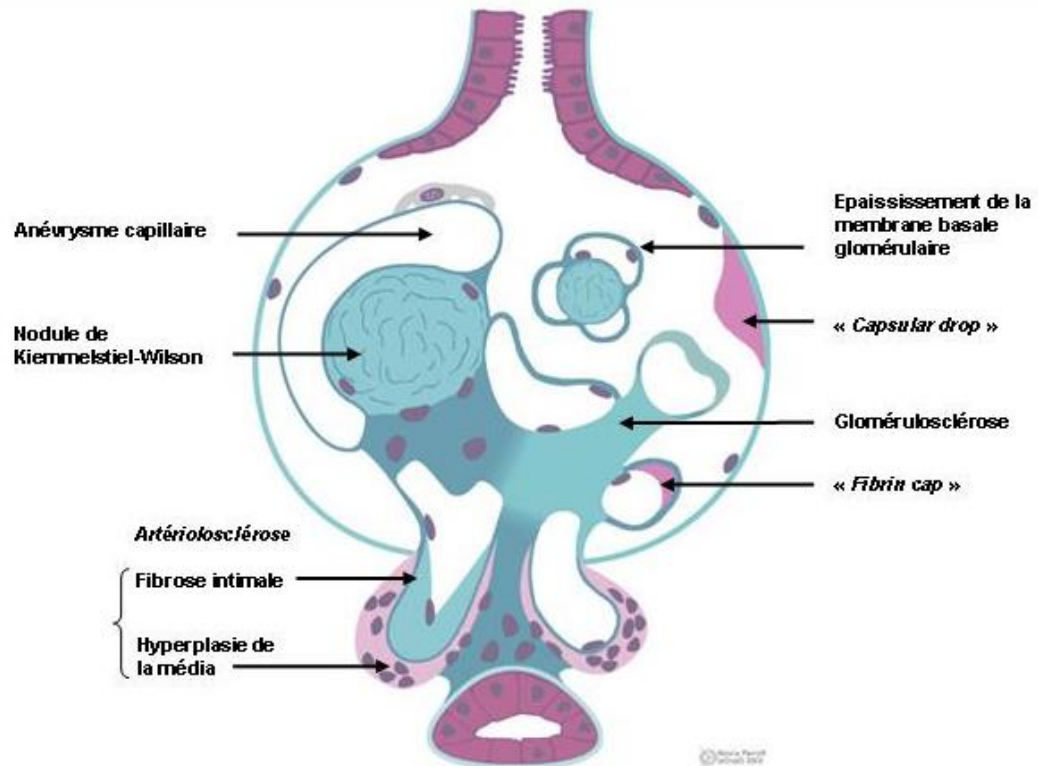
Anses en fil-de-fer
(Wire-loops)



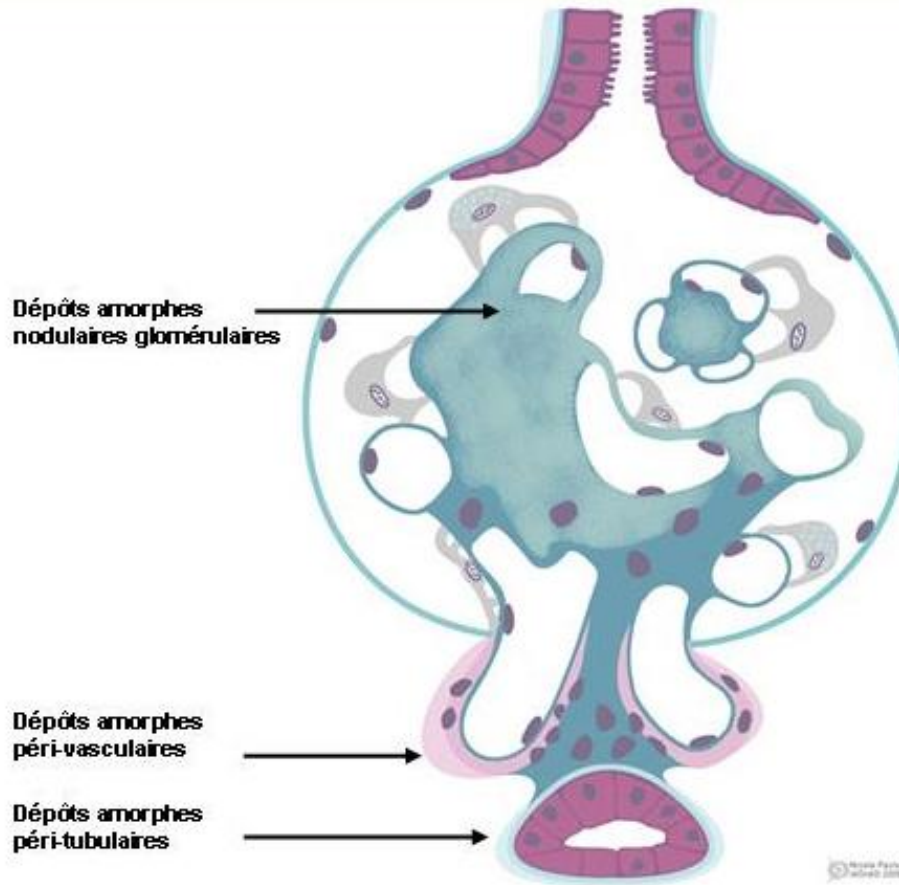
Microangiopathie thrombotique



Glomérulopathie diabétique



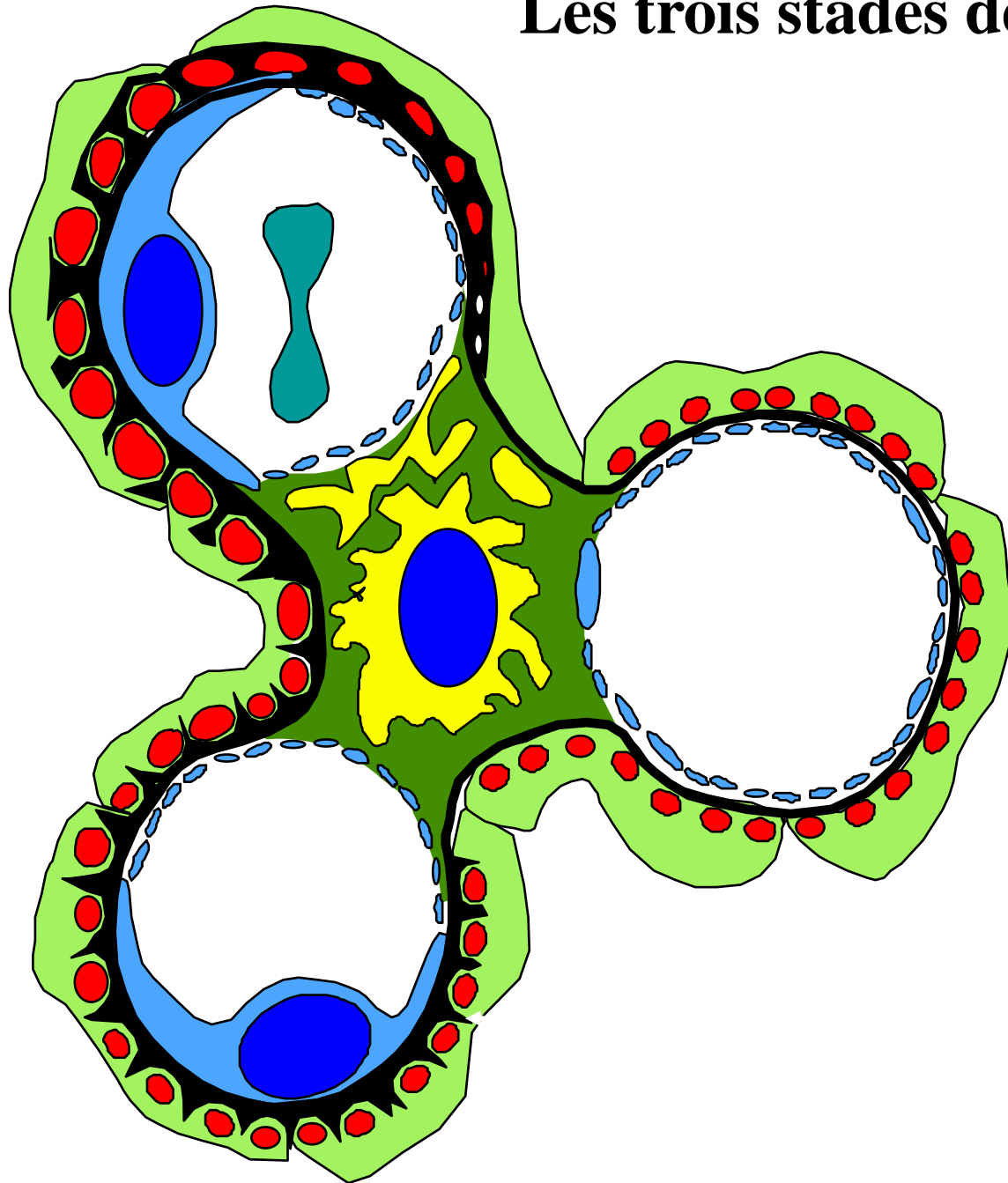
Amylose



G.E.M

Les trois stades de la GEM

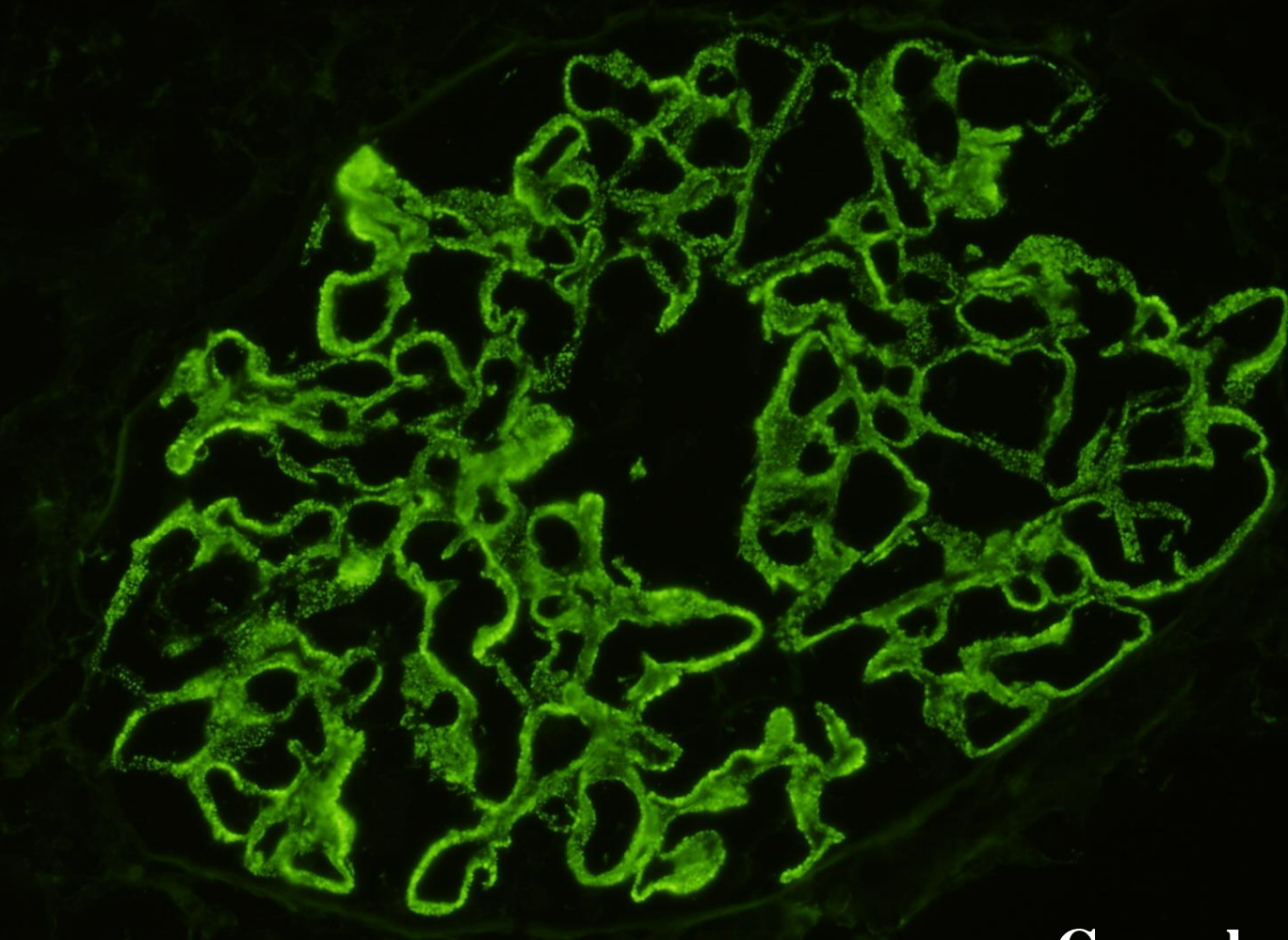
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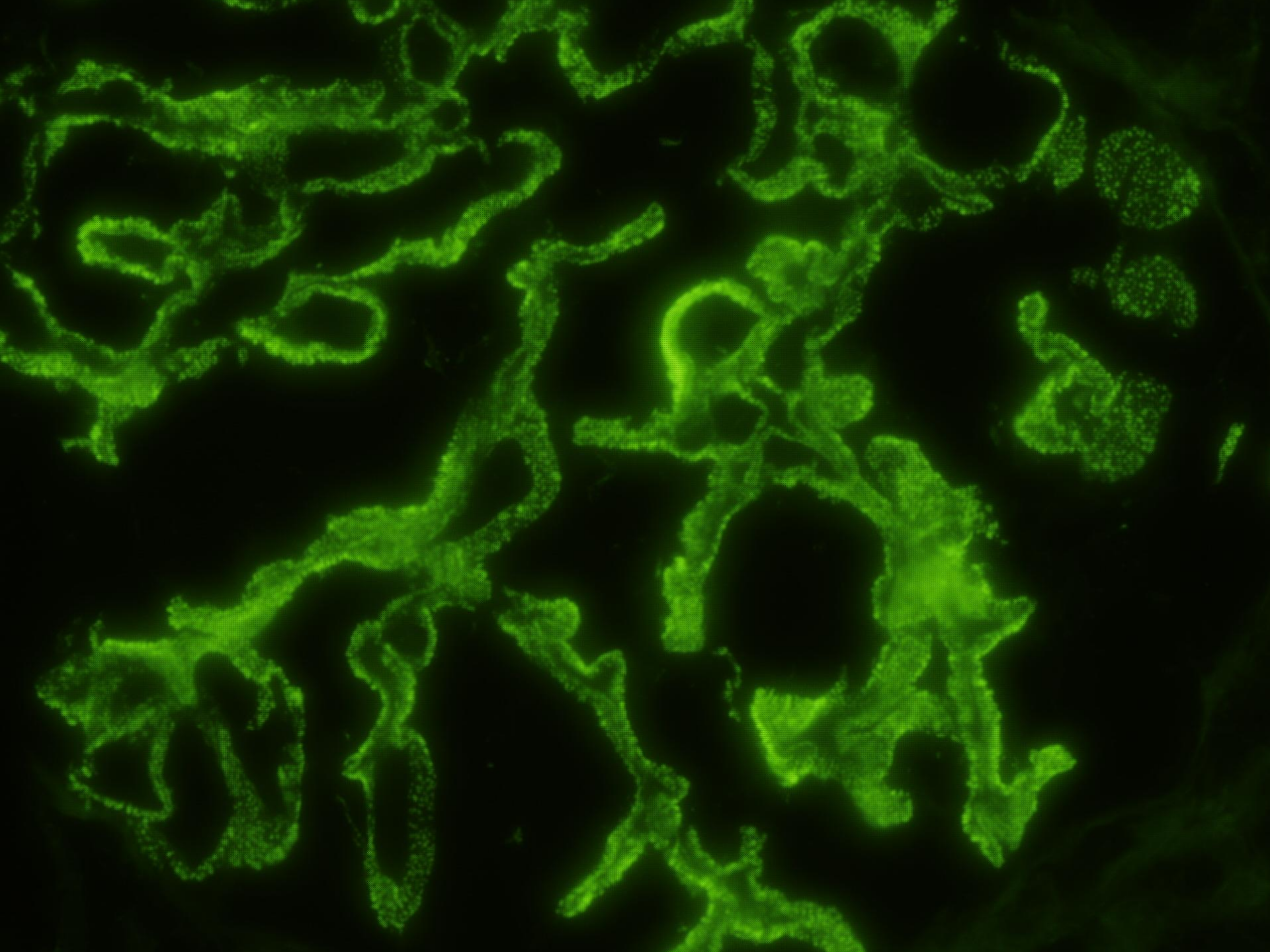
1

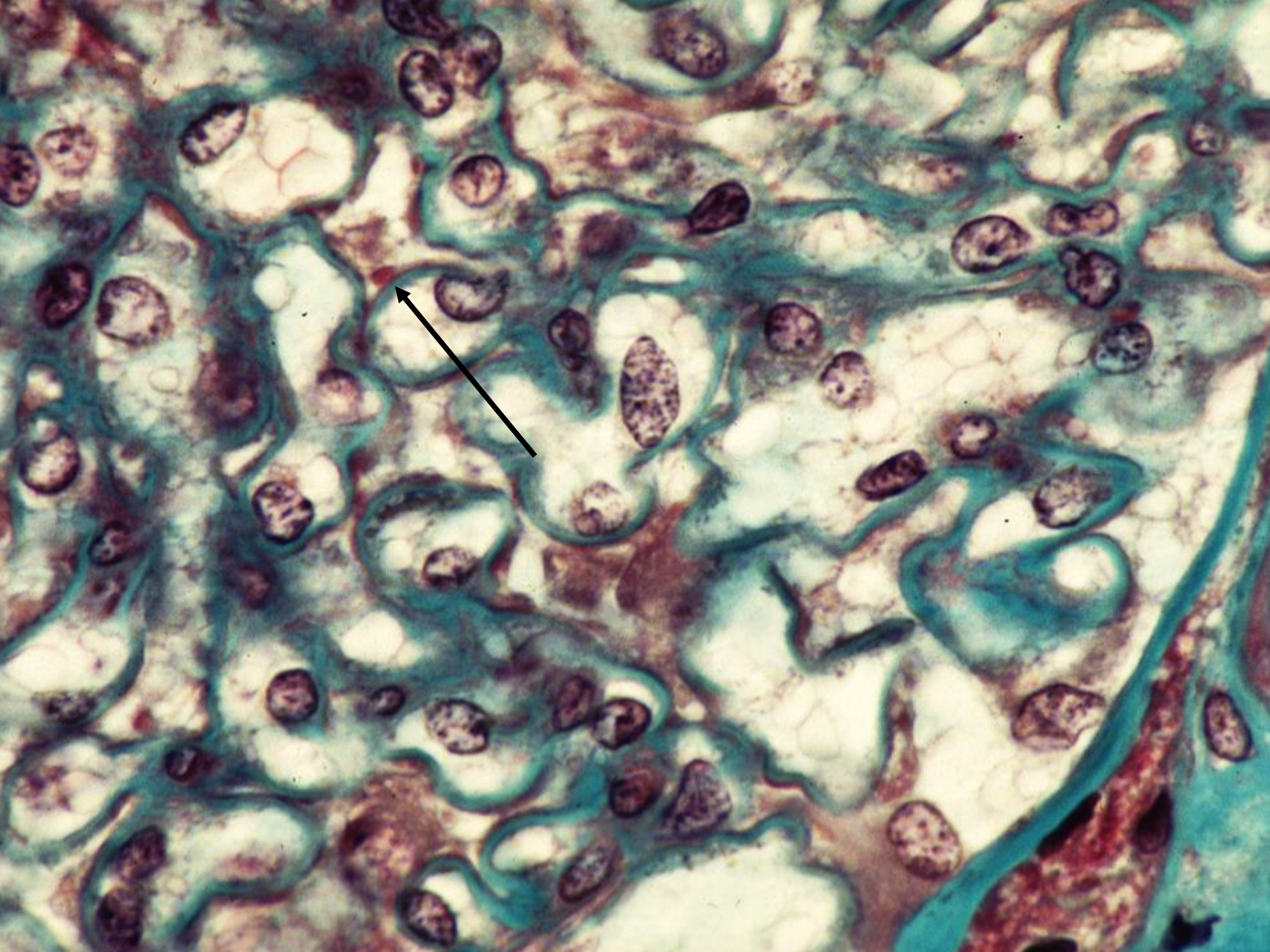
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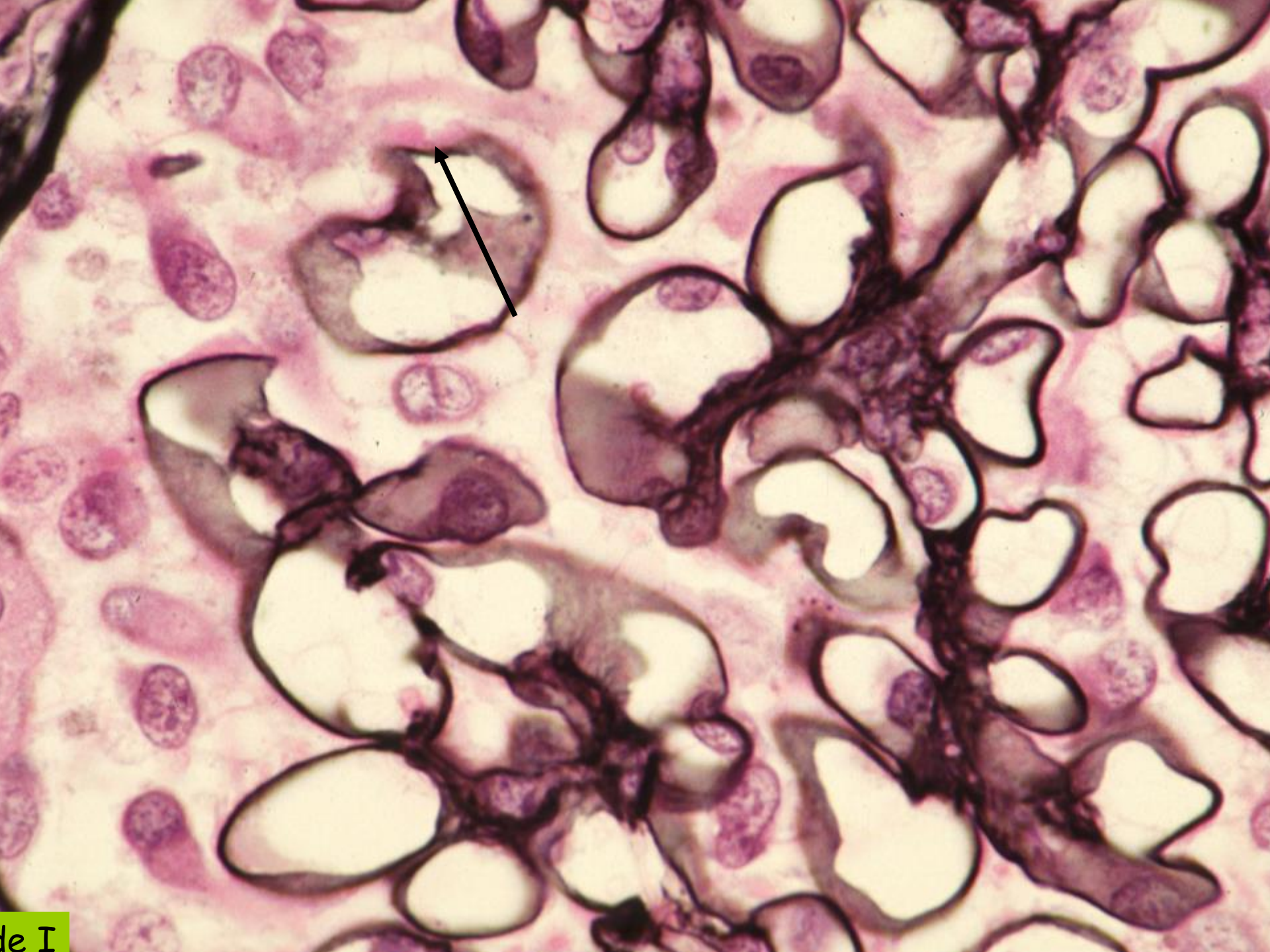
GEM



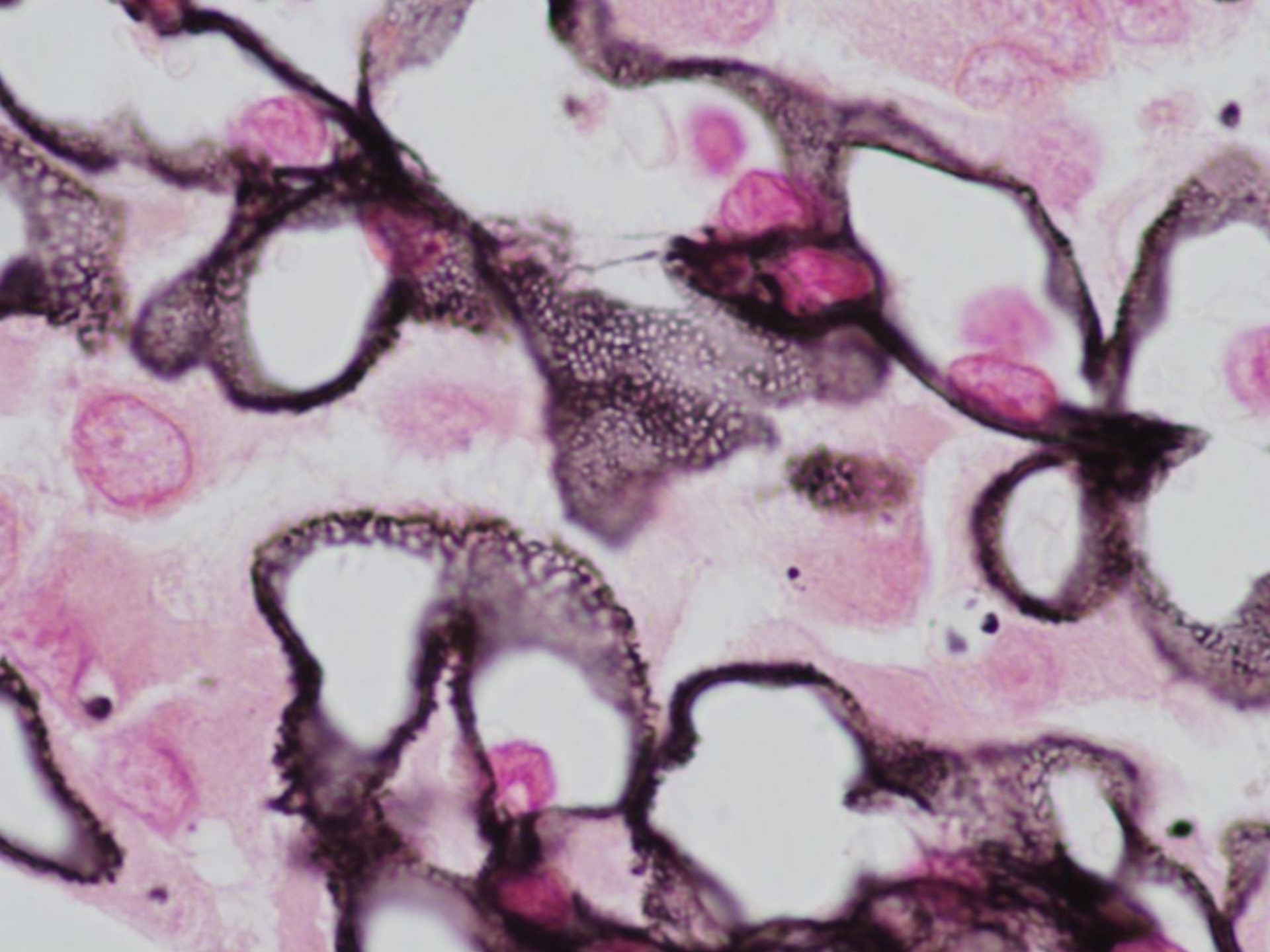
**Granuleux
diffus
pariétaux**

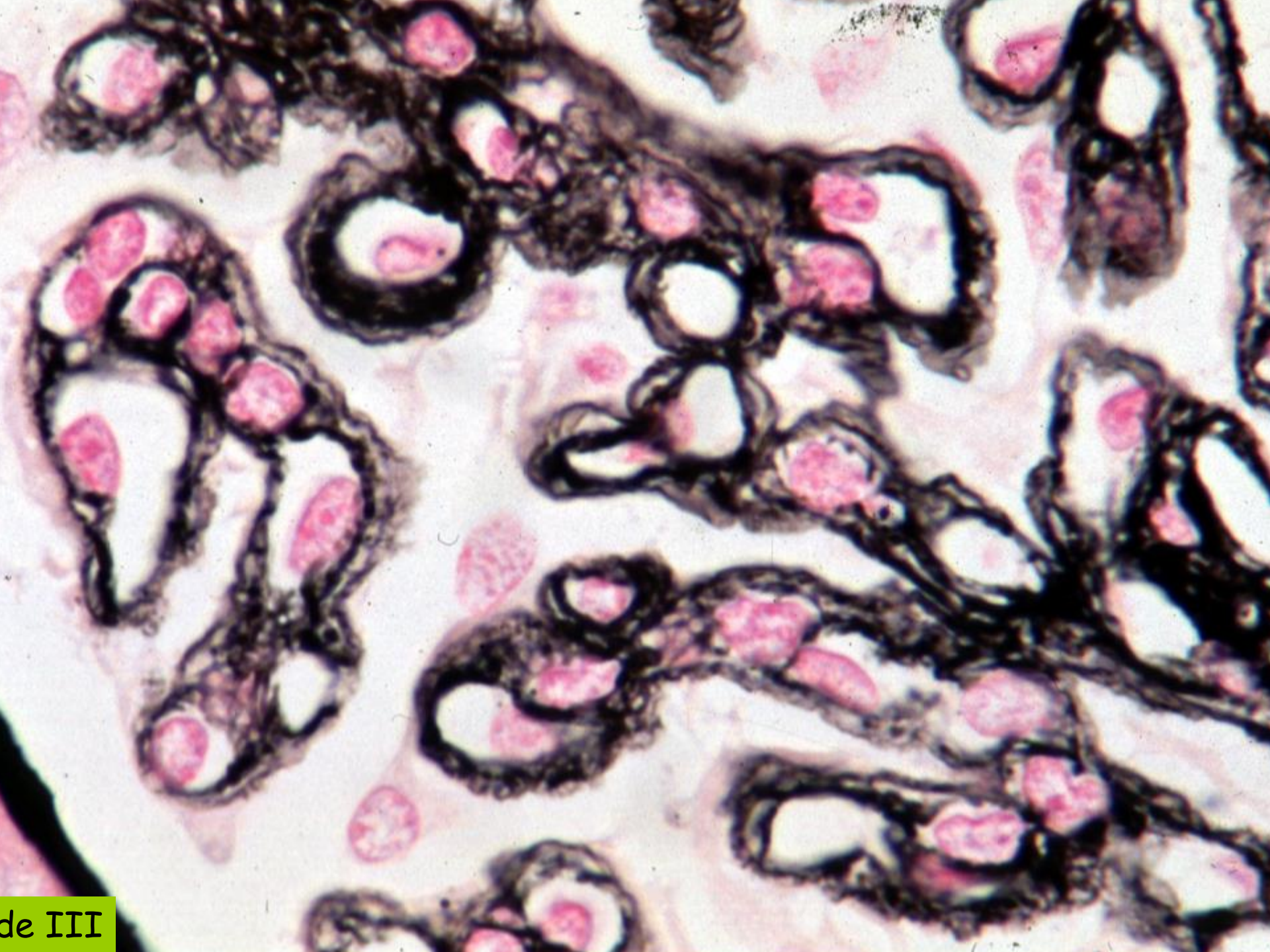






le I

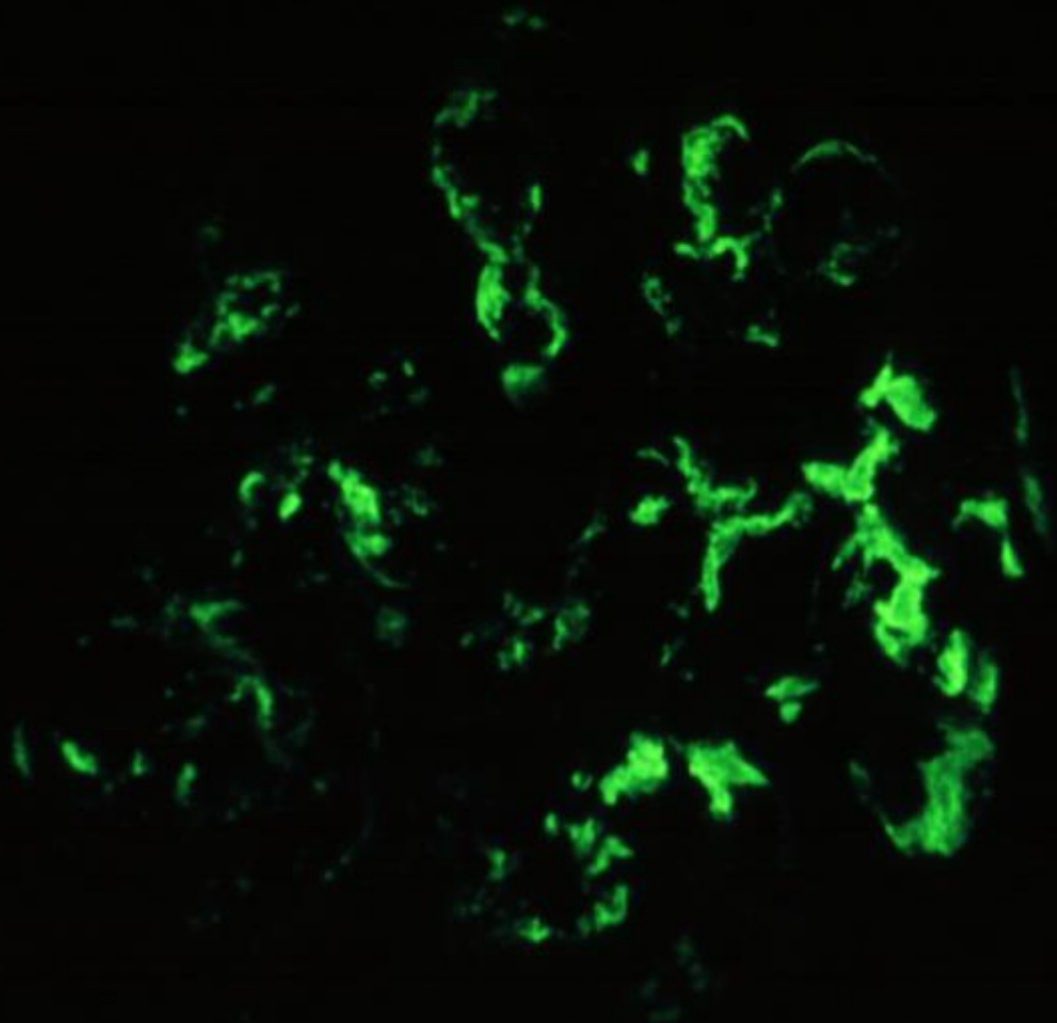




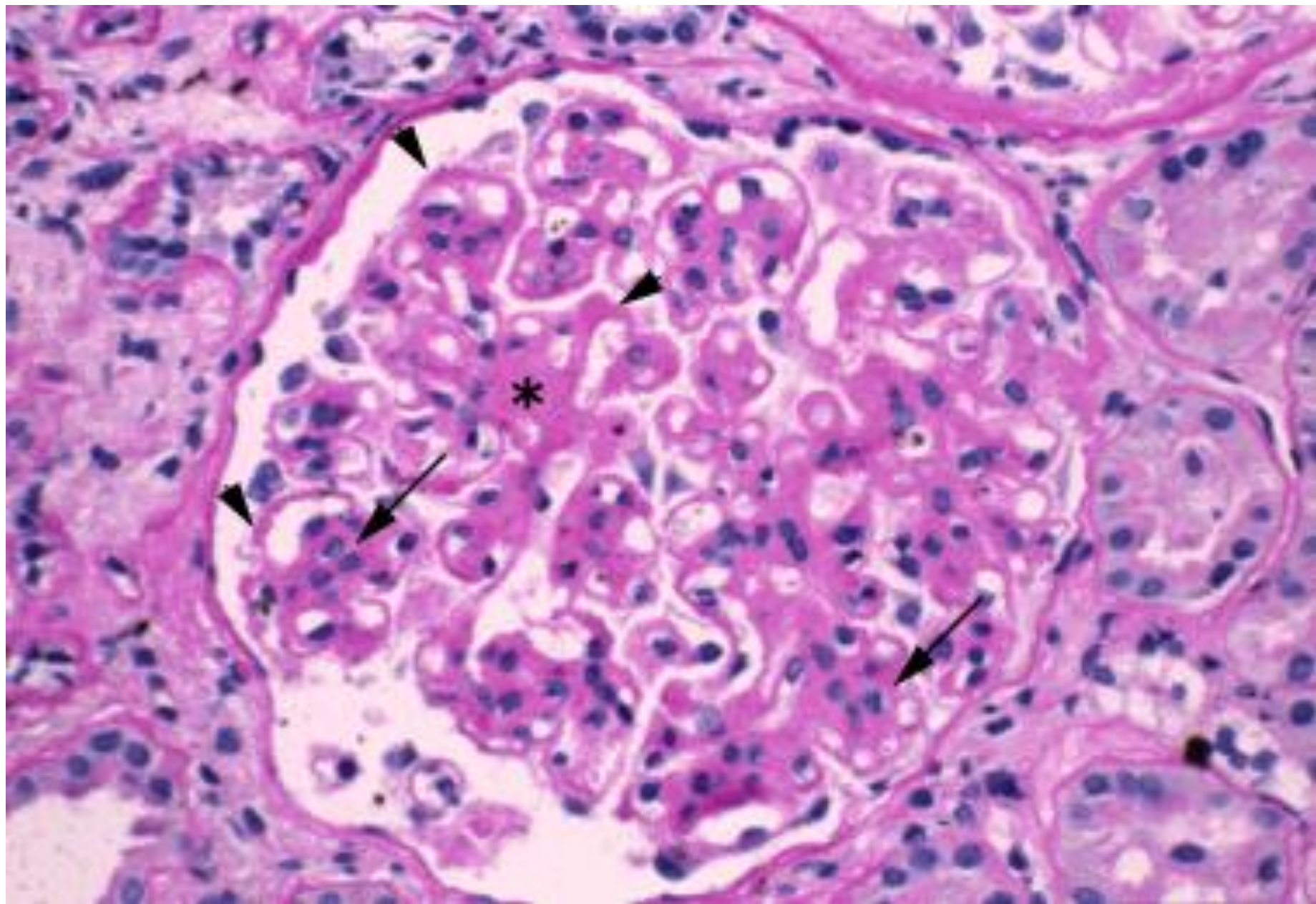
de III

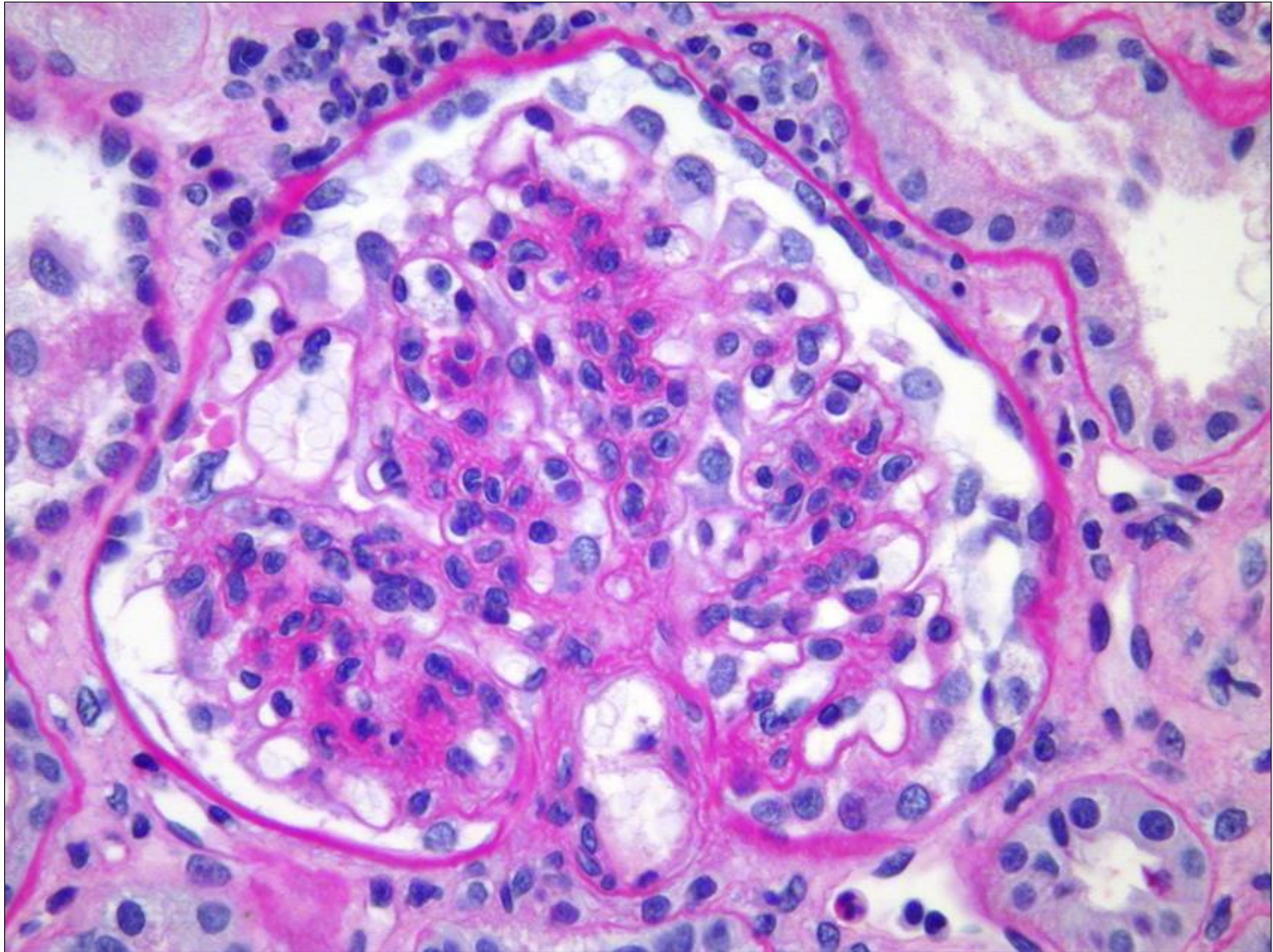
IgAN

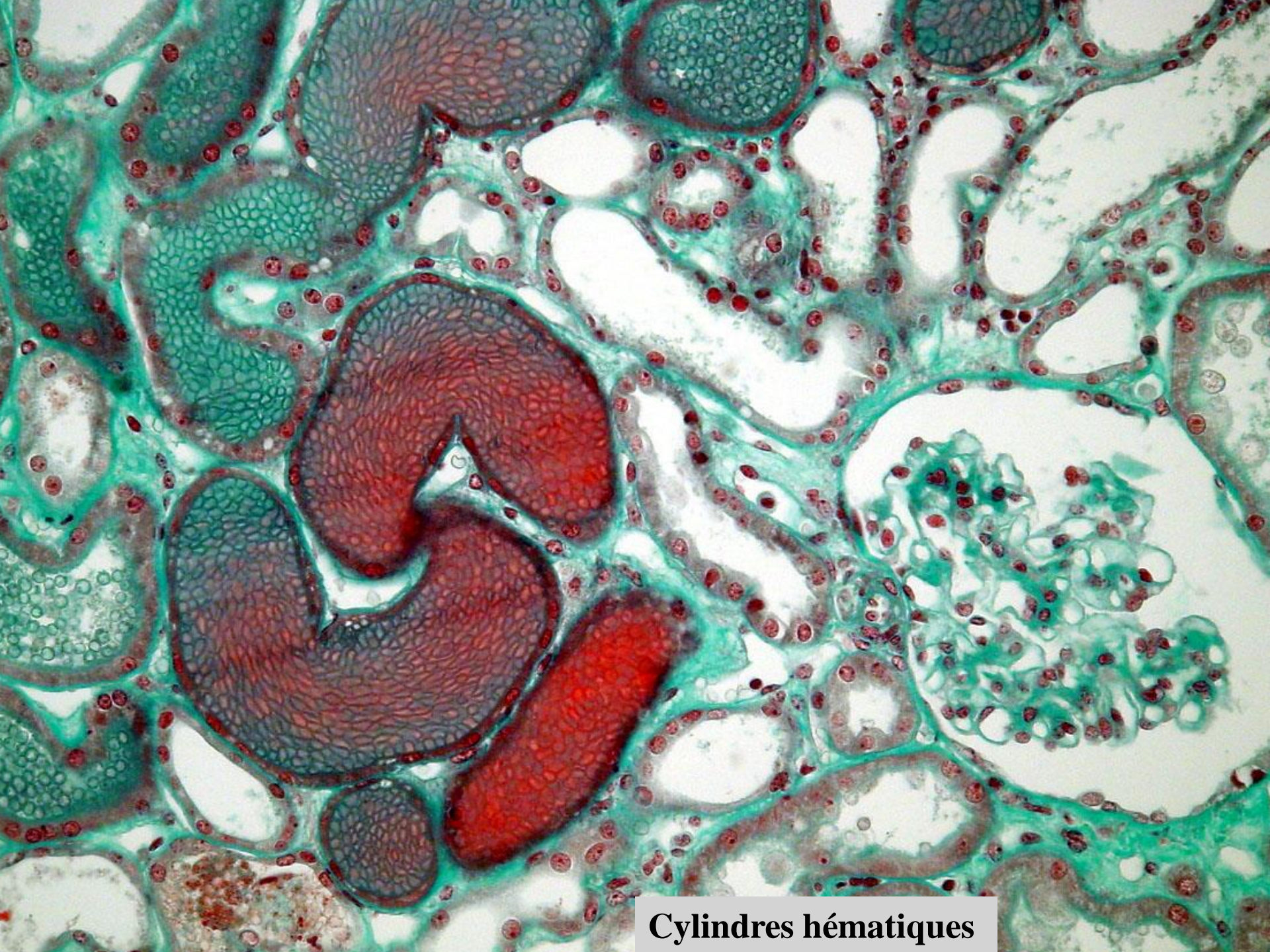
IgA: k < l



dénominateur commun des néphropathies à IgA







Cylindres hématiques

Box 2 | ISKDC histologic classification of HSP nephritis³⁰

- I: Minimal histologic alterations
- II: Pure mesangial proliferation
- III: Focal (IIIa) or diffuse (IIIb) mesangial proliferation with <50% crescentic glomeruli
- IV: Focal (IVa) or diffuse (IVb) mesangial proliferation with 50–75% crescentic glomeruli
- V: Focal (Va) or diffuse (Vb) mesangial proliferation with >75% crescentic glomeruli
- VI: Membranoproliferative-like glomerulonephritis

Abbreviations: HSP, Henoch–Schönlein purpura; ISKDC, International Study of Kidney Disease in Children.

Tableau 11-VI Classification histologique [15].

Classe 1 : glomérules normaux ou discrète hypertrophie mésangiale.

Classe 2 : moins de 50 p. 100 des glomérules atteints avec prolifération mésangiale et/ou prolifération extracapillaire segmentaire.

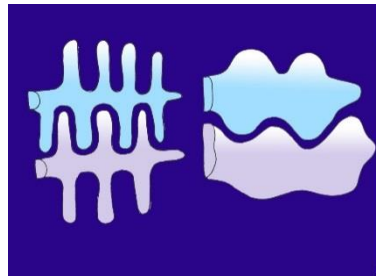
Classe 3a : prolifération endocapillaire modérée globale.

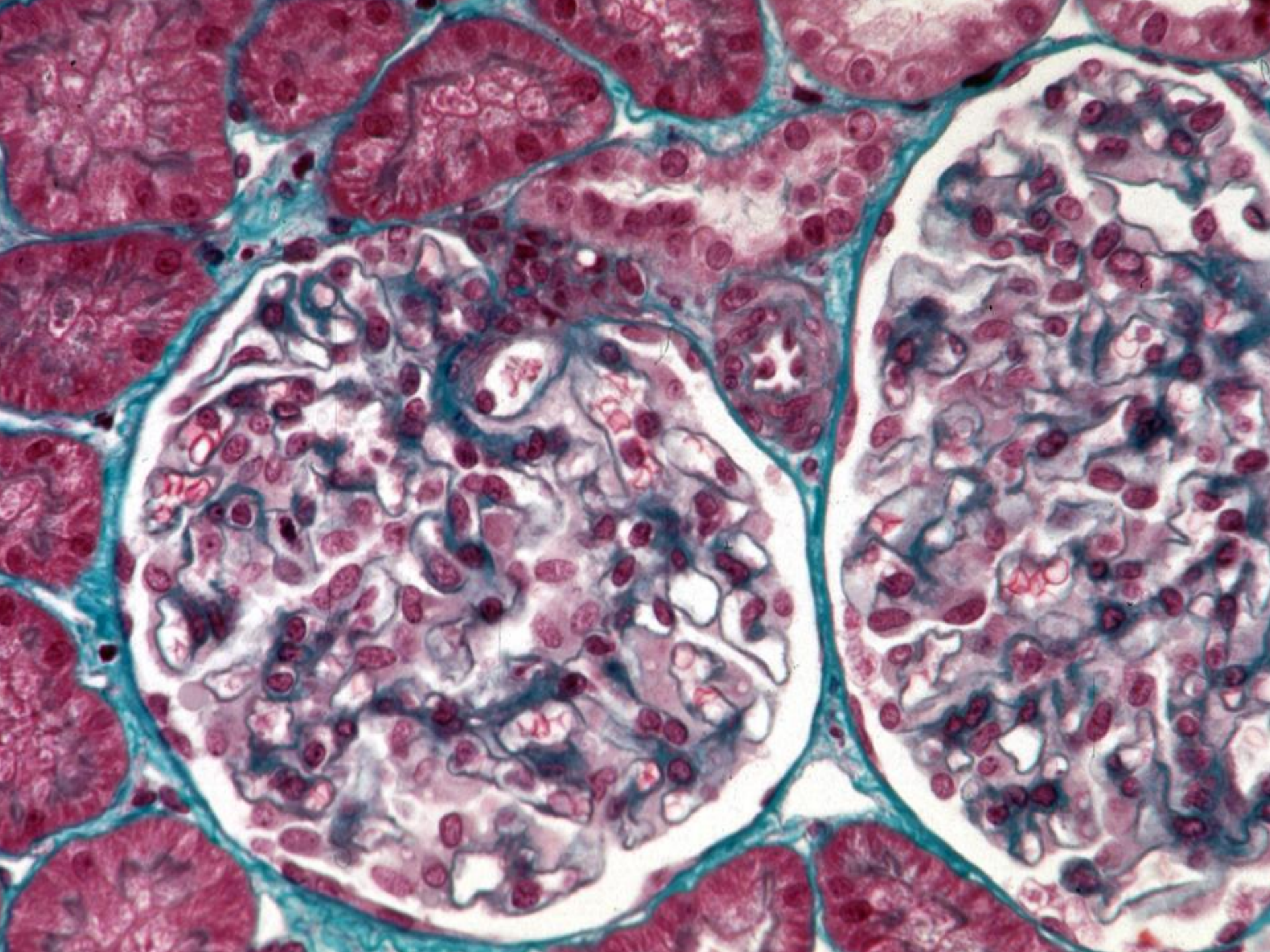
Classe 3b : prolifération endocapillaire sévère globale associée à des foyers de prolifération extracapillaire touchant moins de 50 p. 100 des glomérules.

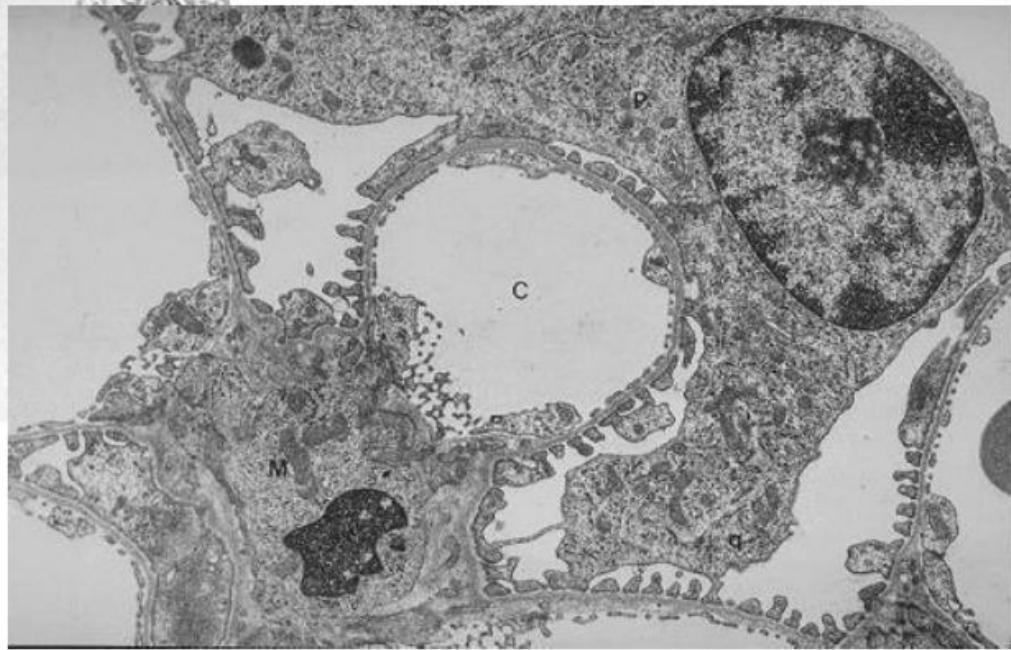
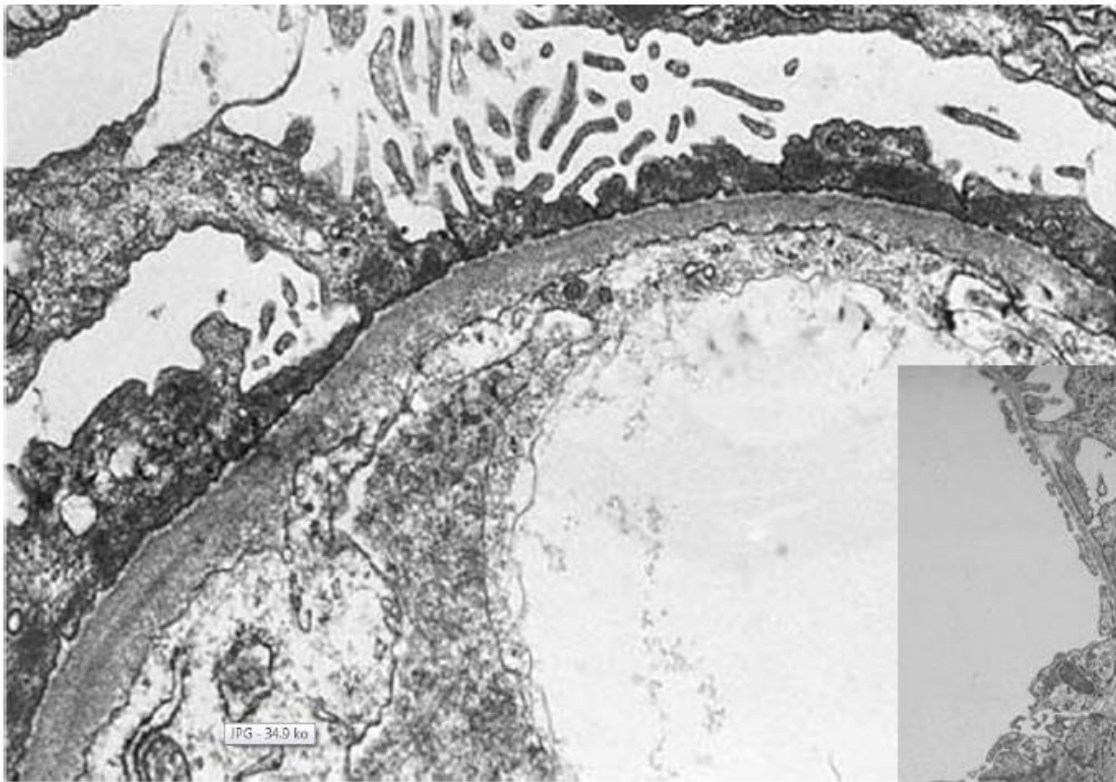
Classe 4 : lésions identiques à celles de la classe 3, mais avec des croissants dans plus de 50 p. 100 des glomérules.

Classe 5 : glomérules scléreux touchant plus de 50 p. 100 des glomérules

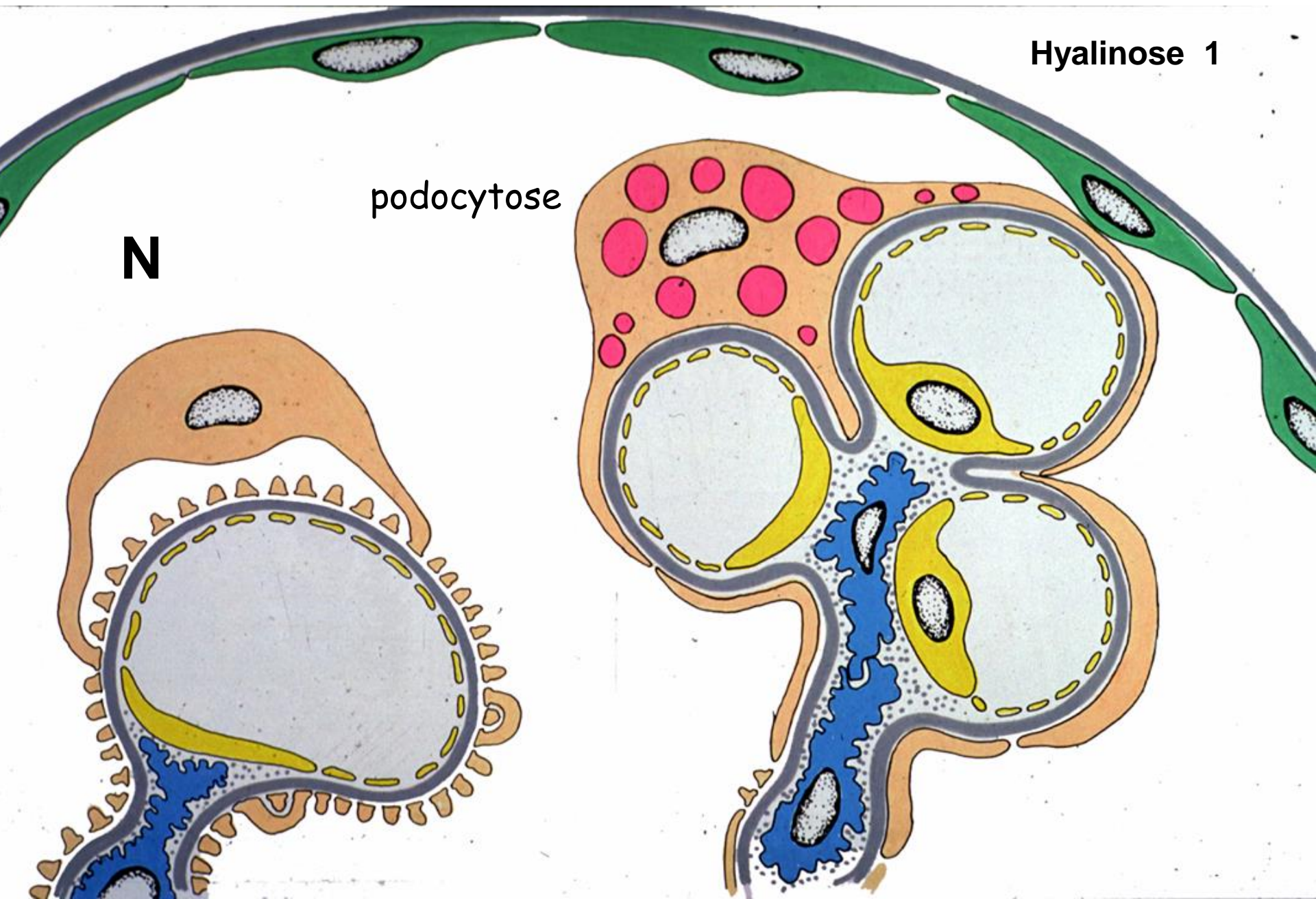
L.G.M







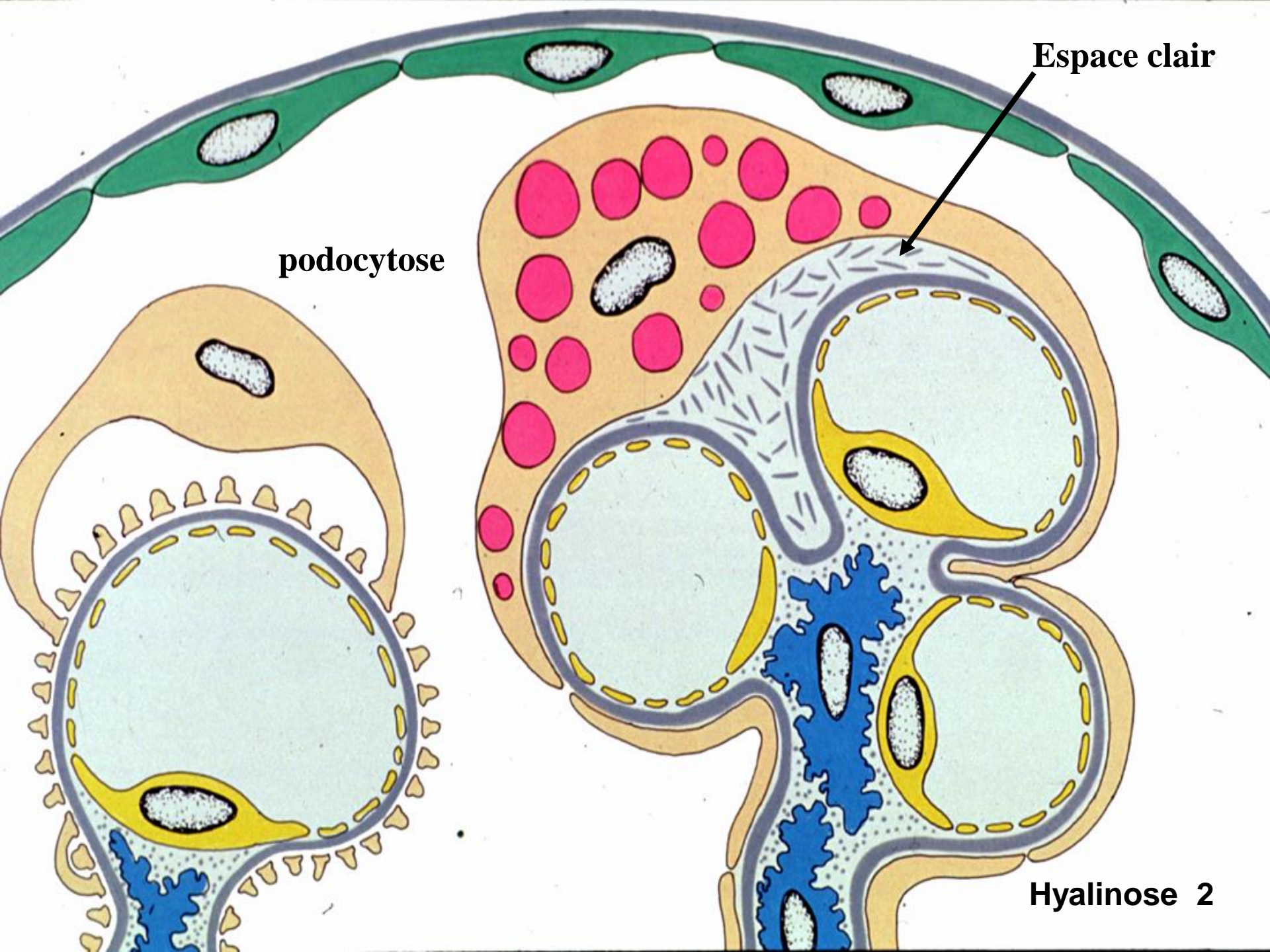
H.S.F



Hyalinose 1

podocyte

N

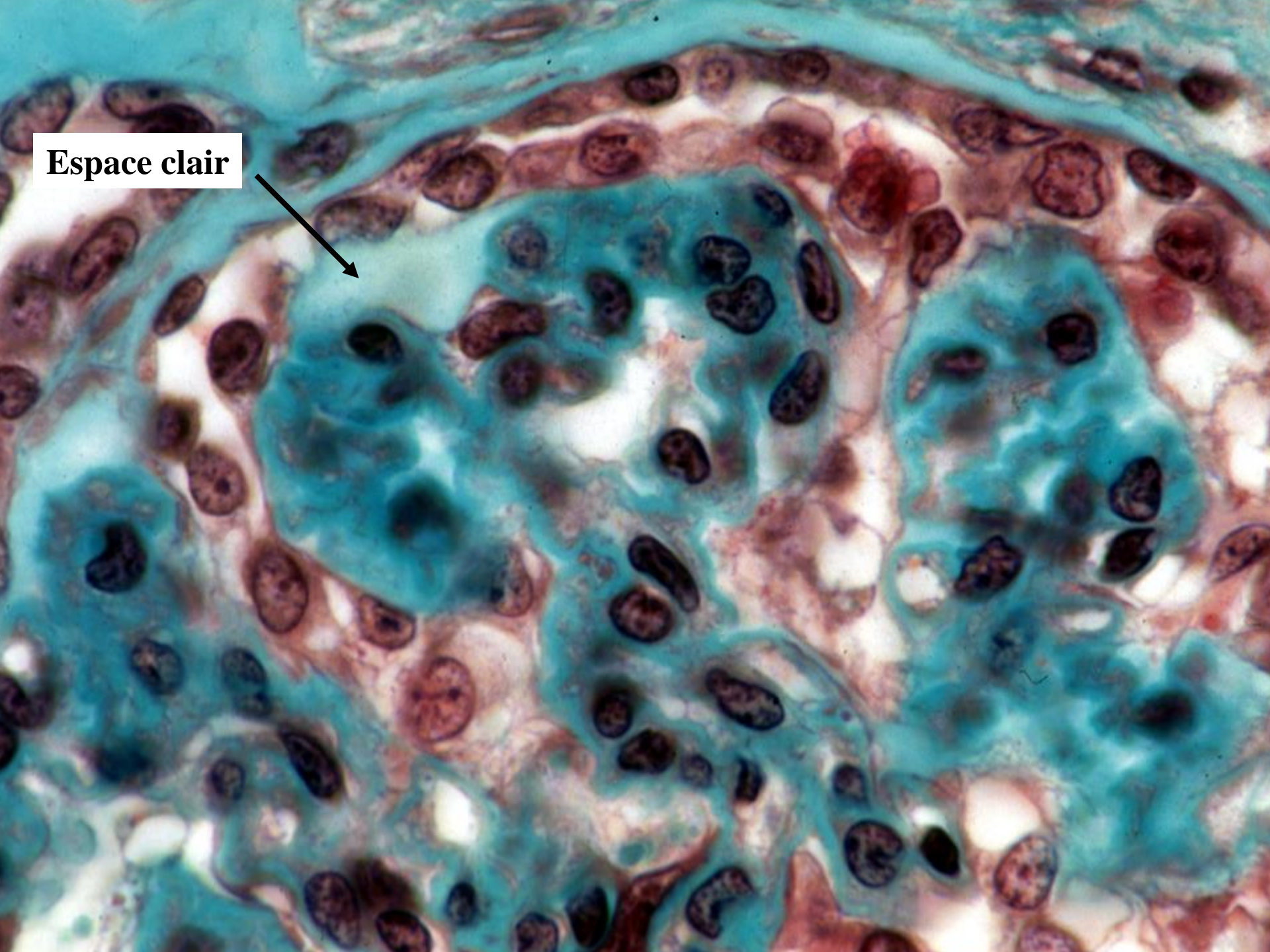
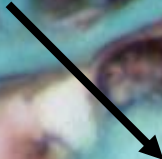


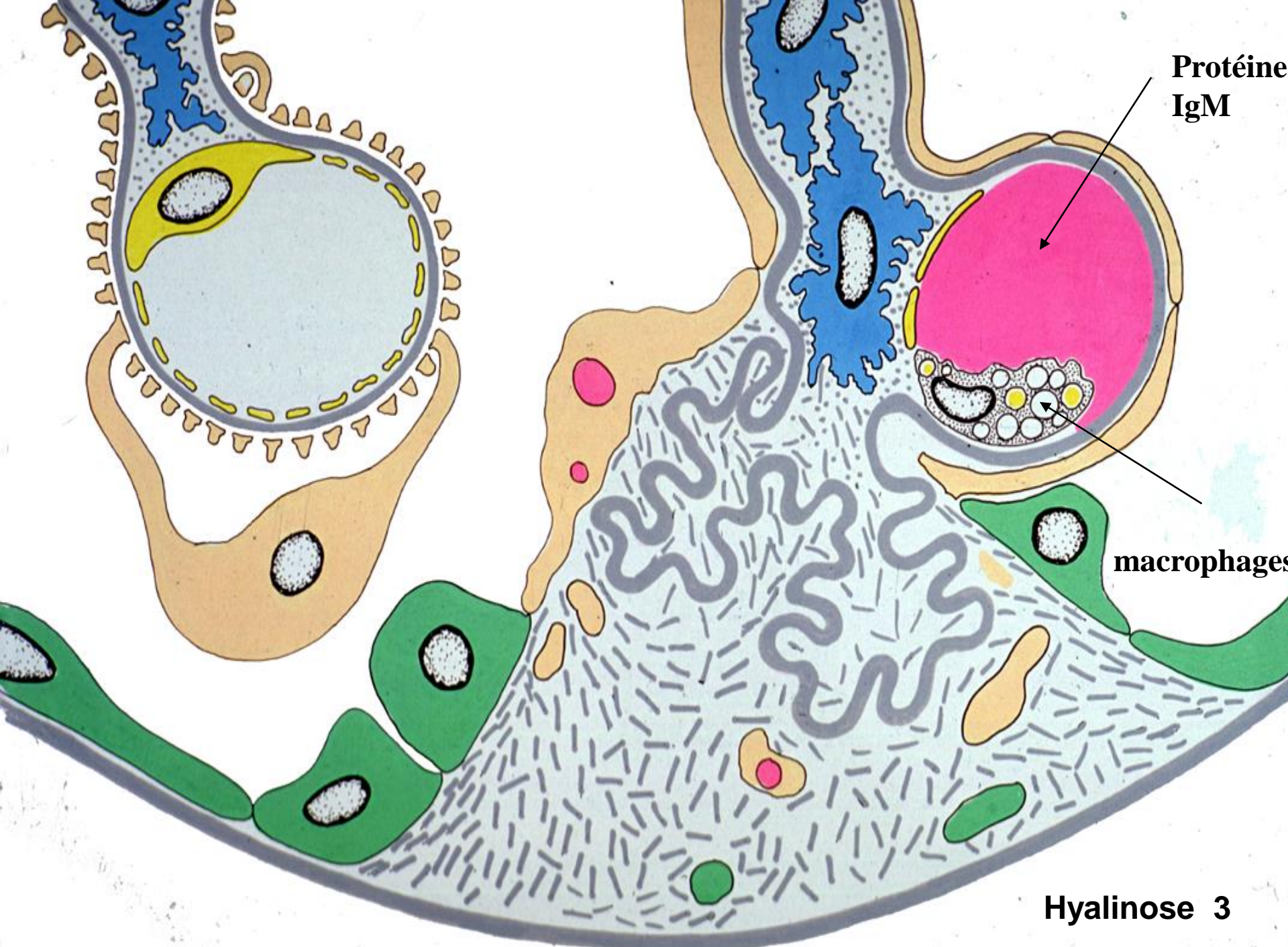
Espace clair

podocyte

Hyalinose 2

Espace clair

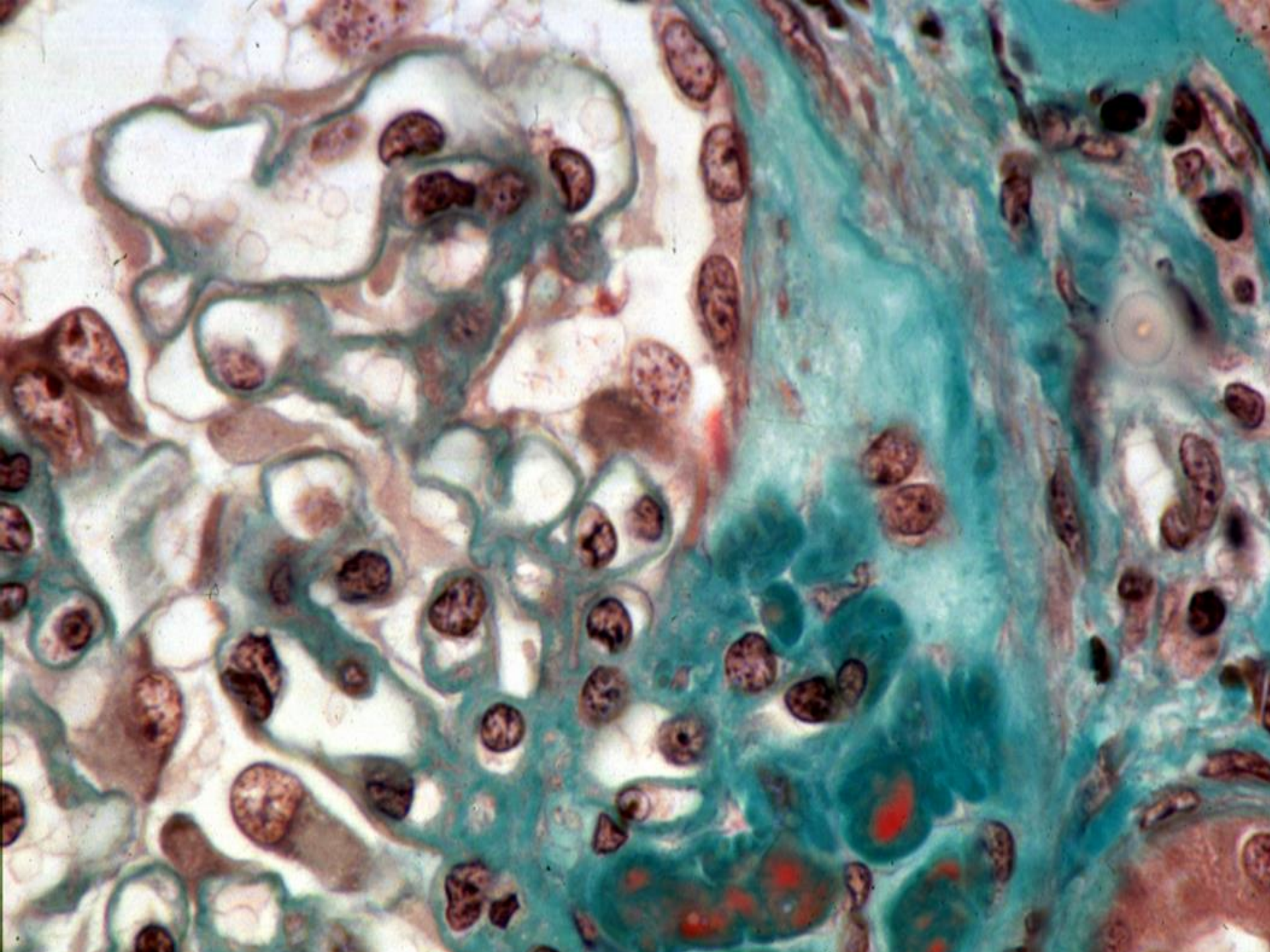


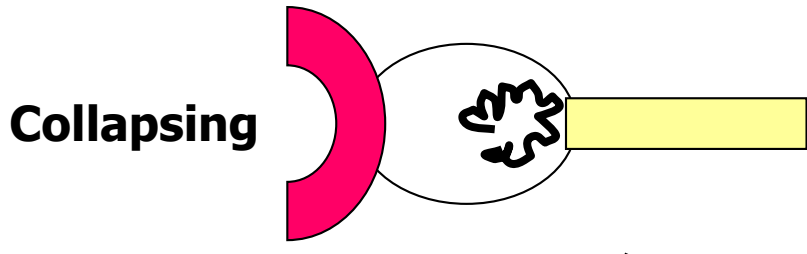
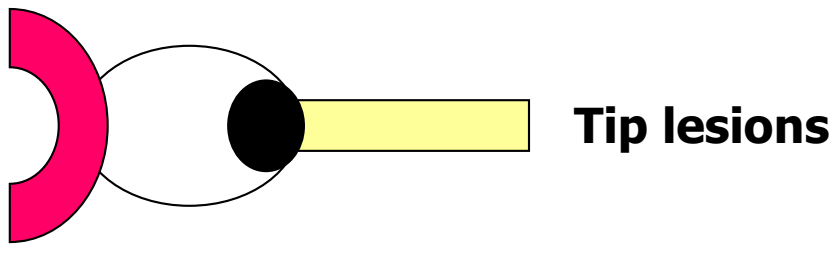
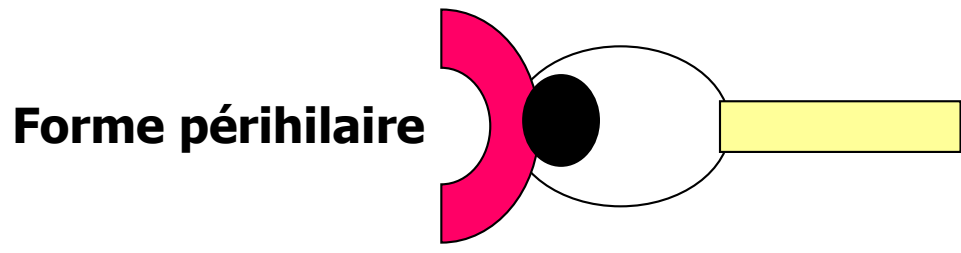


Protéine
IgM

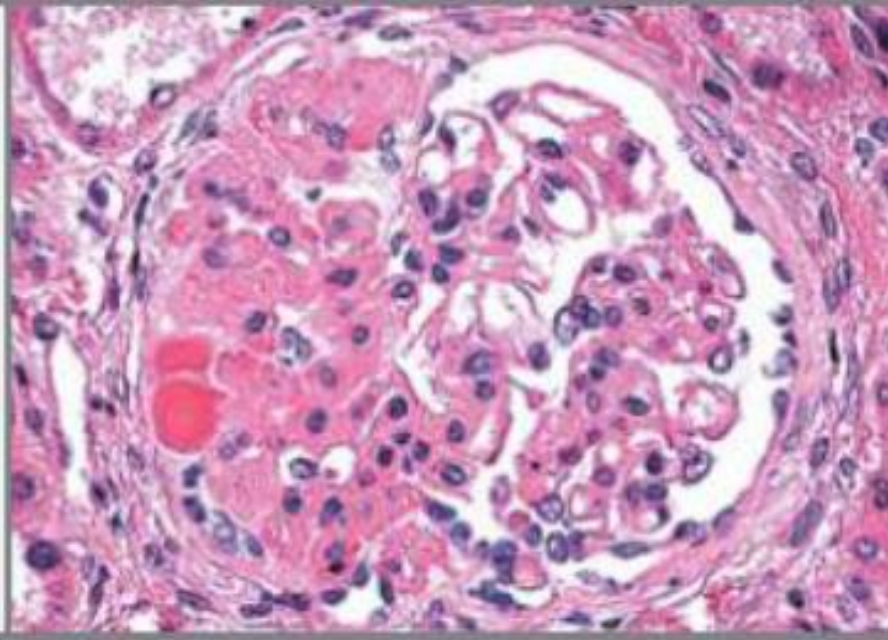
macrophages

Hyalinose 3



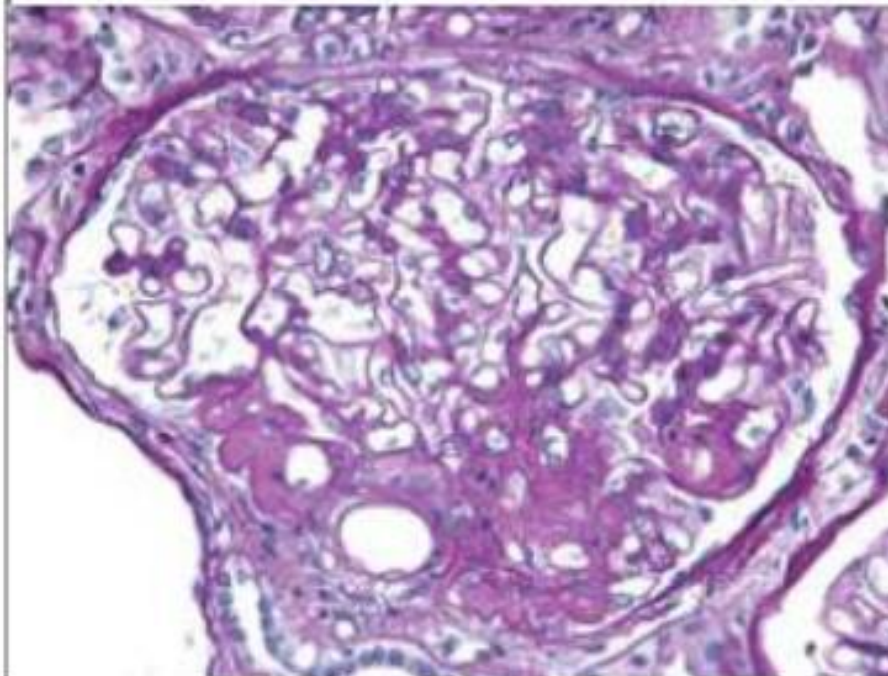


NOS

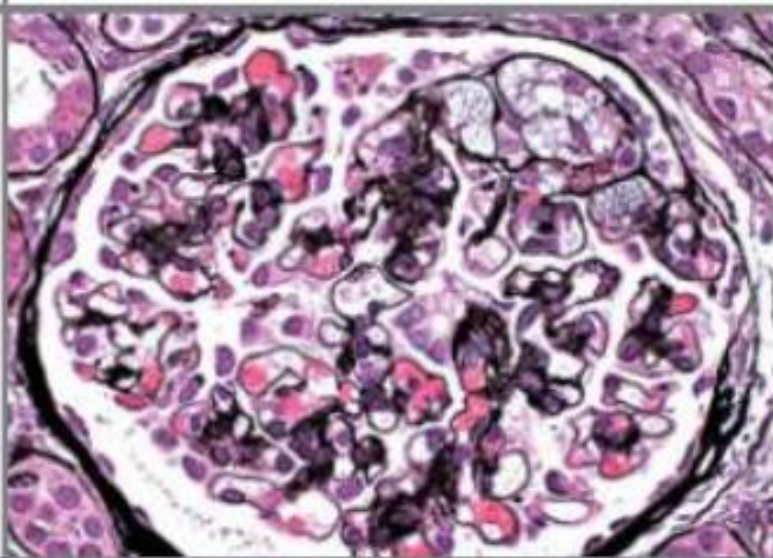


Perihilar

= au pôle
vasculaire

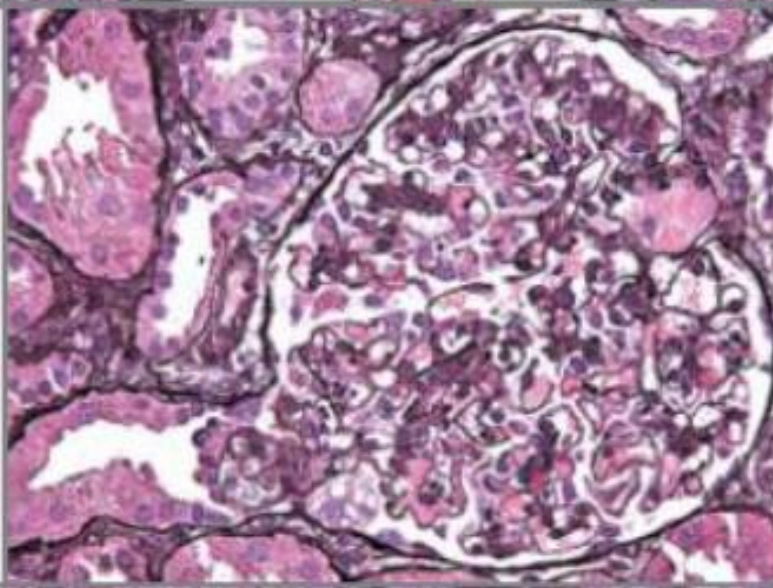


Cellular

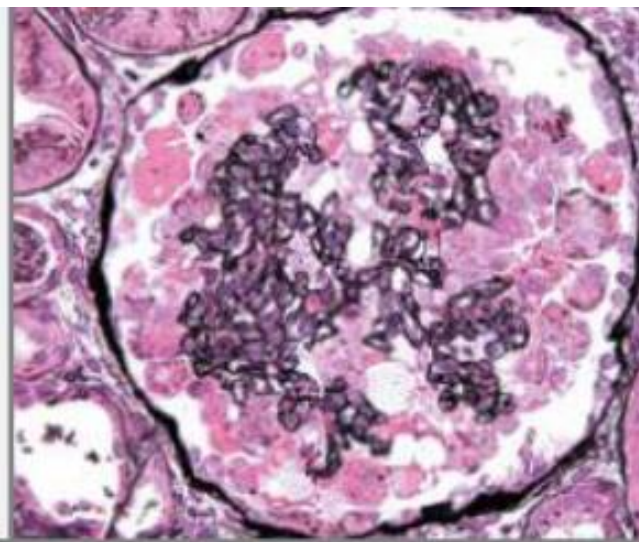


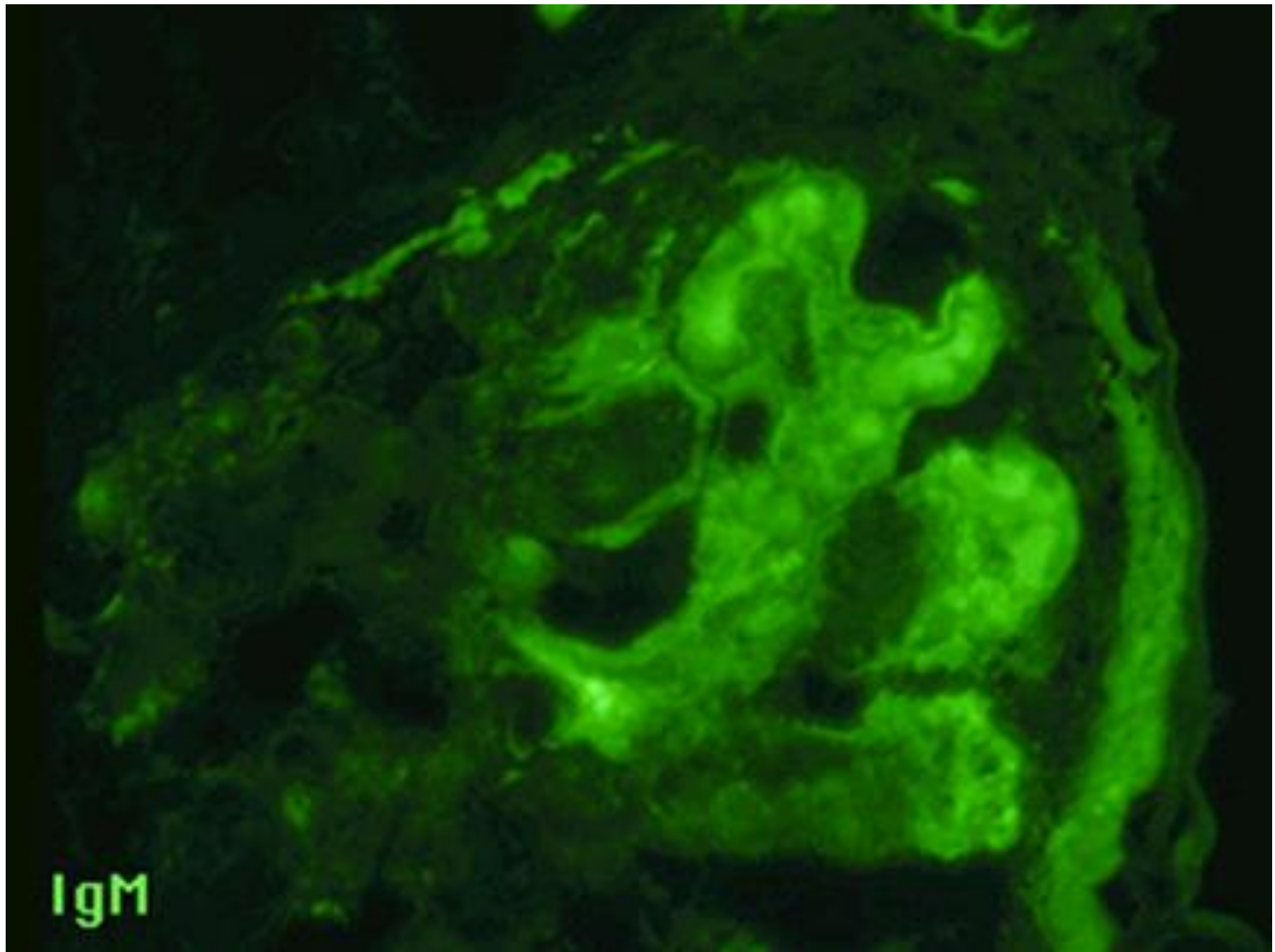
Tip

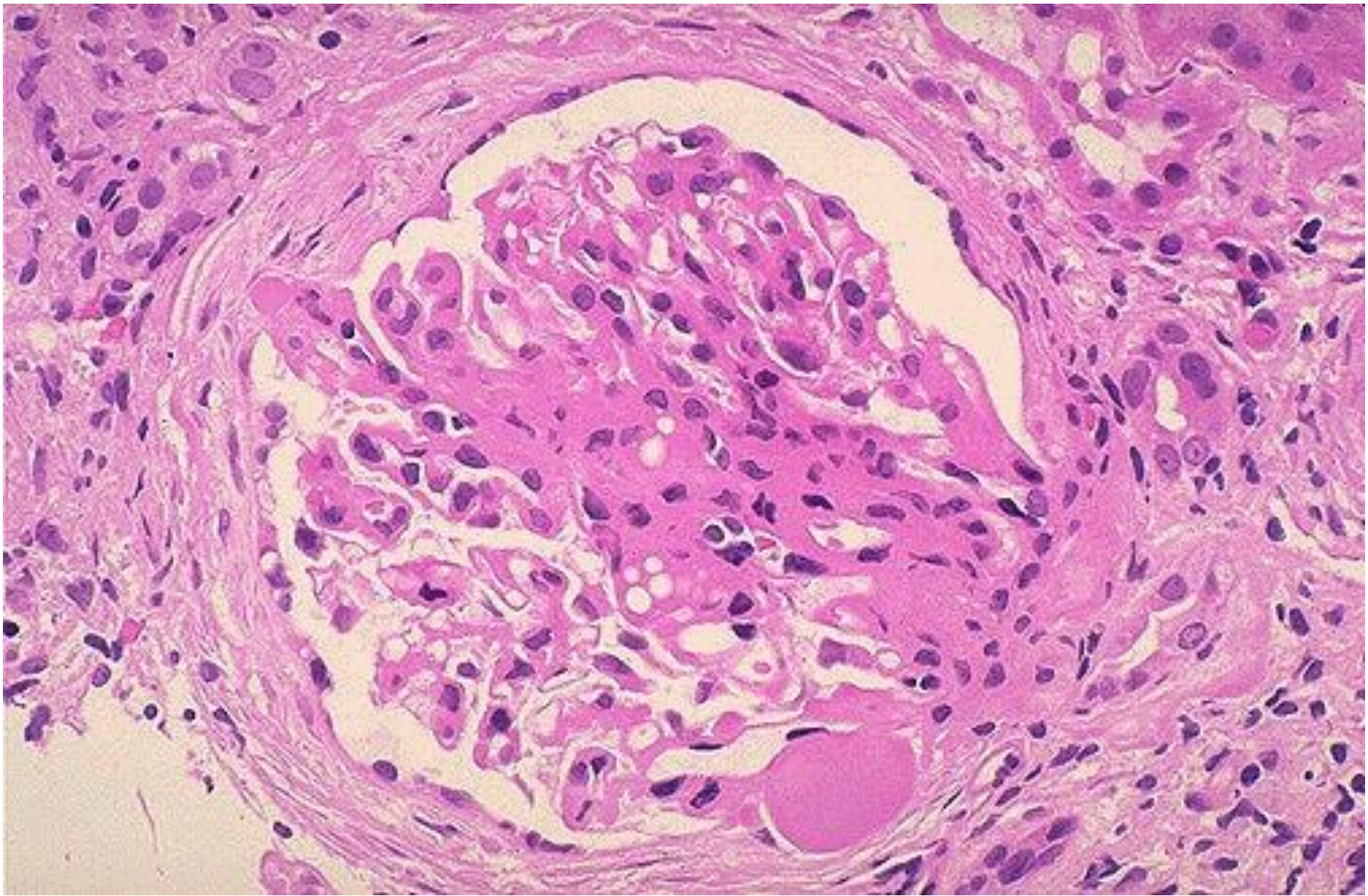
= au pôle urinaire

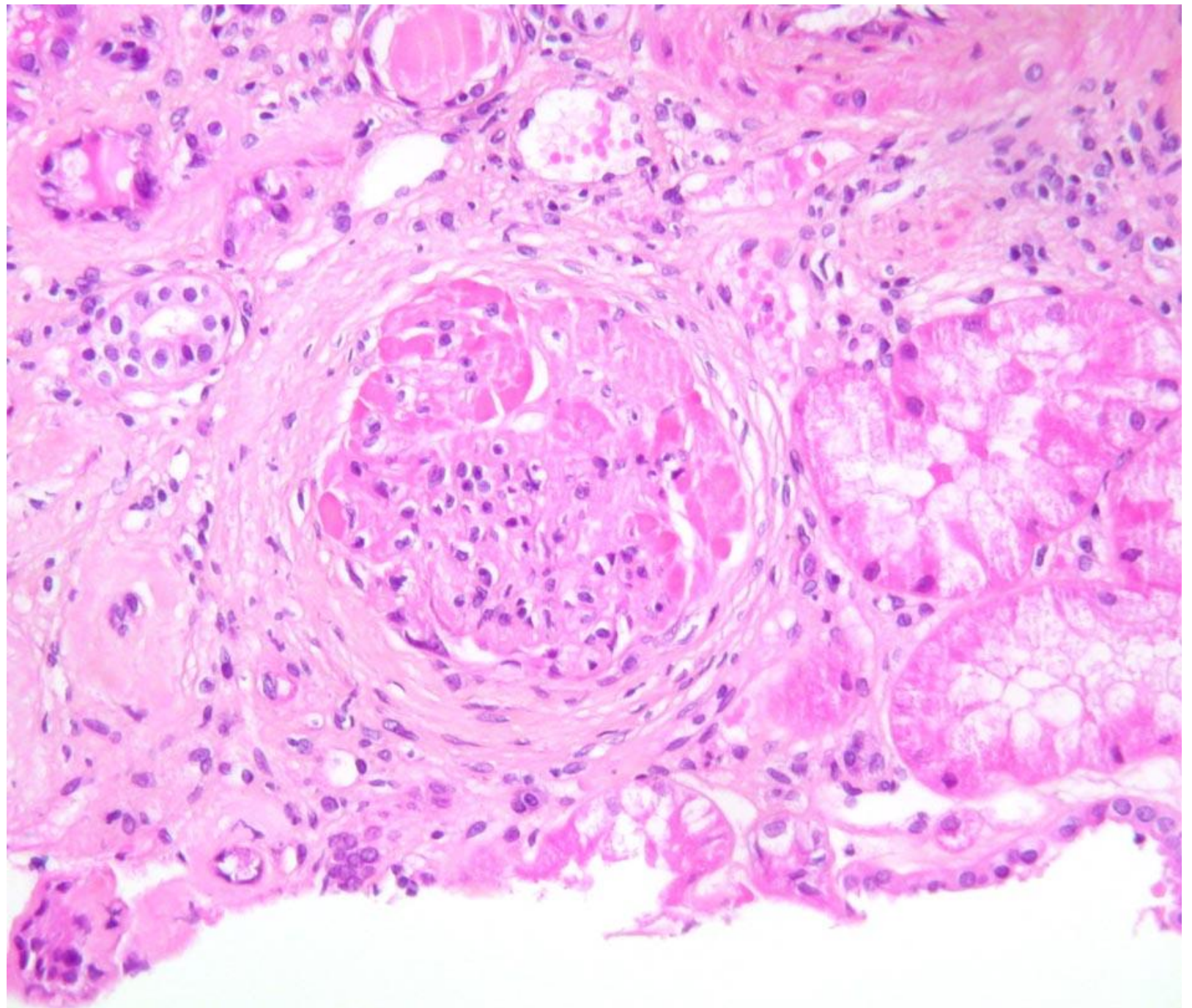


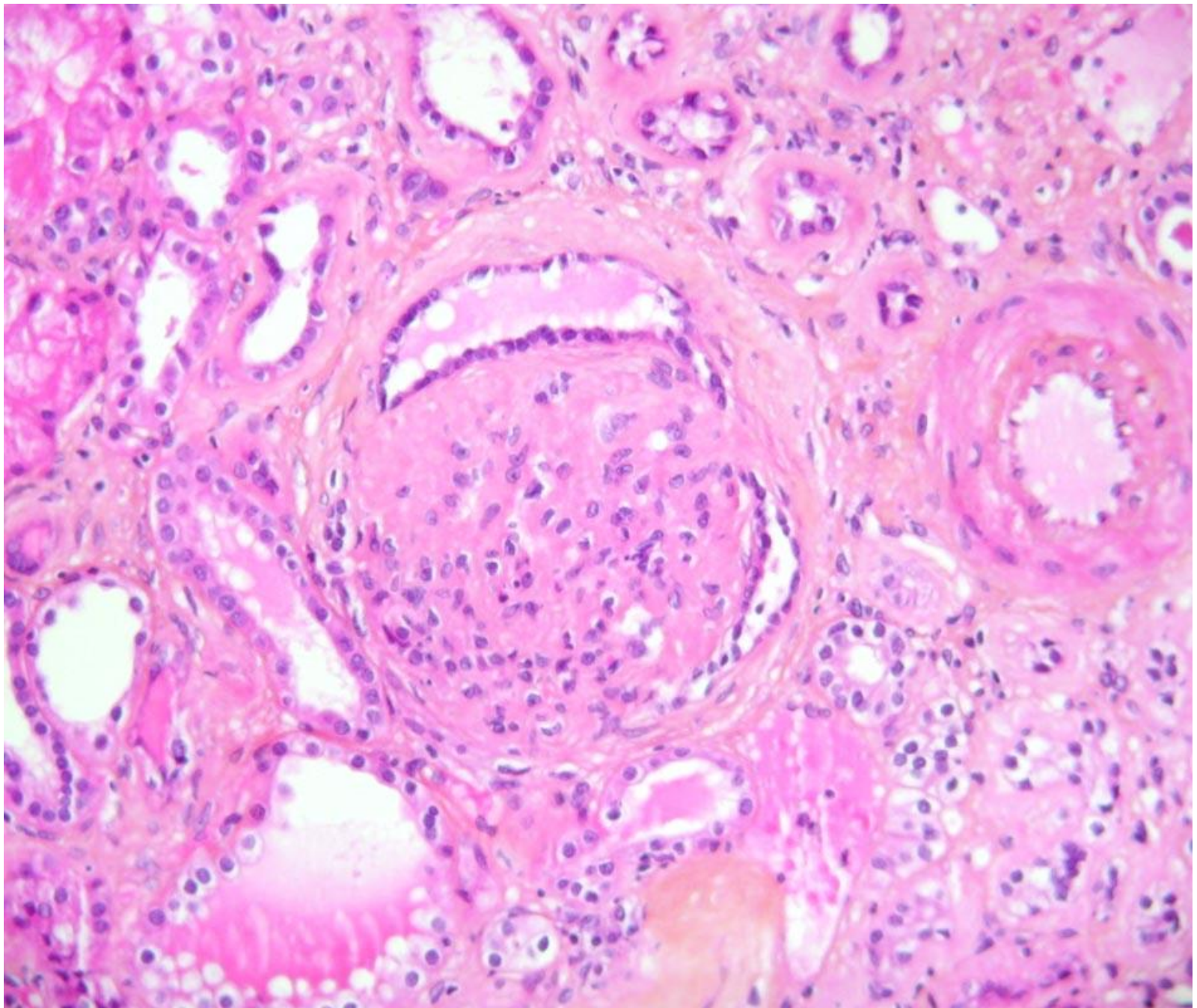
Collapse

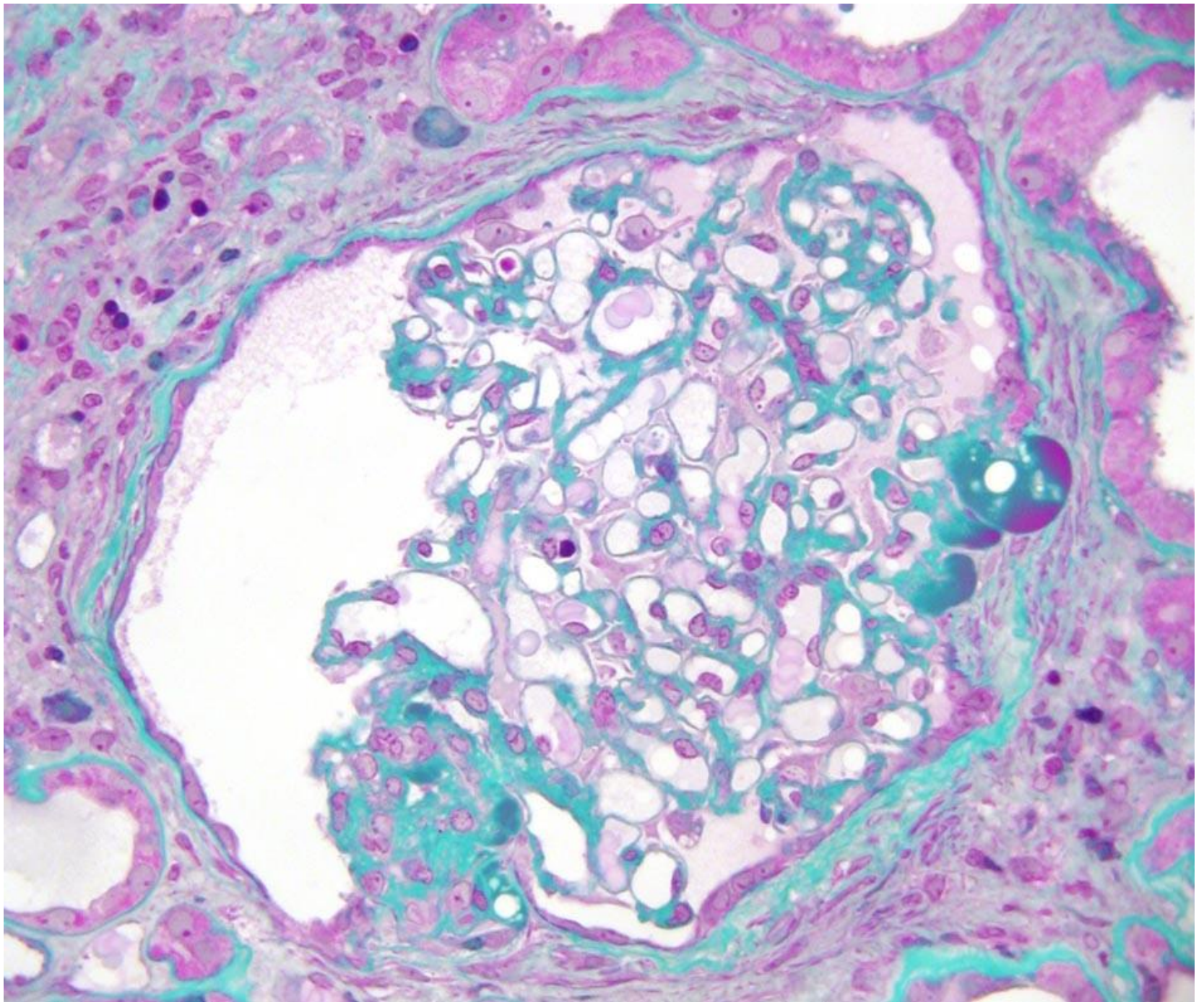


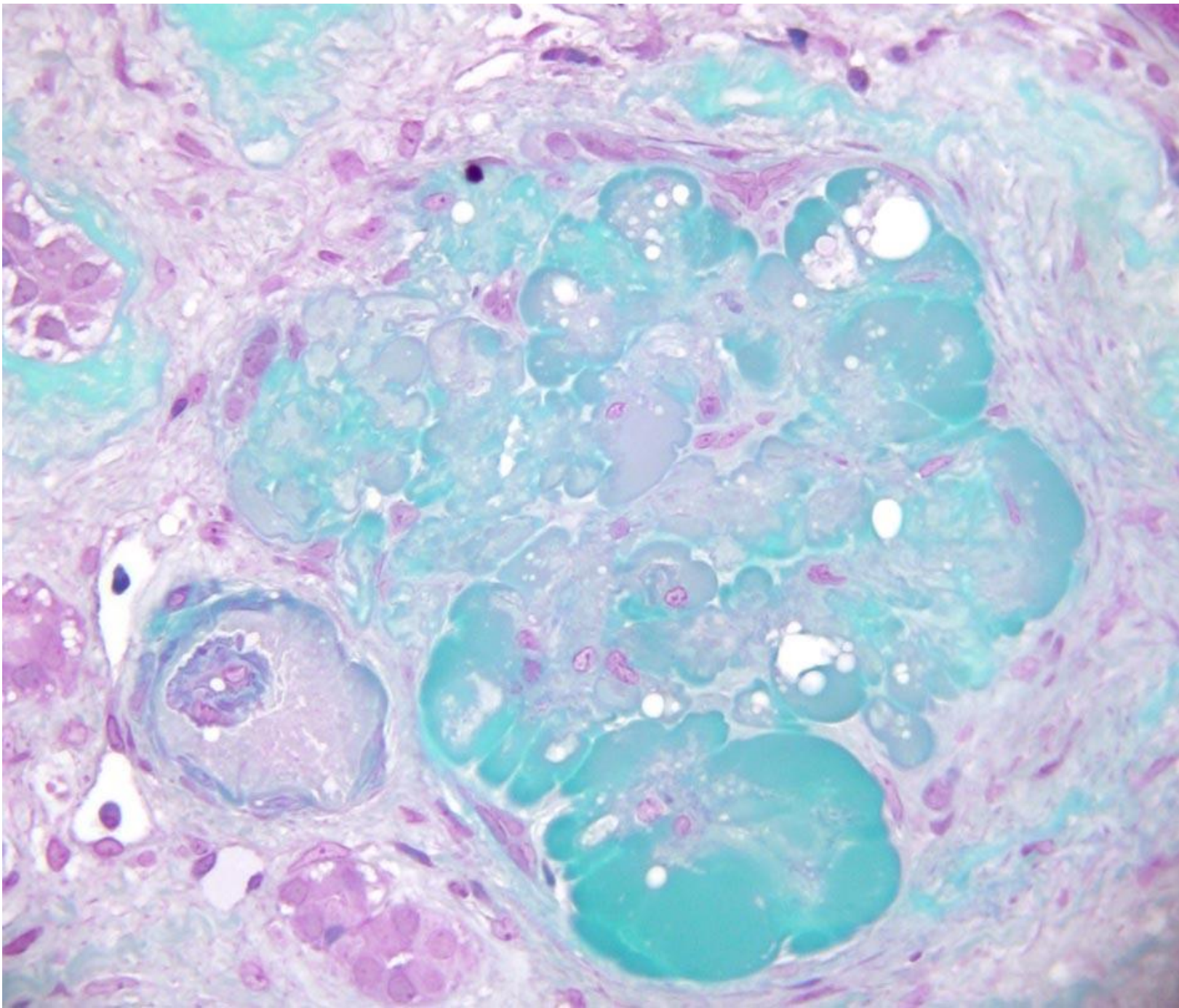


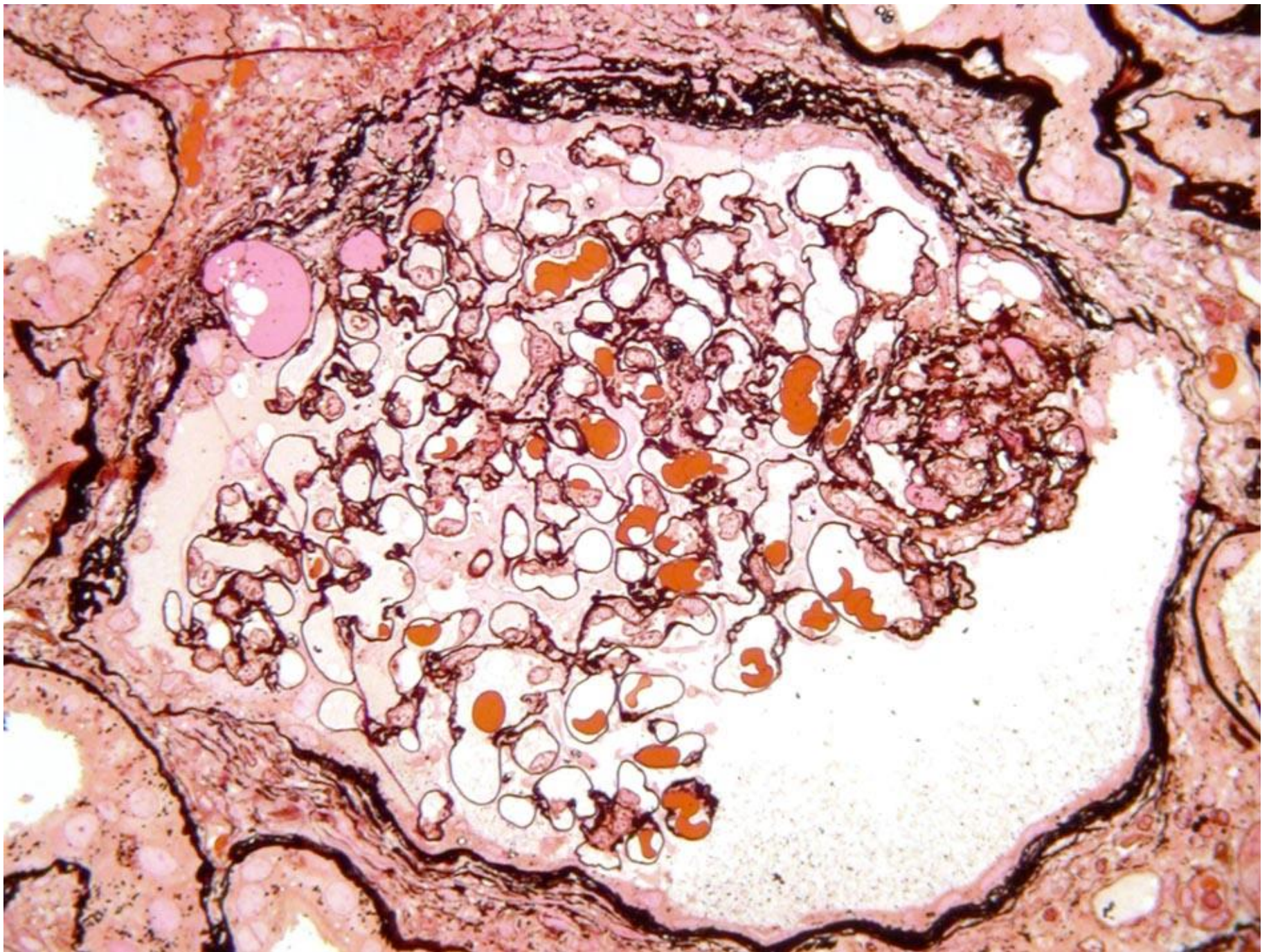


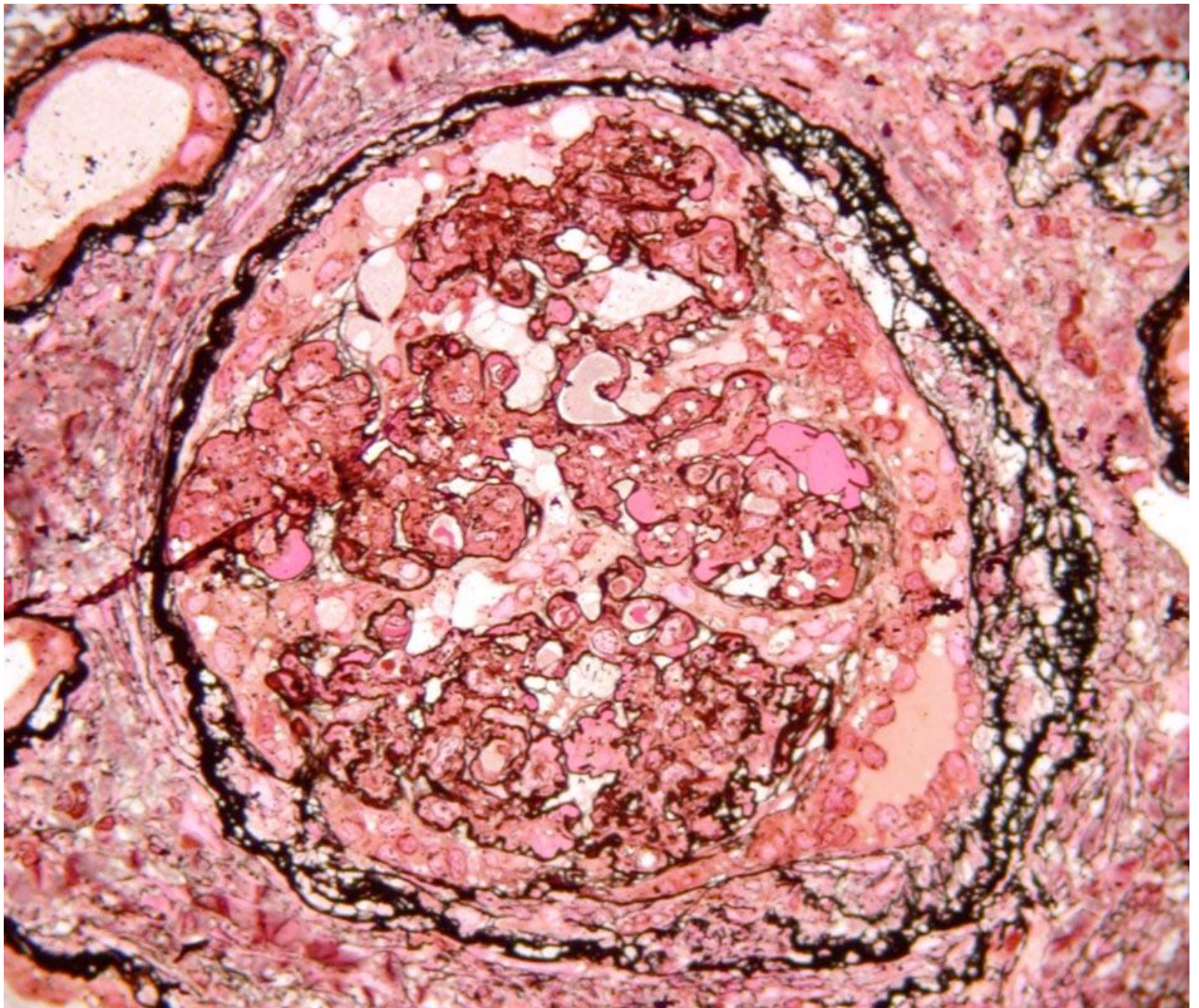


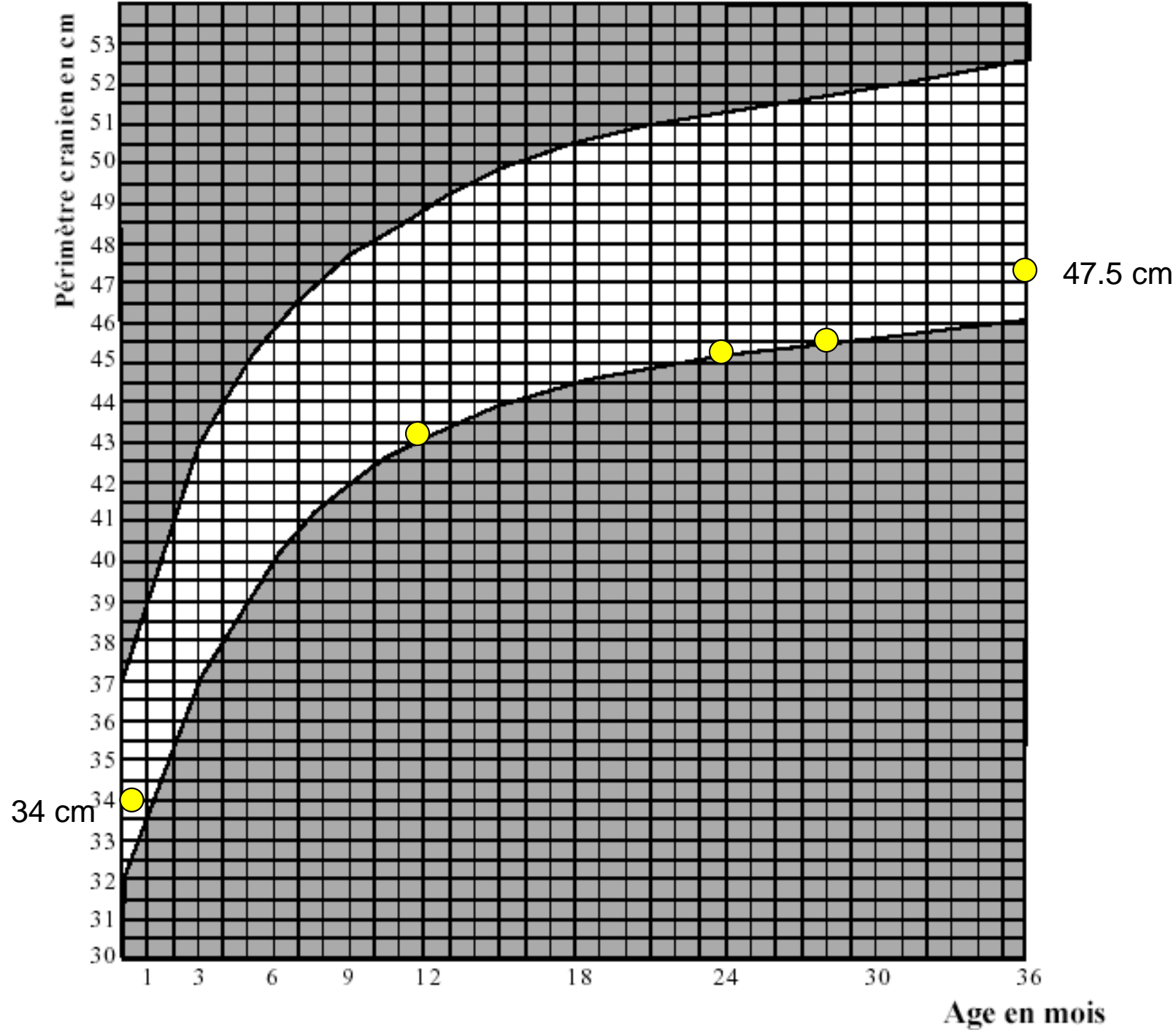




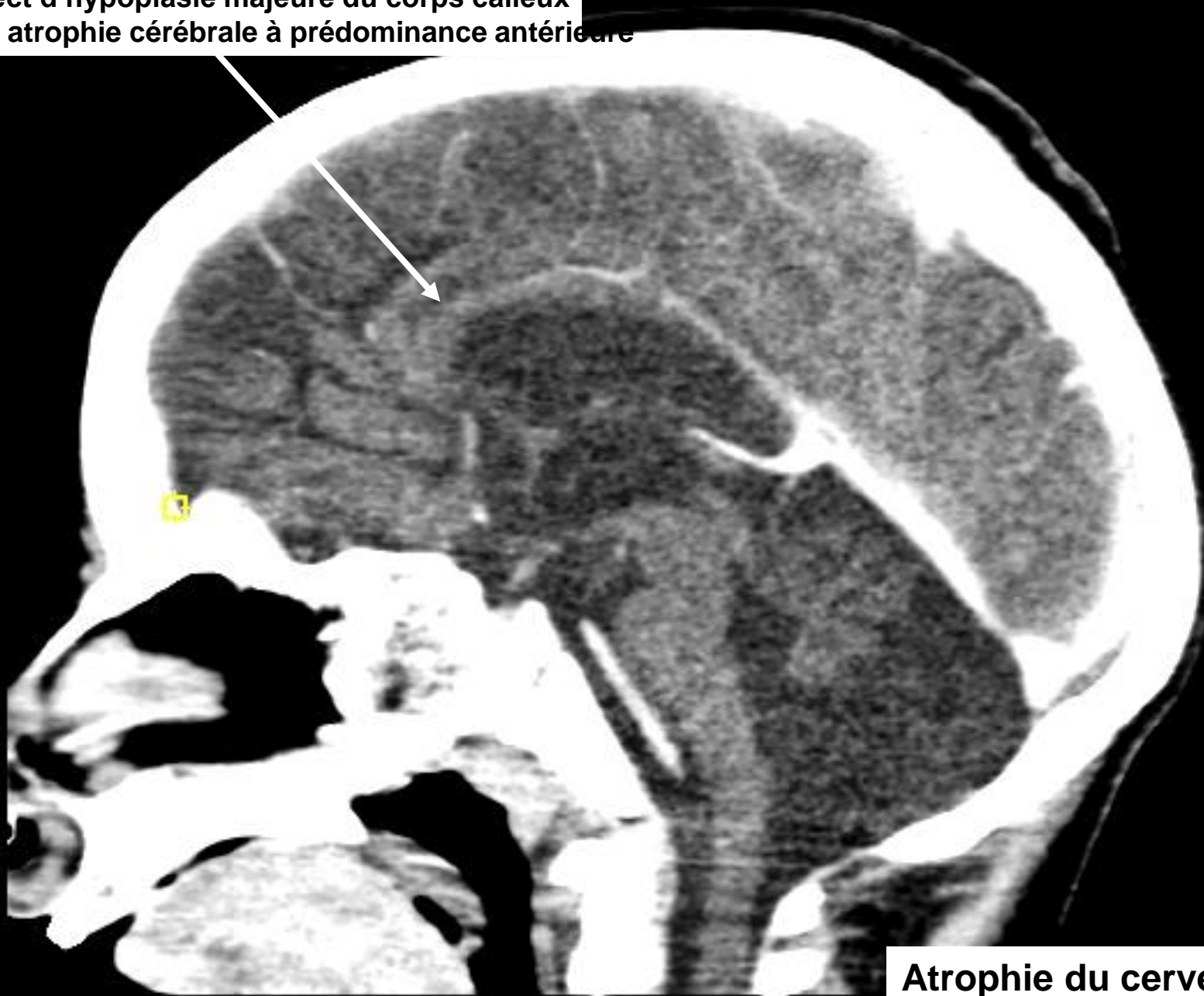




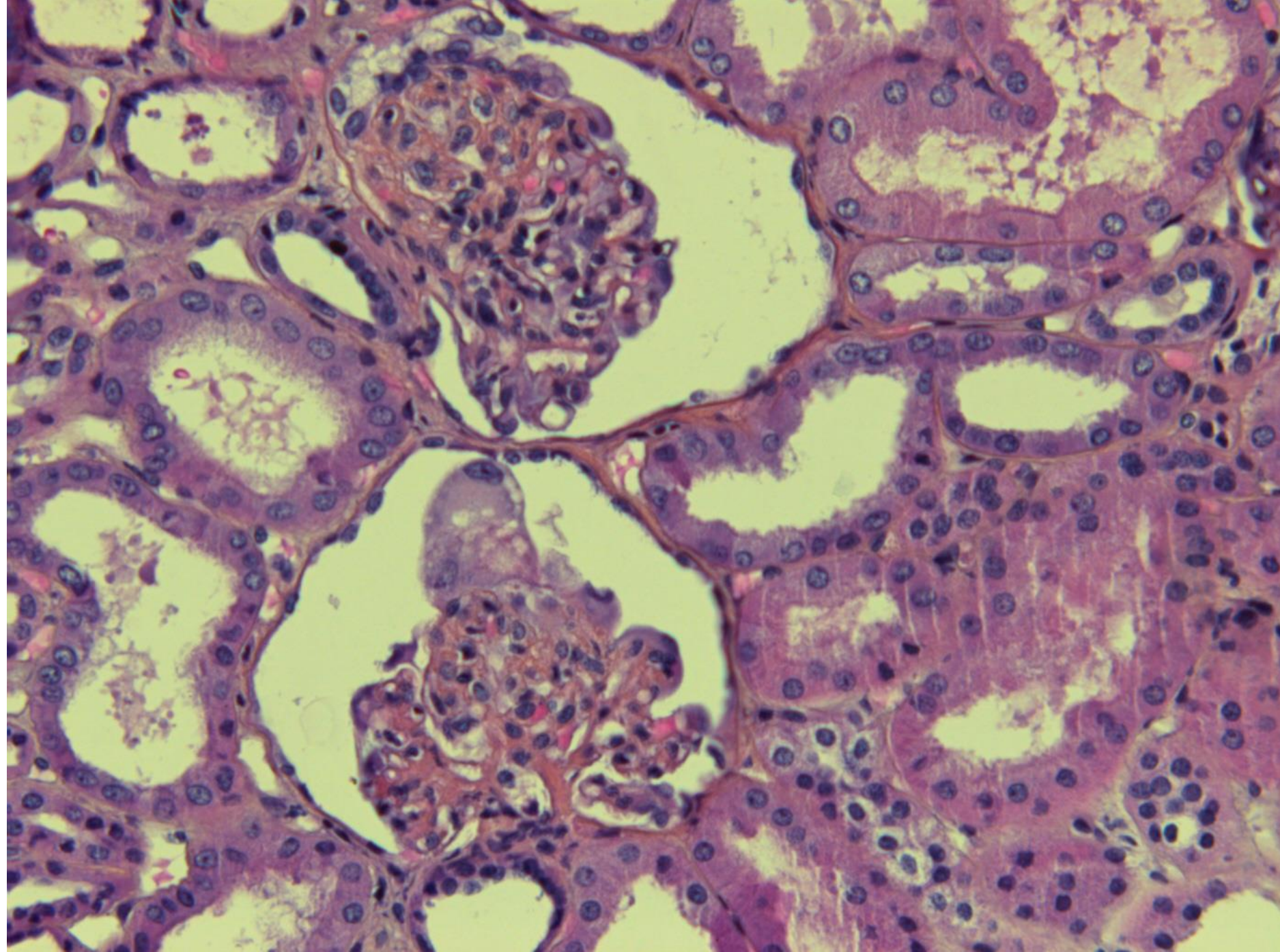




Aspect d'hypoplasie majeure du corps calleux
avec atrophie cérébrale à prédominance antérieure



Atrophie du cervelet

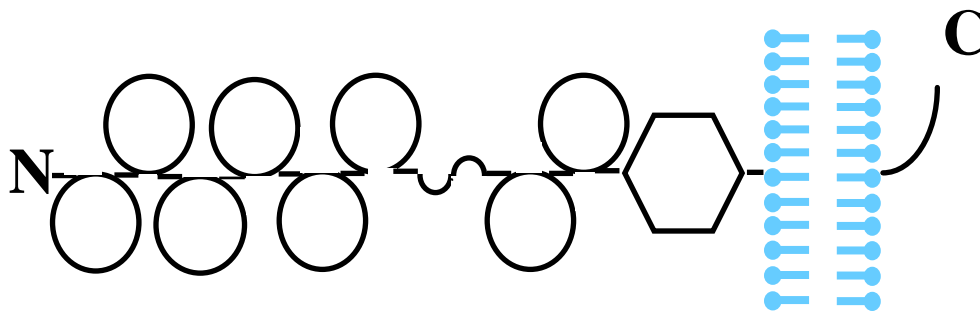
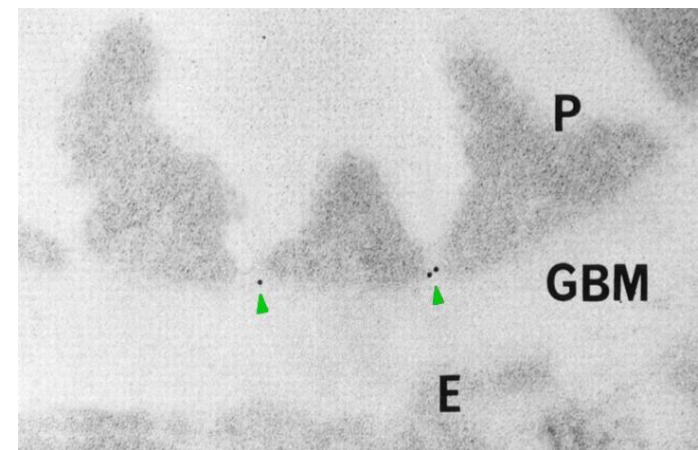
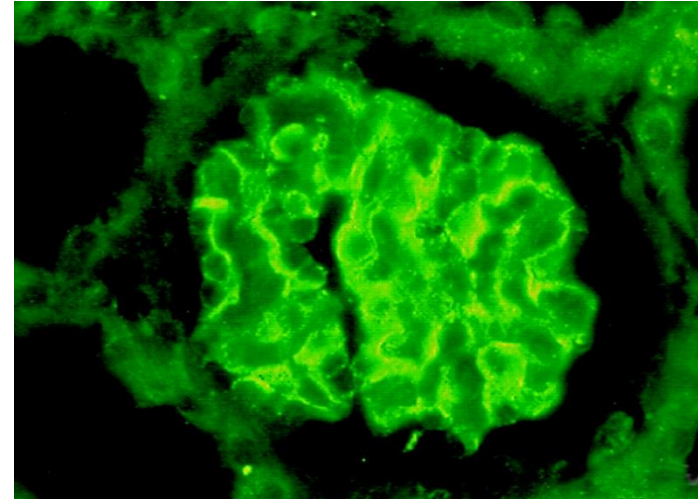


Formes pédiatriques..

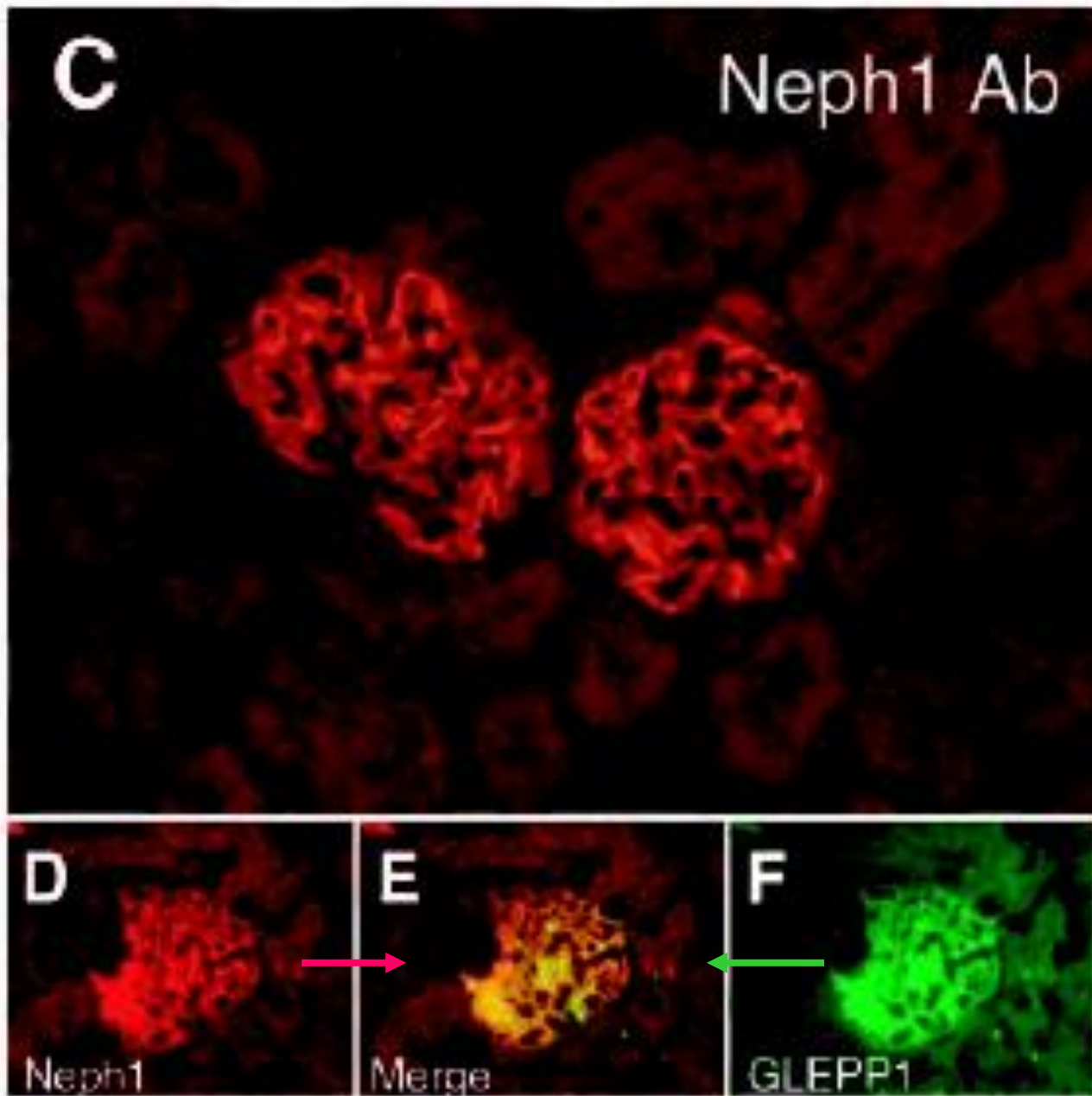
Syndromes Récessifs Autosomiques

Syndrome Néphrotique Finlandais (NPHS1)

- Autosomique récessif.
- 1 : 10 000 Finlande.
- SN massif avec oedème placentaire.
- Devenir péjoratif.
- Mutations de NPHS1 (Néphrine).
- 2 hot-spots en finlande :
 - Fin-maj: 121delCT.
 - Fin-min: R1109X.

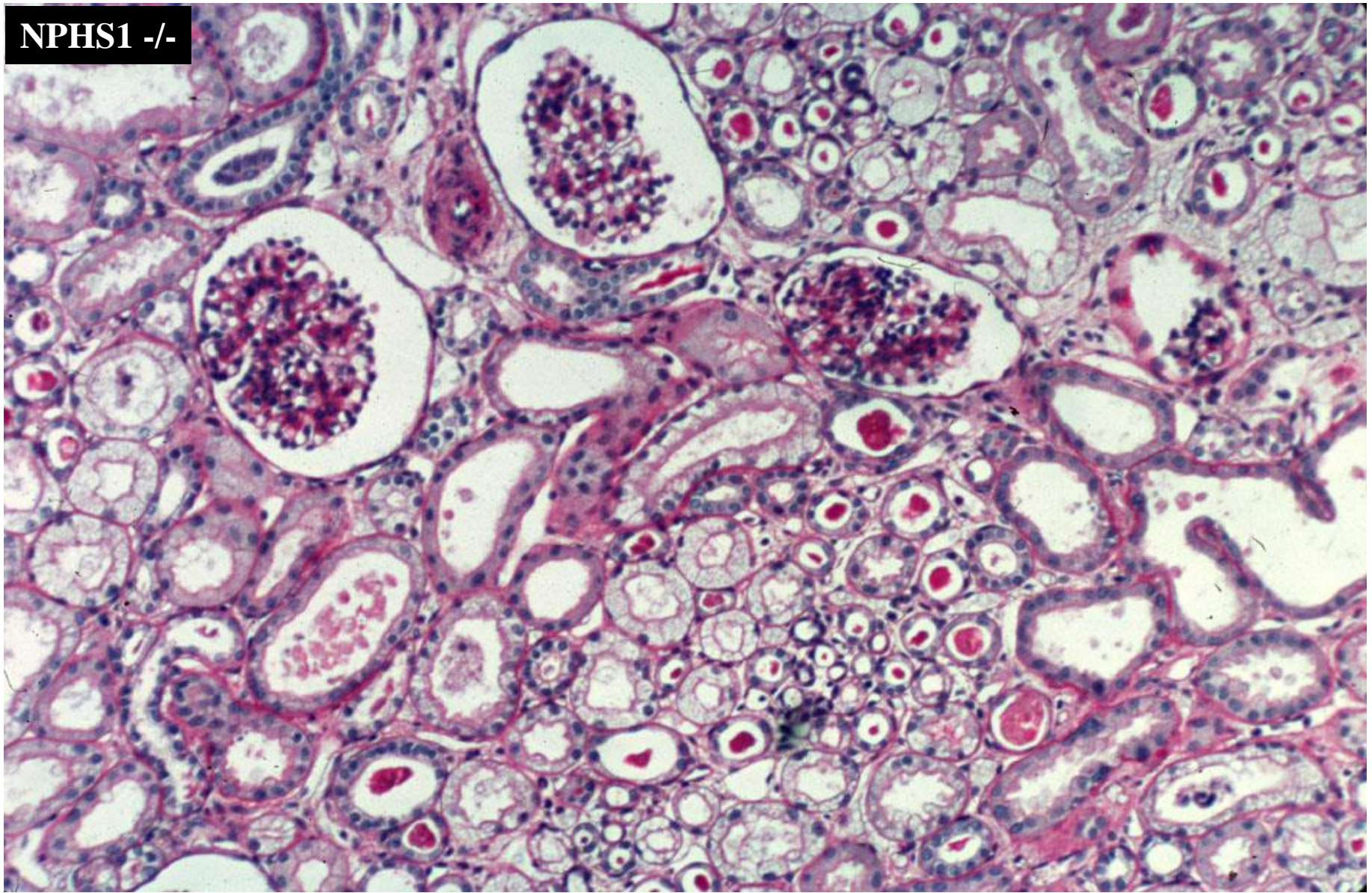


NPHS1: Molécule d'adhérence de la SF des Ig.



Gravité en fonction de la rétention cytoplasmique de NPHS1

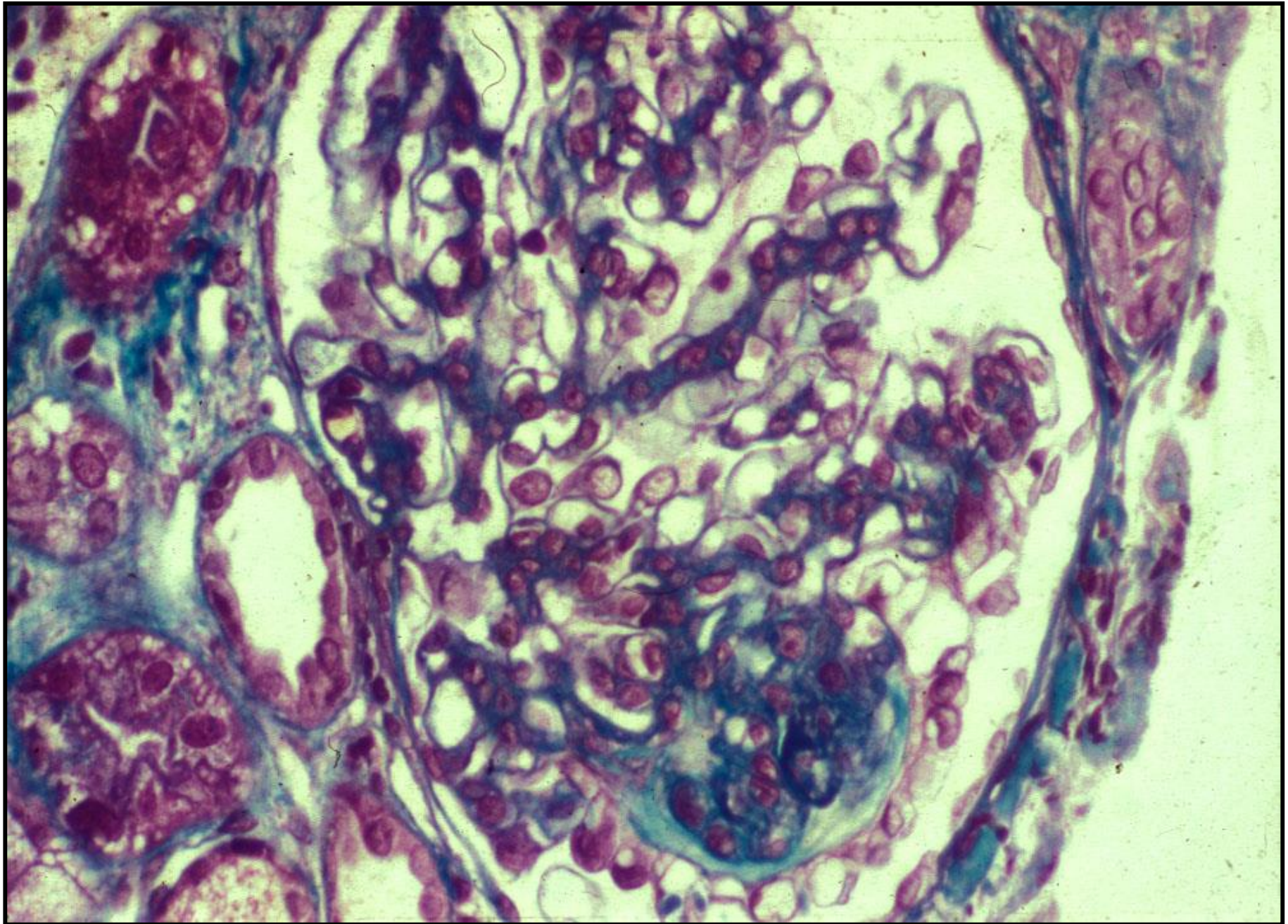
NPHS1 -/-



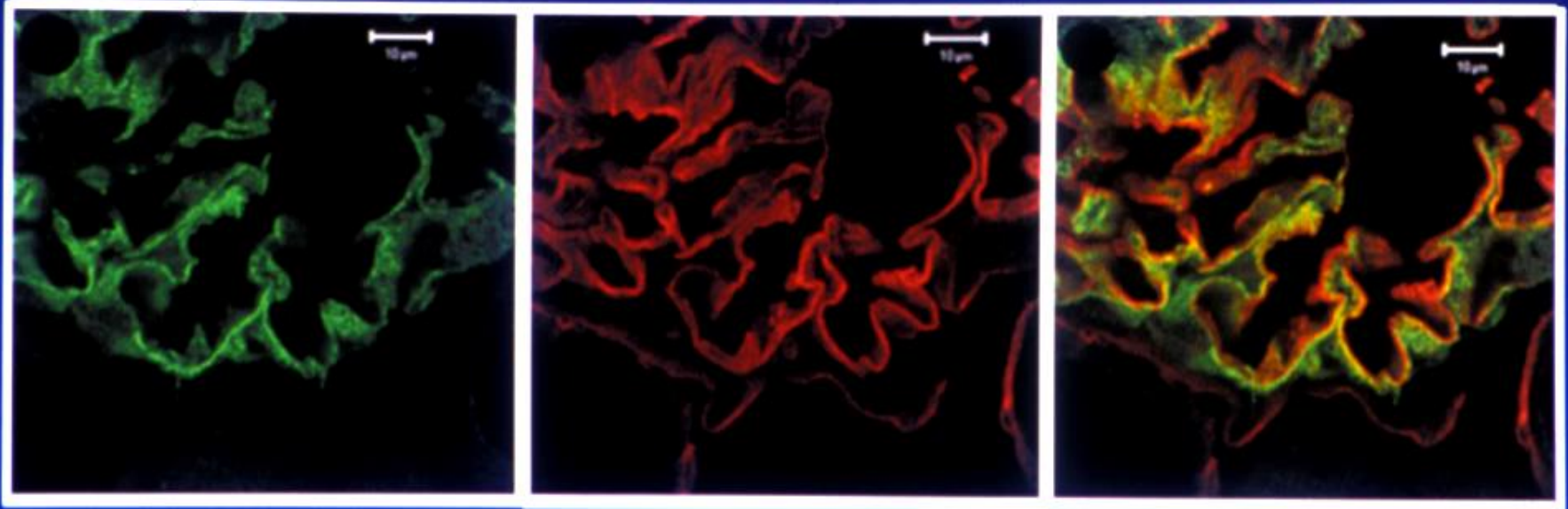
Lésions non spécifiques ++.
Dilatations des tubes +/-.

Syndrome Néphrotique Cortico-Résistant lié à NPHS2 (podocine)

- **Début précoce (1ère ou 2ème année) .**
- **Résistance à tout traitement ++.**
- **Progression rapide vers l'IRCT (< 10 ans)**
- **LGM puis Hyalinose segmentaire et focale typique.**
- **Mais : ...**
 - **Formes « sporadiques » .**
 - **Formes congénitales précoces, ou tardives.**
 - **Récidives post-transplantation (25%)**
- **Formes de l'adulte avec 1 mutation + polymorphisme R229Q (polymorphisme retrouvé dans 4% de la population et qui diminue l'efficacité de la liaison à la néphrine).**



NPHS2 dans le rein mature



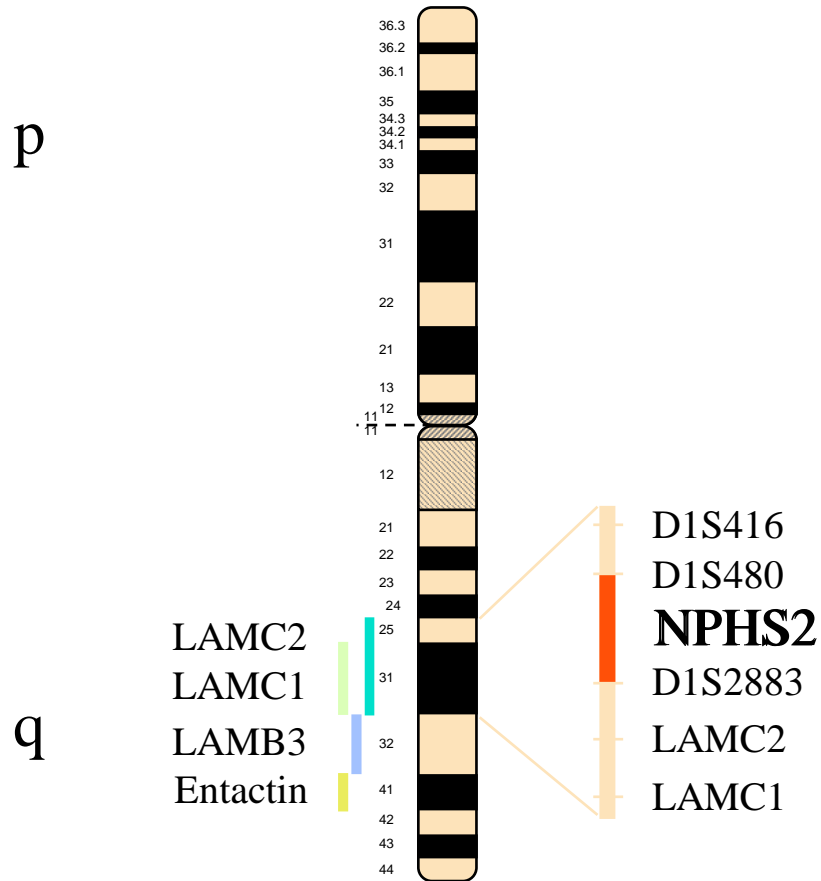
Podocin

Col (IV) α 3

Merge

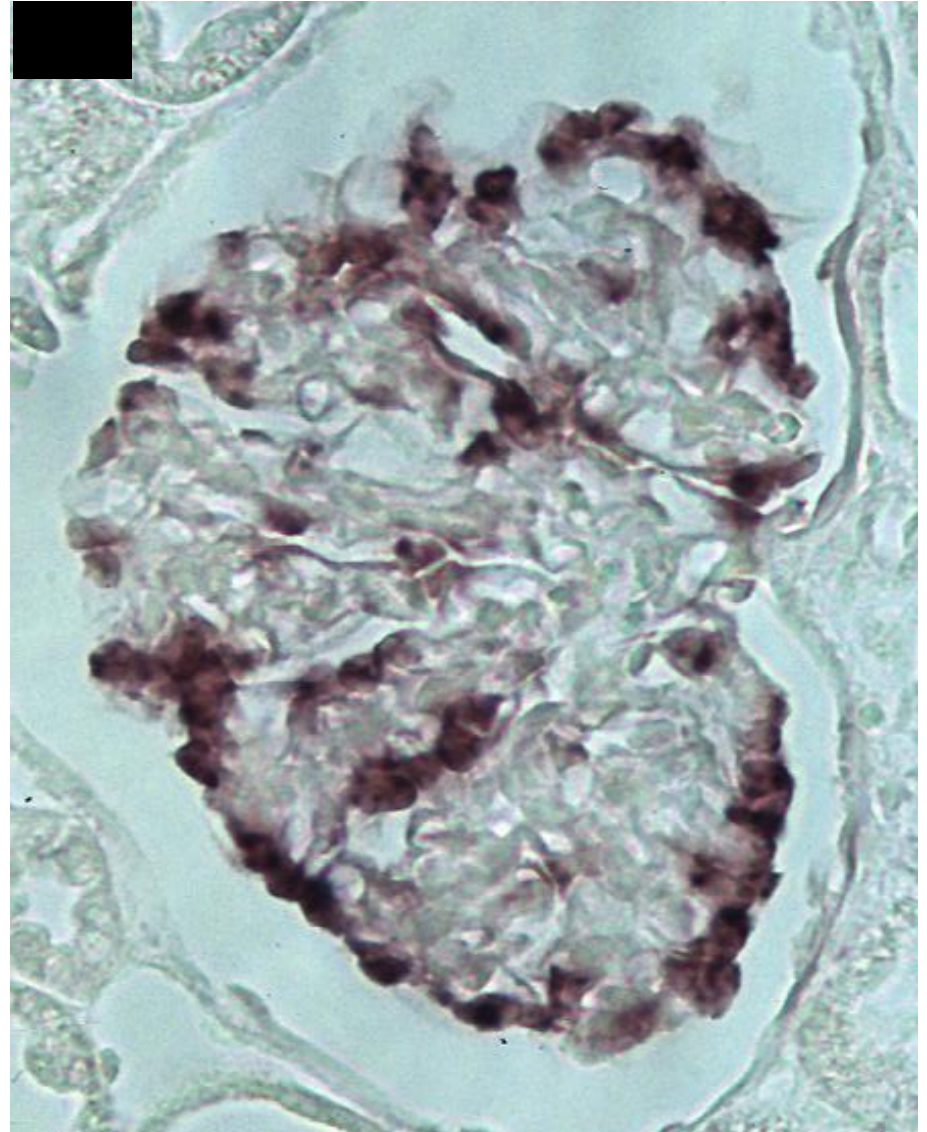
Si mutation de la podocine: IF variable: normale ou séquestration anormale dans le cytoplasme. L'expression de NPHS1 peut-être également modifiée.

La podocine est régulée par le facteur lmx 1B (Nail-patela syndrome)



CHROMOSOME 1

NPHS2: REGION 1q25-31



- **Homme de 21 ans.**
- **Protéinurie 2.04g/L.**
 - **protidémie 56g/L.**
 - **albuminémie 32 g/L.**
 - **sans leucocyturie ni hématurie.**
- **Fonction rénale normale.**
 - **Créatininémie: 66 $\mu\text{mol/L}$.**

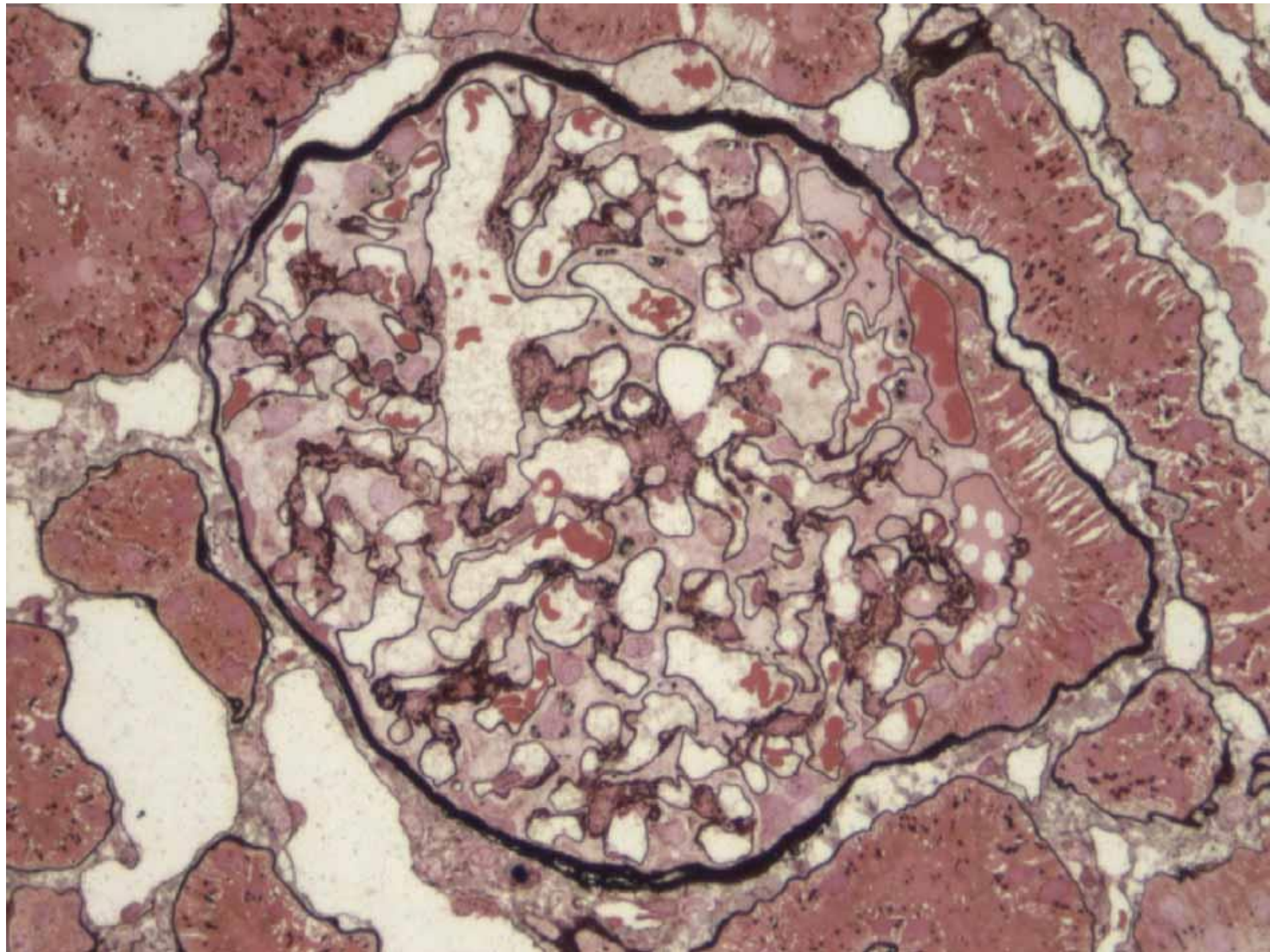


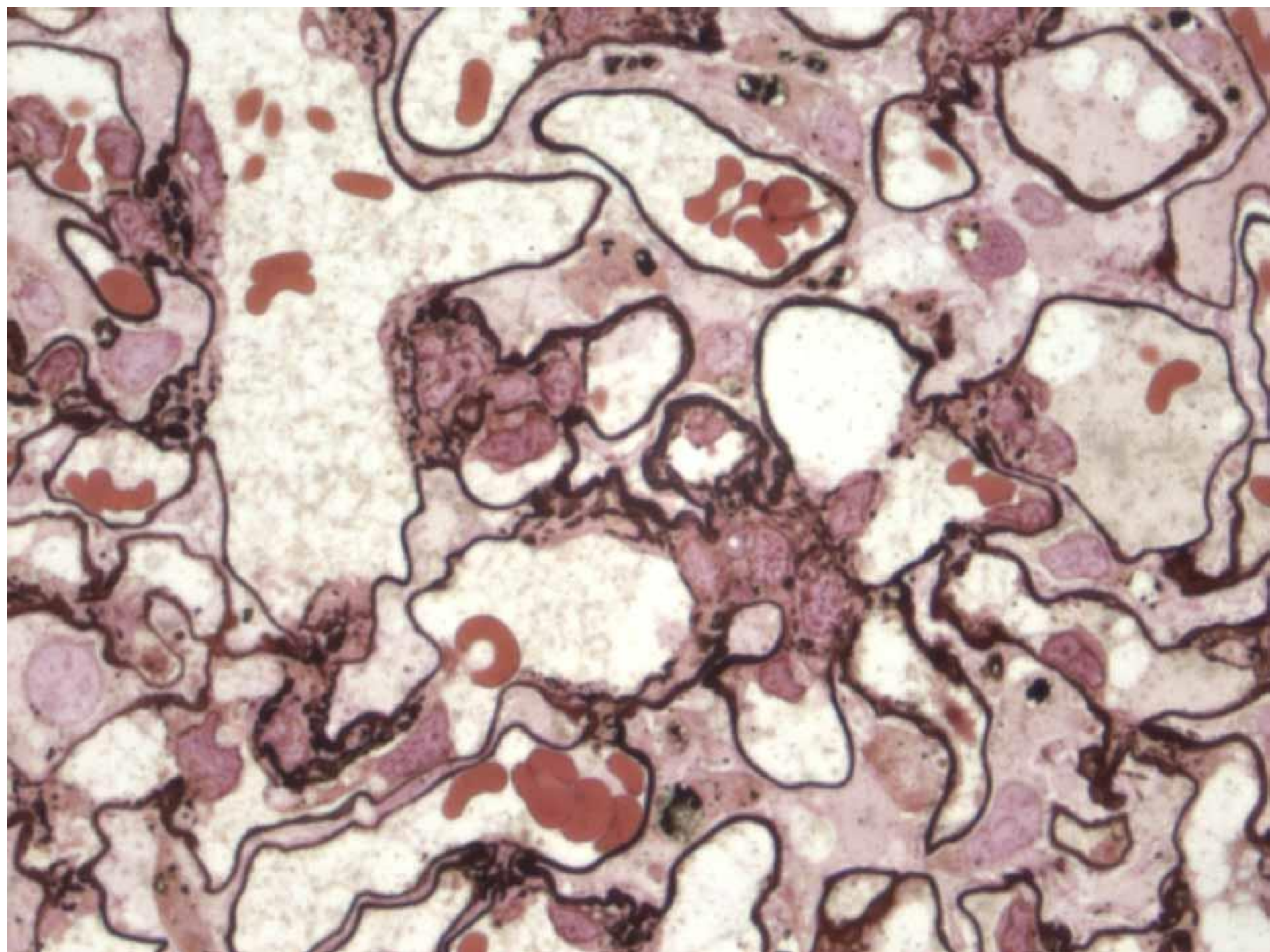
D

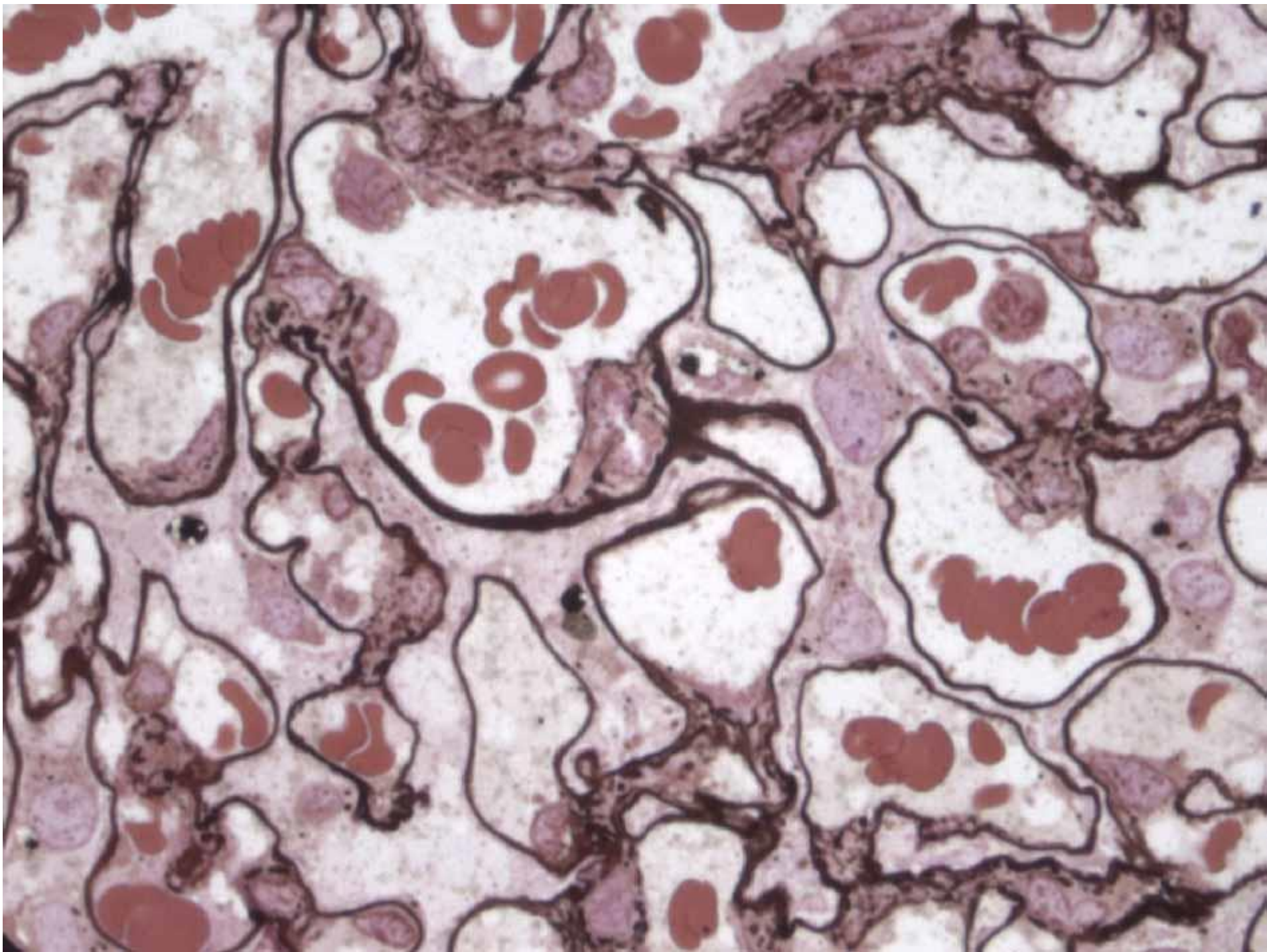
DEBOUT

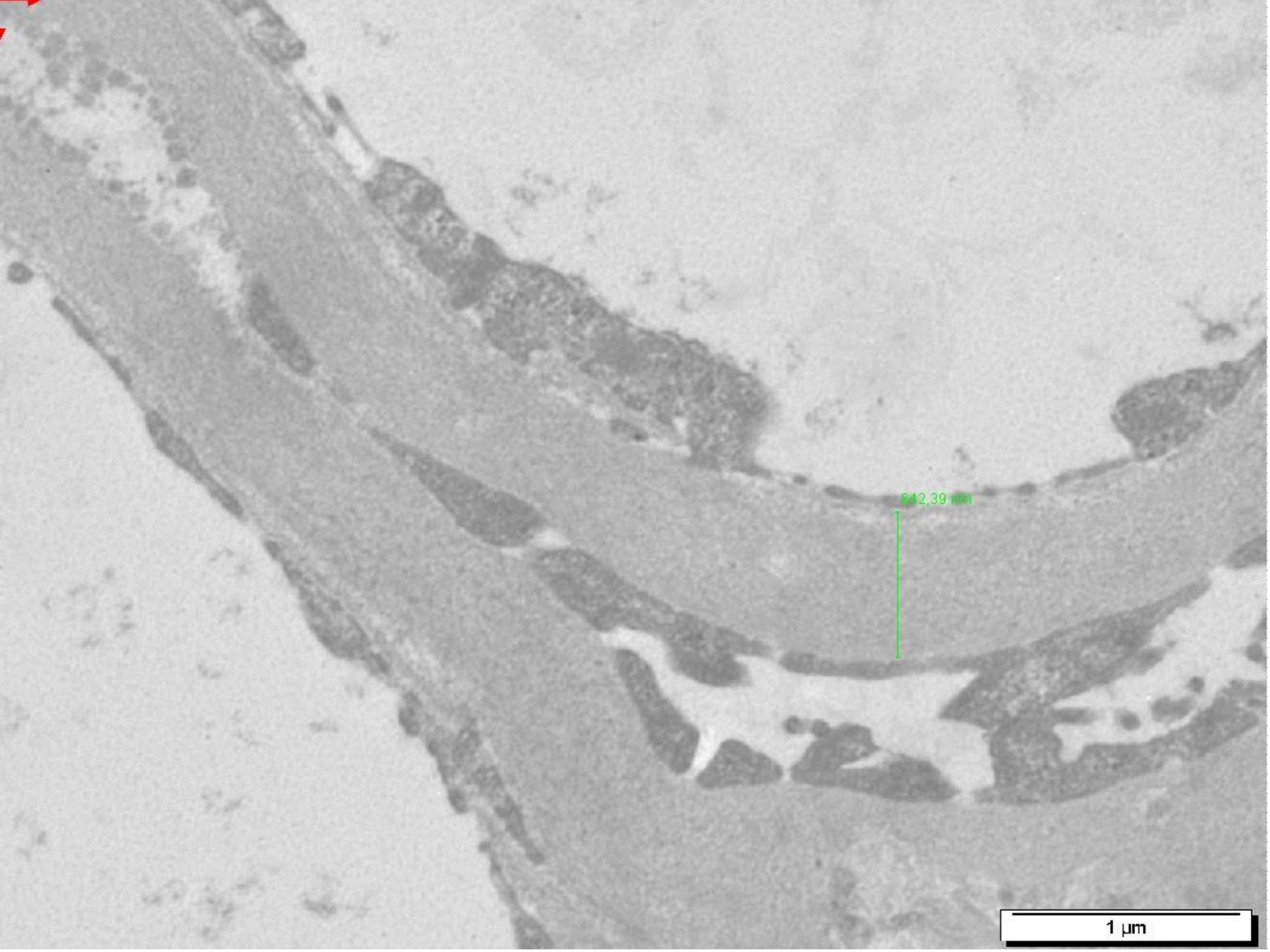
C





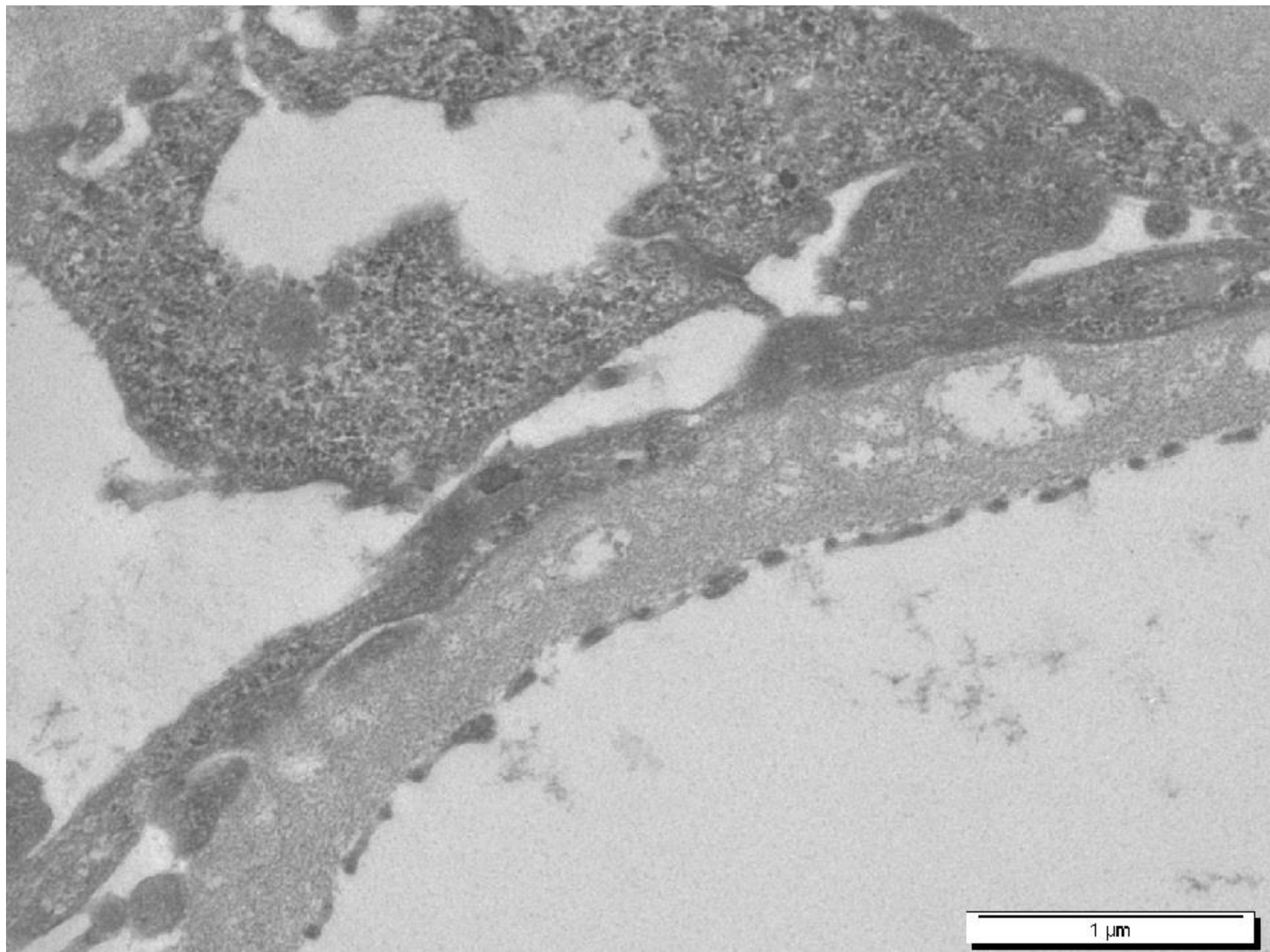




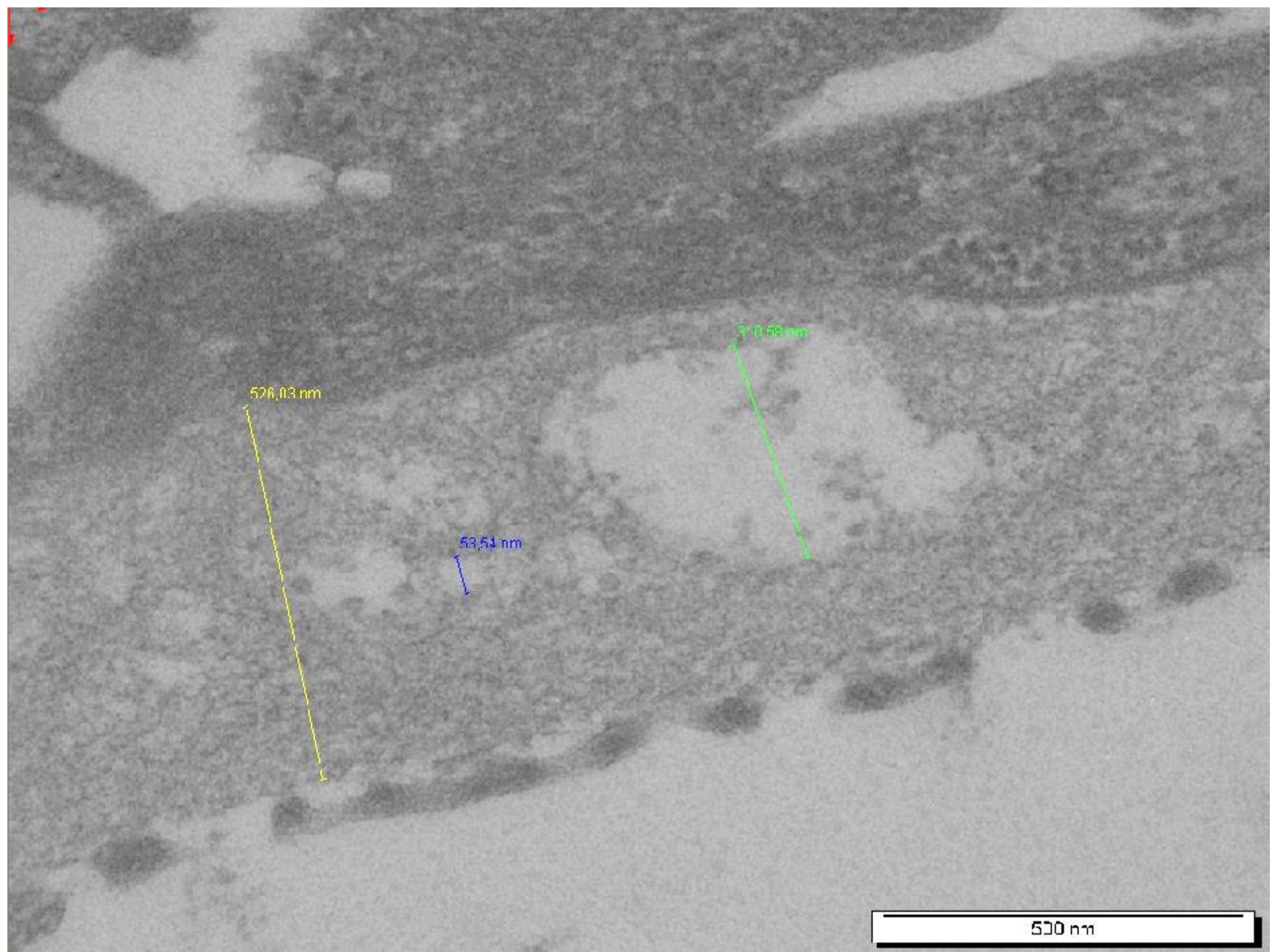


642.39 nm

1 μm



1 μm

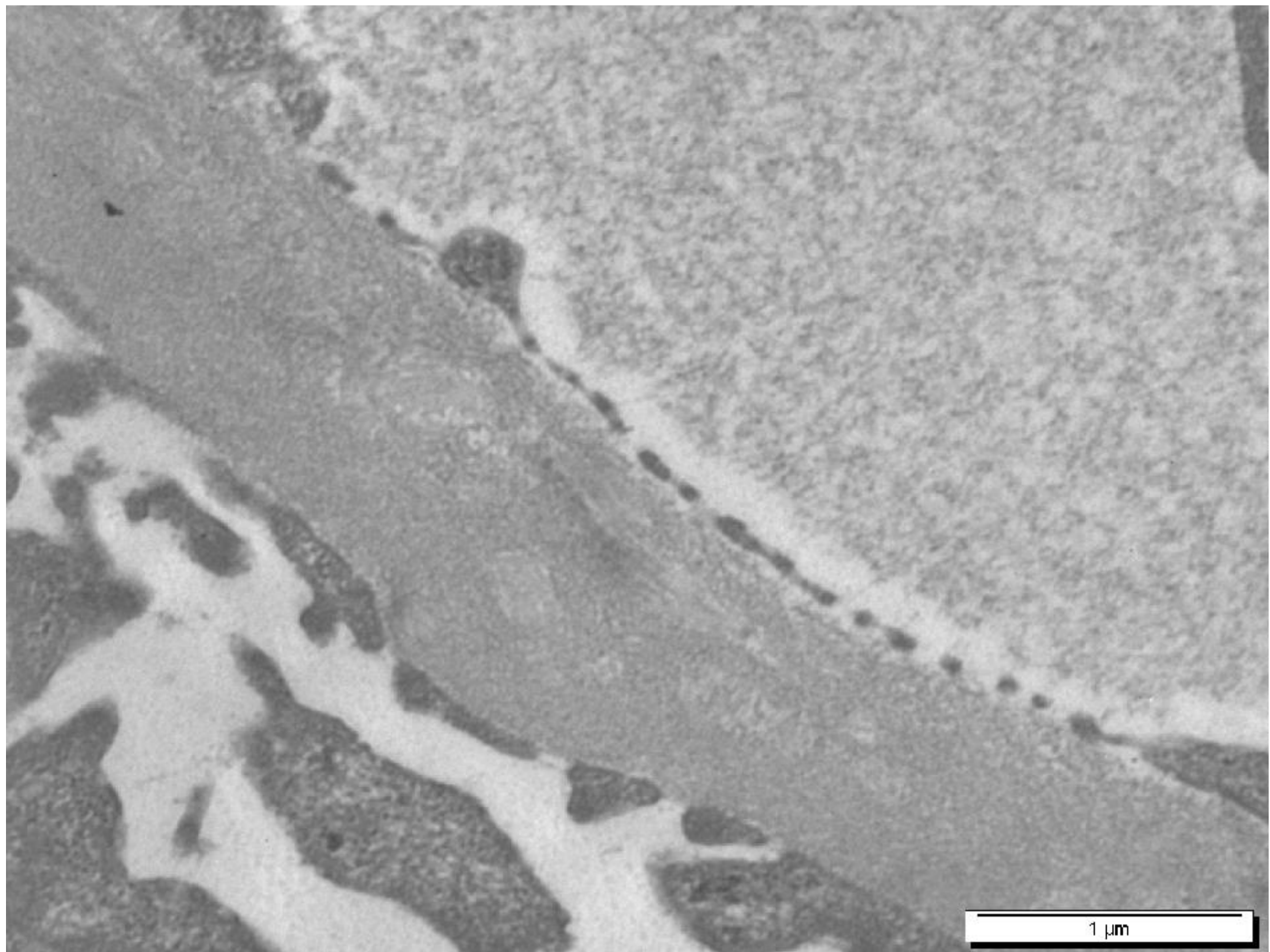


526.03 nm

53.54 nm

910.58 nm

500 nm



Syndrome de Pierson

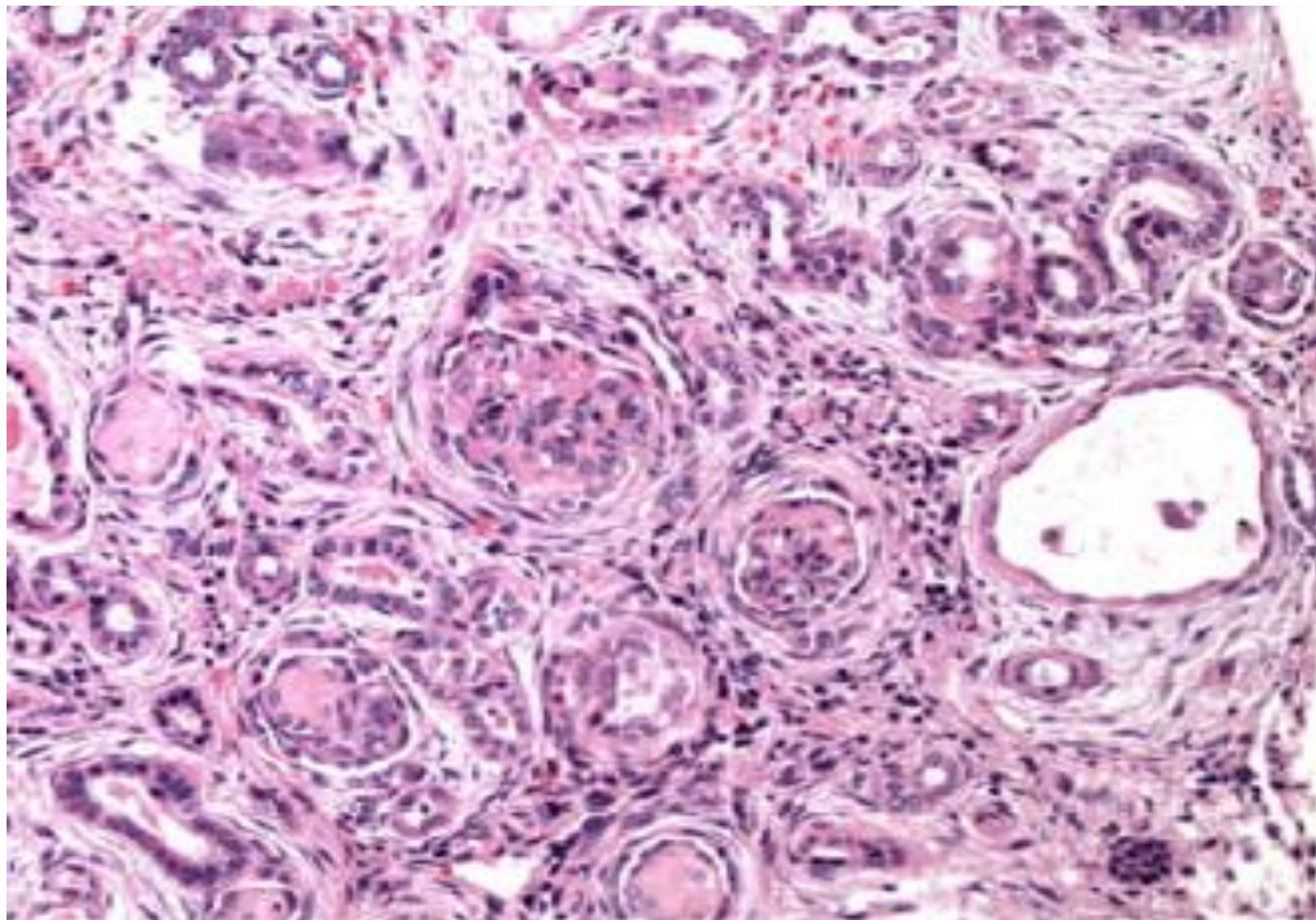
Mutations de LAMB2 codant la laminine $\beta 2$ exprimée dans la MBG



+/- Aspect de buphtalmie



**Microcorie aréactive
Retard psycho-moteur
Anomalies musculaires**



- **SN congénital ou post-natal immédiat.**
- **Sclérose mésangiale diffuse et micro-glomérules.**
(autre cas avec SMD: PLCe1)

**Syndromes
Autosomiques
Dominants**

HSF et mutations de ACTN-4 (19q13)

- **Détection, dans 3 grandes familles (oklahoma, californie, et îles canaries), de 3 mutations différentes dans l'exon 8, modifiant des acides aminés très conservés (de l'homme à C elegans).**
- **Augmente la liaison de l' α -actinine-4 aux filaments d'actine.**
- **Altération des caractéristiques mécaniques du podocyte ?
Aggrégation et rapide dégradation de la protéine.**
- **HSF classique.**

Locus FSGS-2 = gène TRPC6.

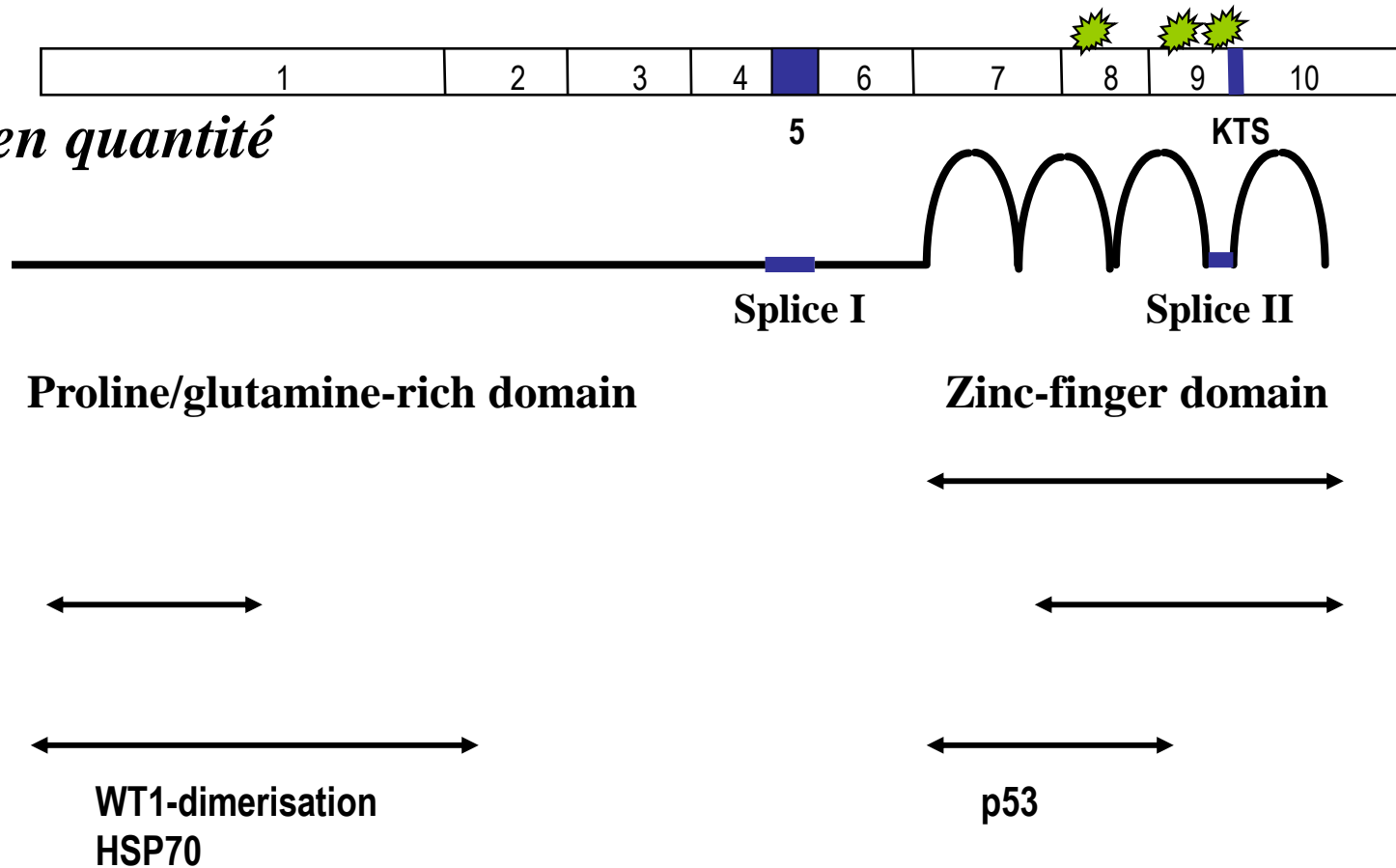
- Localisation d'une gène en 11q21-q22 dans une grande famille présentant une protéinurie abondante à l'âge adulte, avec progression vers l'IRCT dans 60% des cas. (Conlon PJ, KI 1999; 55: 1241.)
- Mutation faux-sens du gène *TRPC6* codant un canal ionique (Winn et al., ASN, St-Louis, 2004)
- Cas caucasiens (Italie +++; PACA) et asiatiques.

Protéine CD2AP: modèle murin et 2 pts avec mutations hétérozygotes (rôle de la protéine dans la dégradation de la néphrine).

Cas publiés avec des LGM (Gigante M et al, NDT 2009).

WT1

(4 isoformes en quantité régulée)

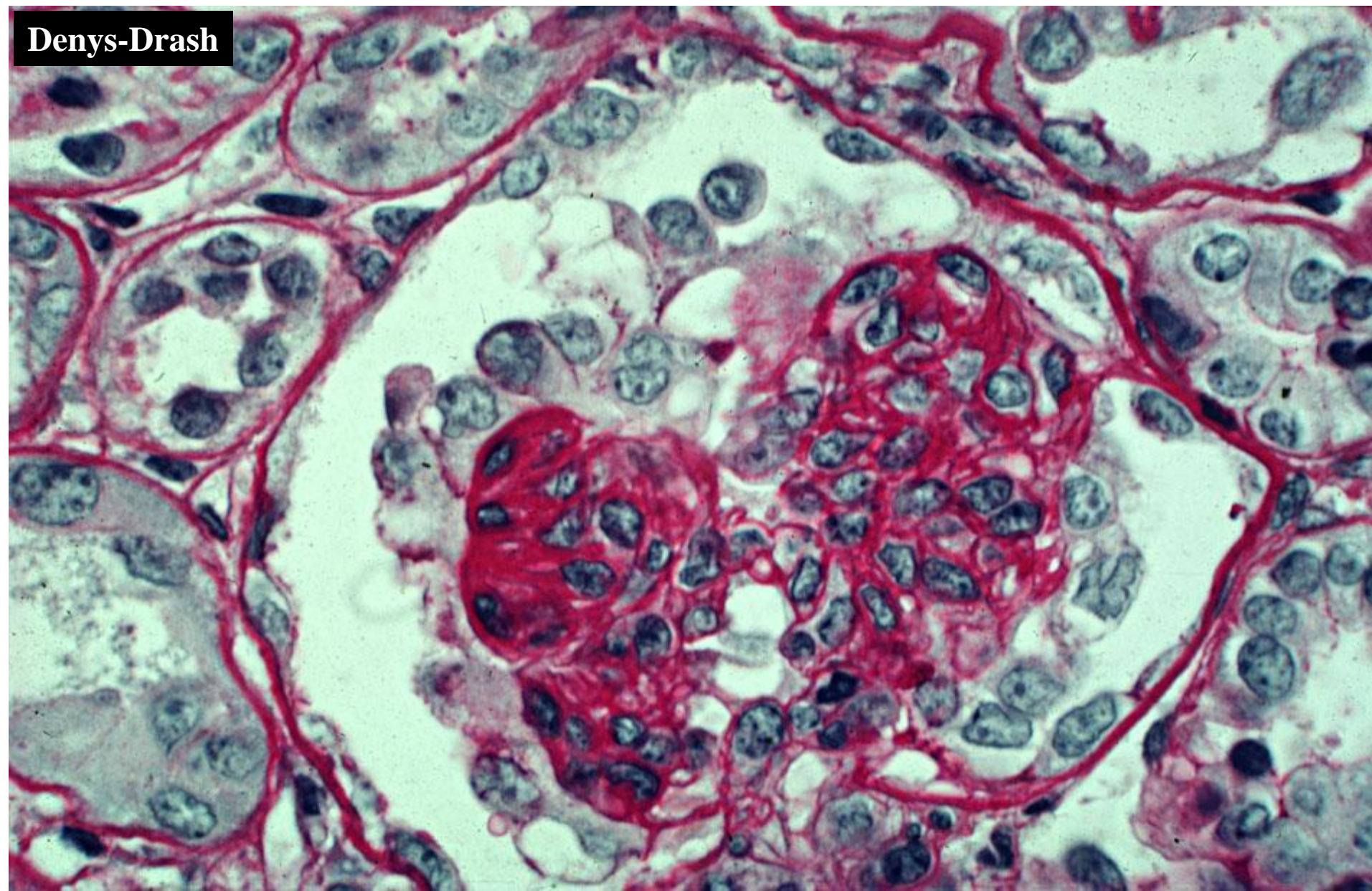


Syndrome de Denys Drash : mutations dans les exons 8-9. **SMD**

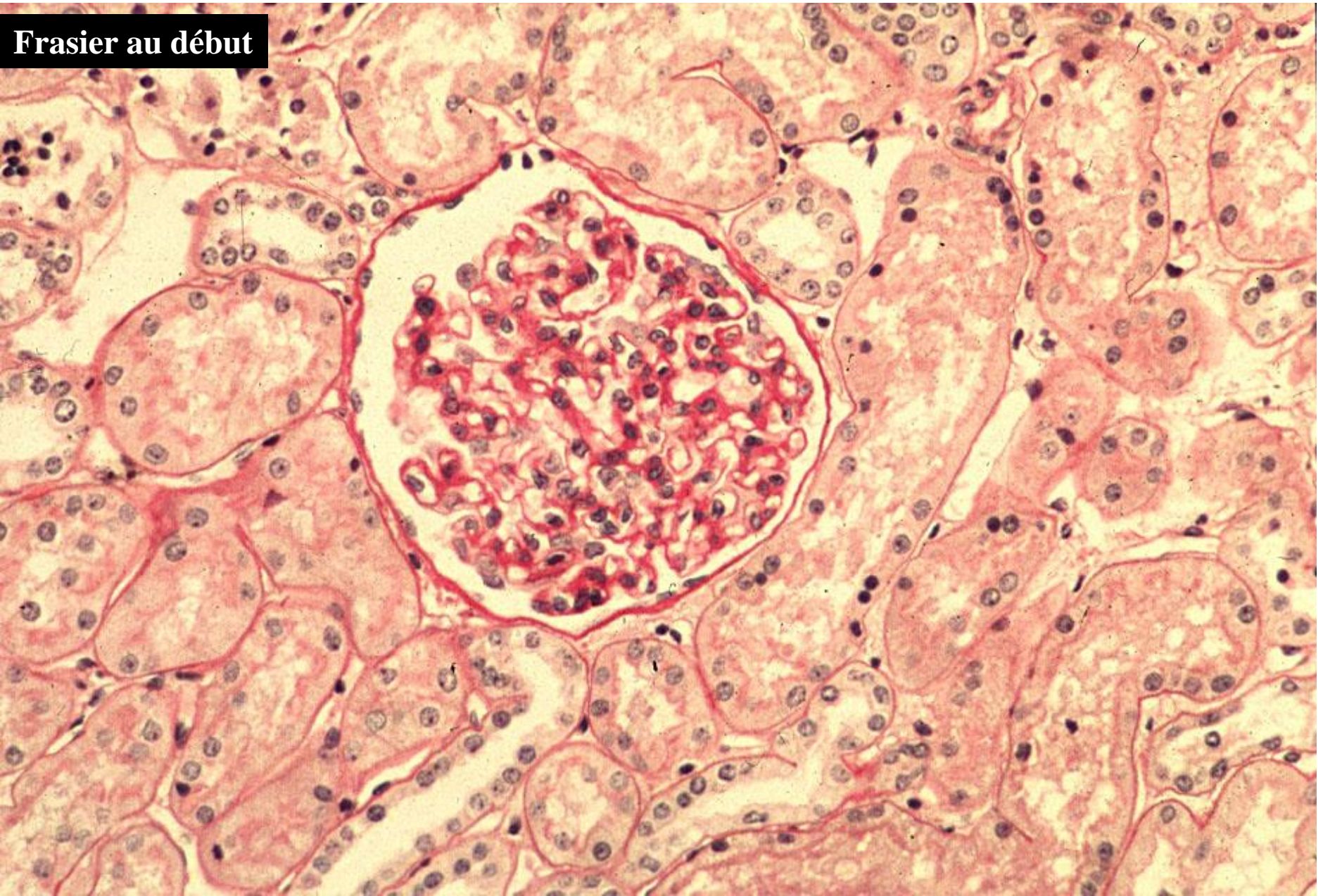
Syndrome de Frasier : mutations dans le site d'épissage « KTS » de l'intron 9. **HSF typique**

WT1 est un facteur de régulation de NPHS1.

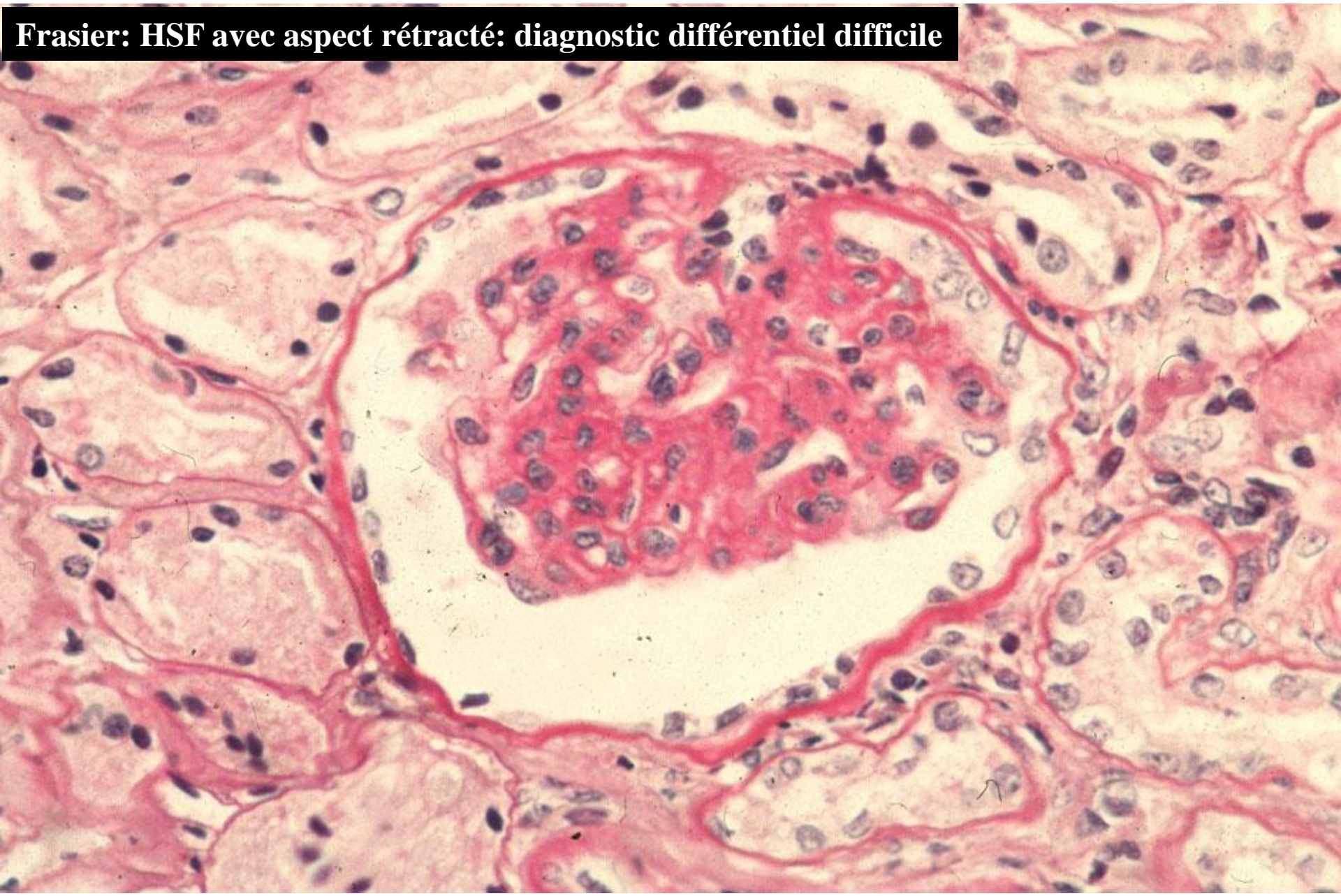
Denys-Drash



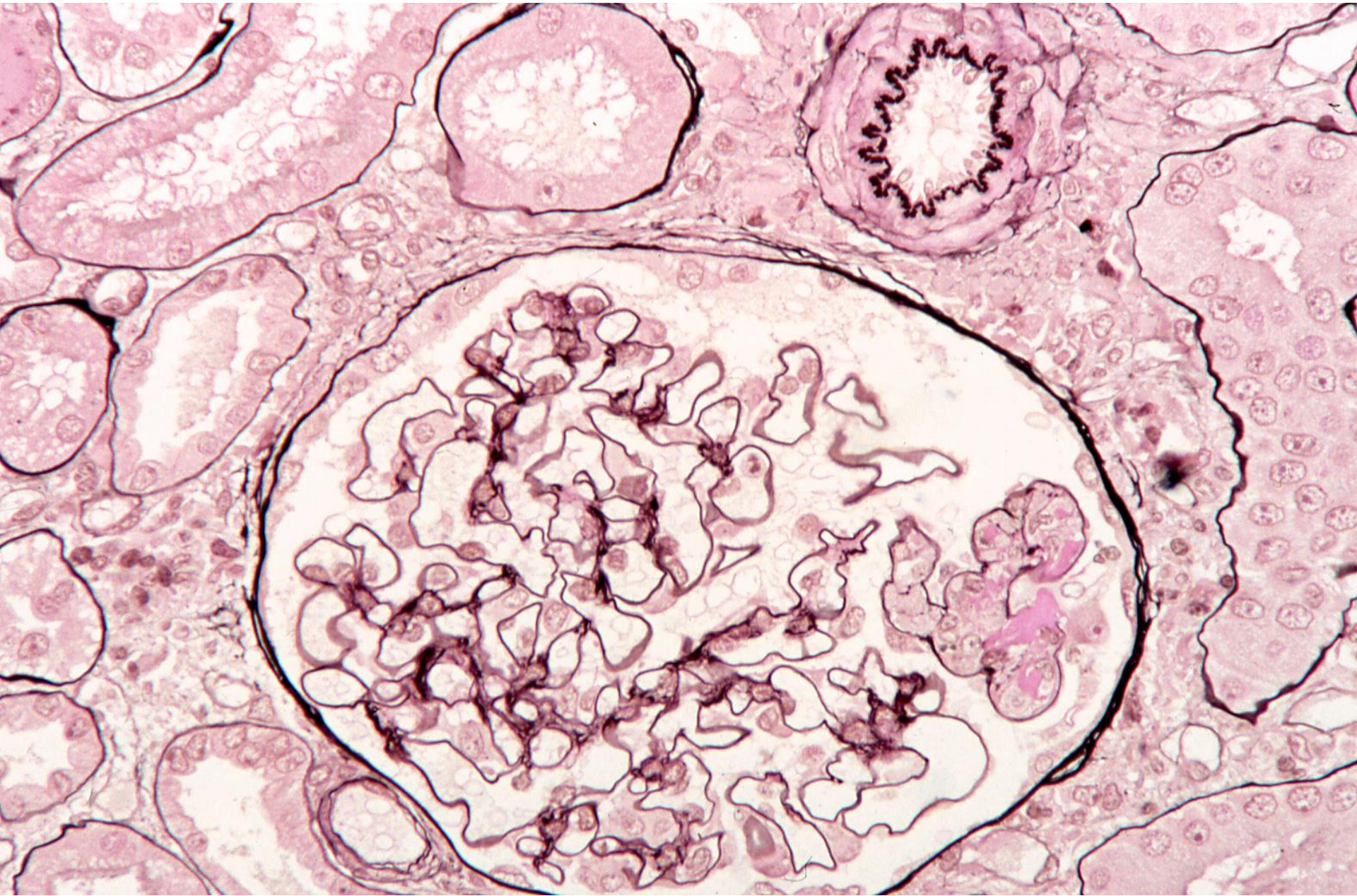
Frasier au début

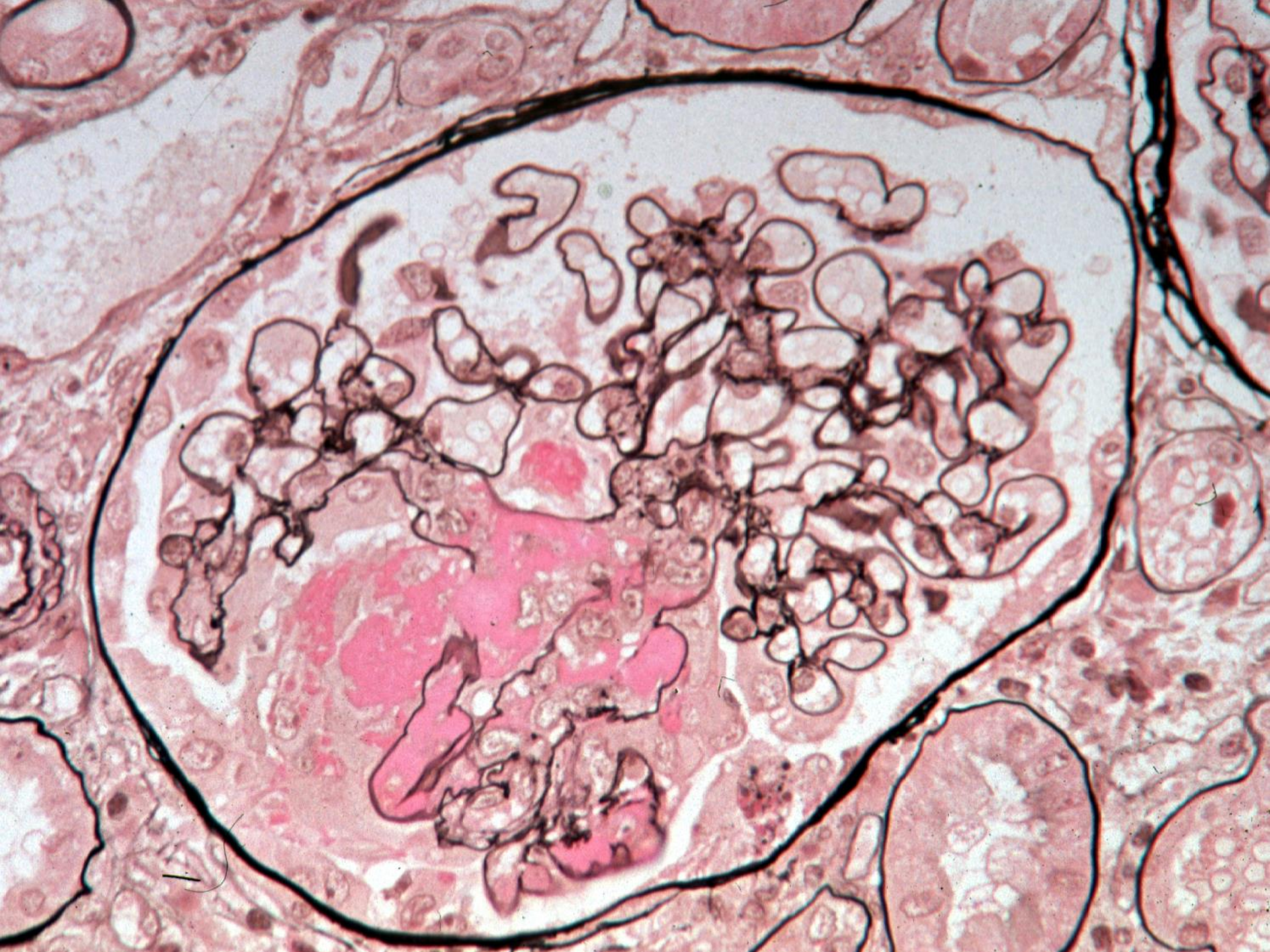


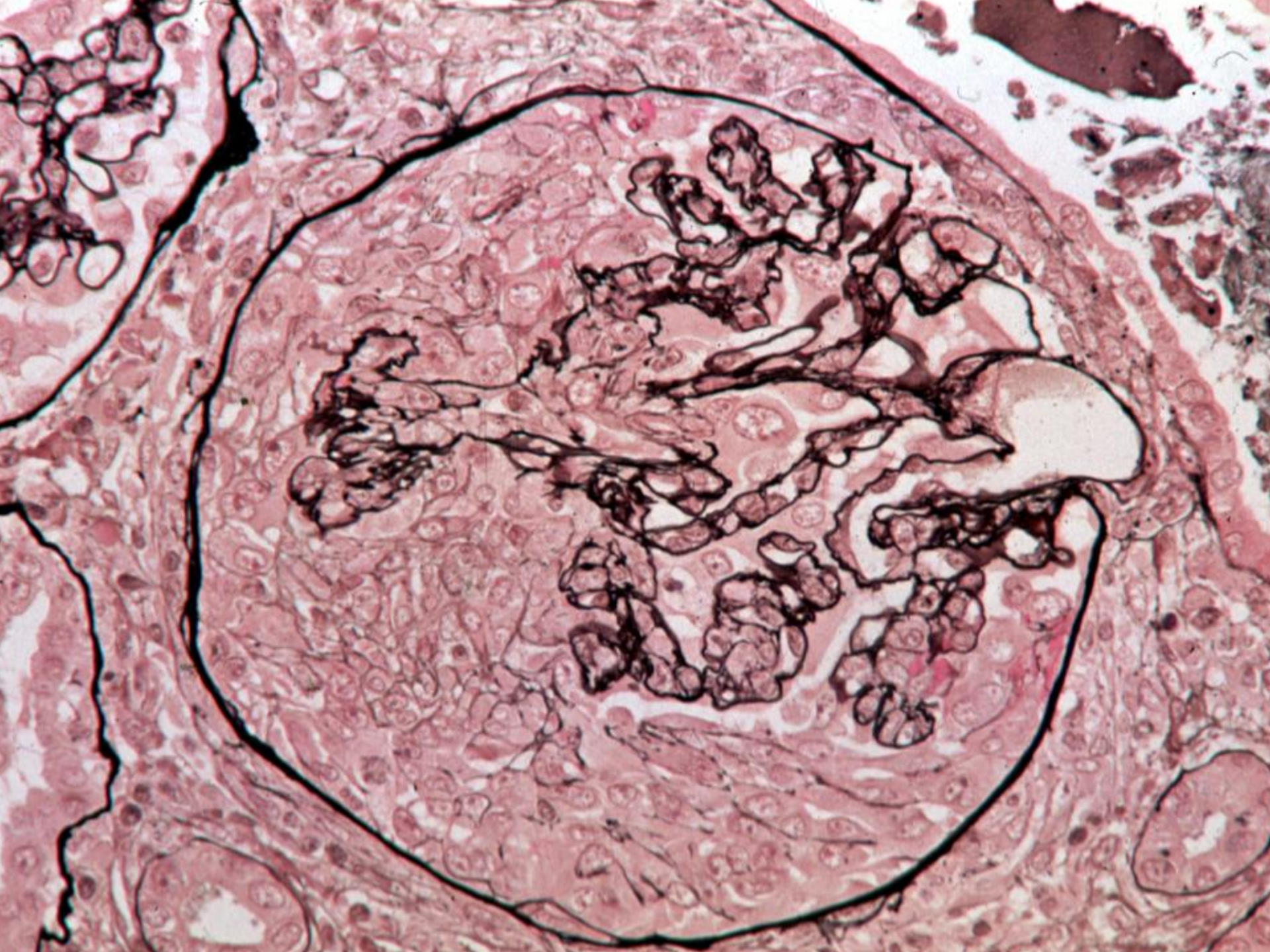
Frasier: HSF avec aspect rétracté: diagnostic différentiel difficile

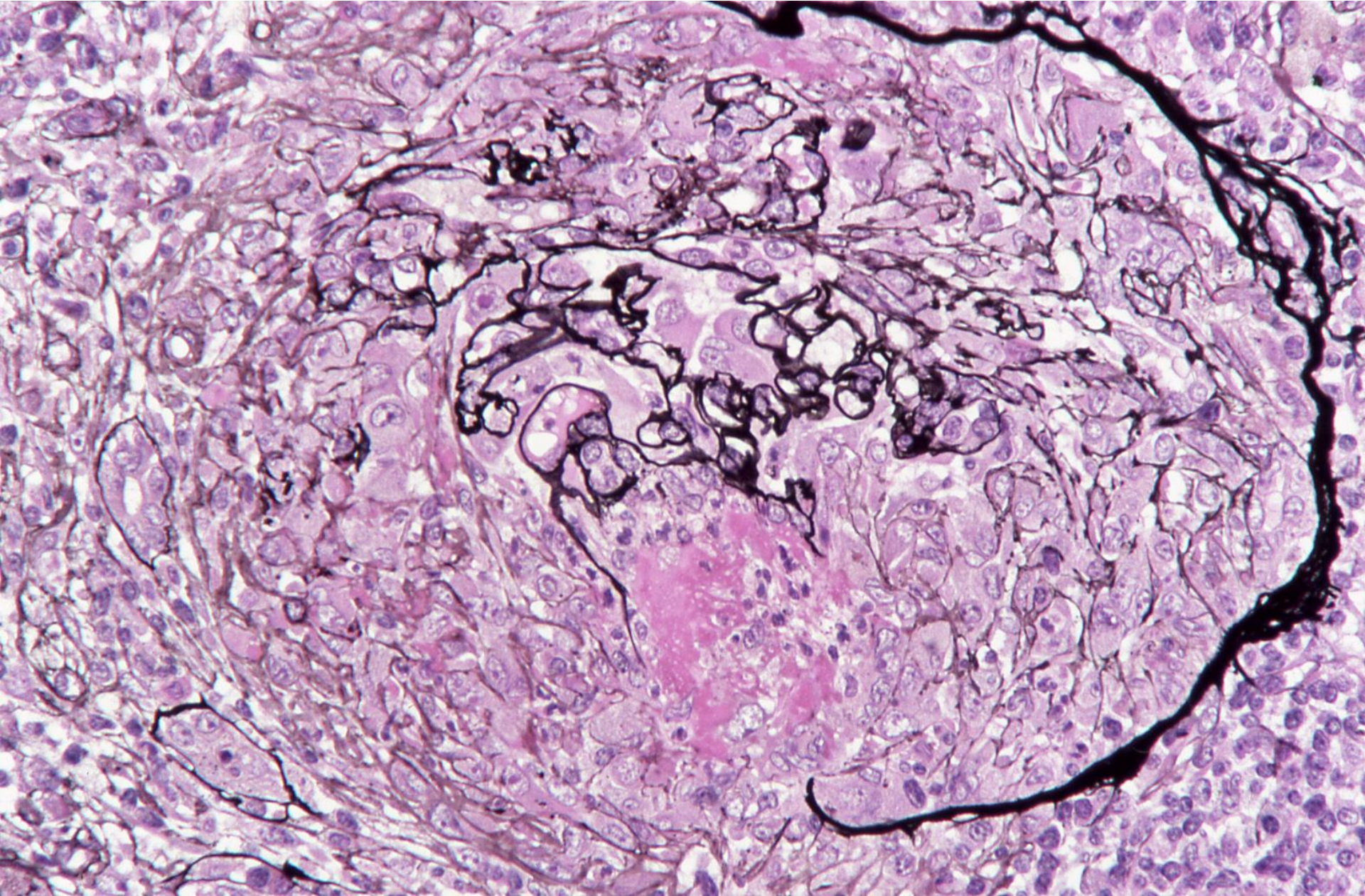


Les glomérulopathies proliférantes extracapillaires

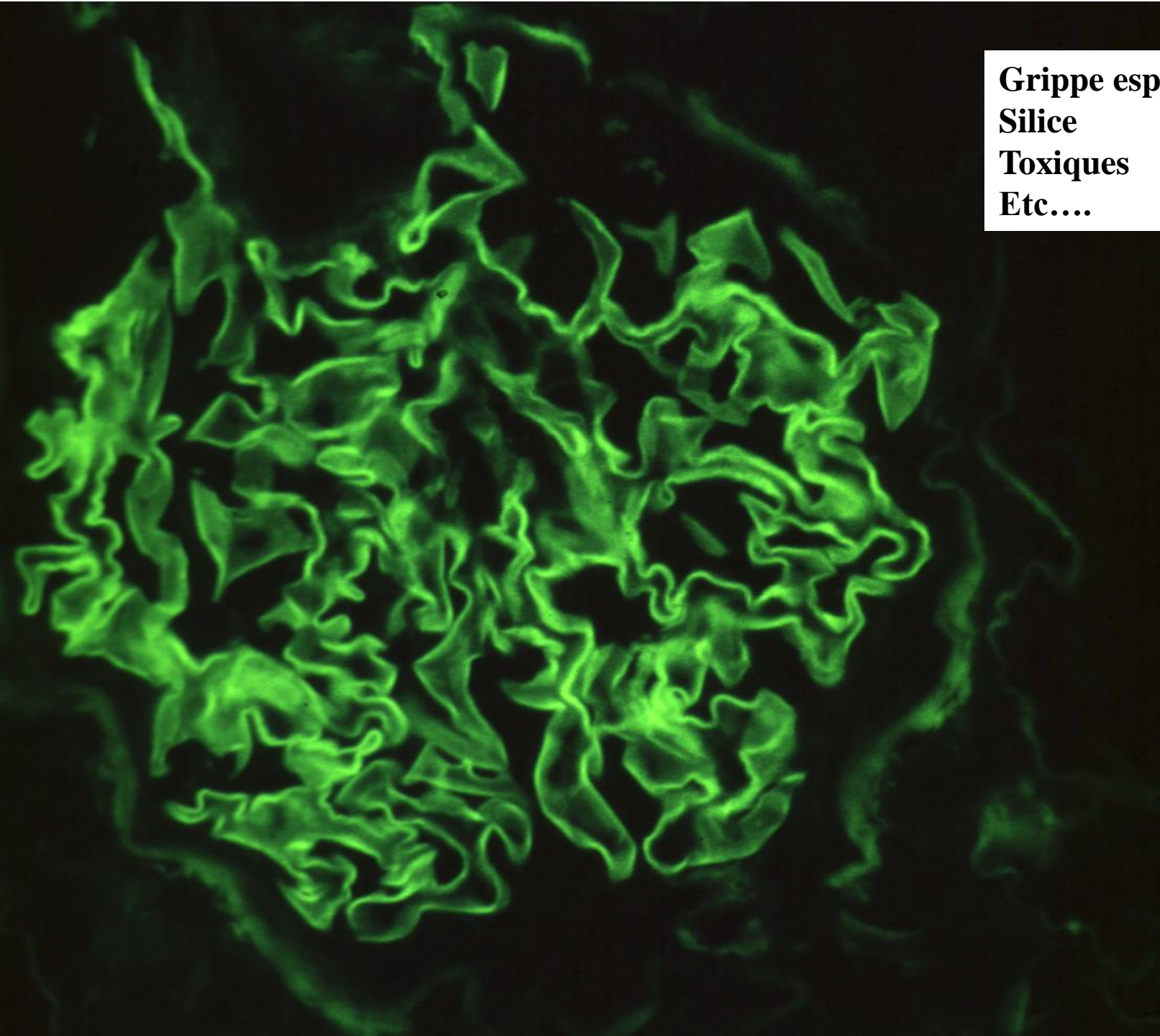


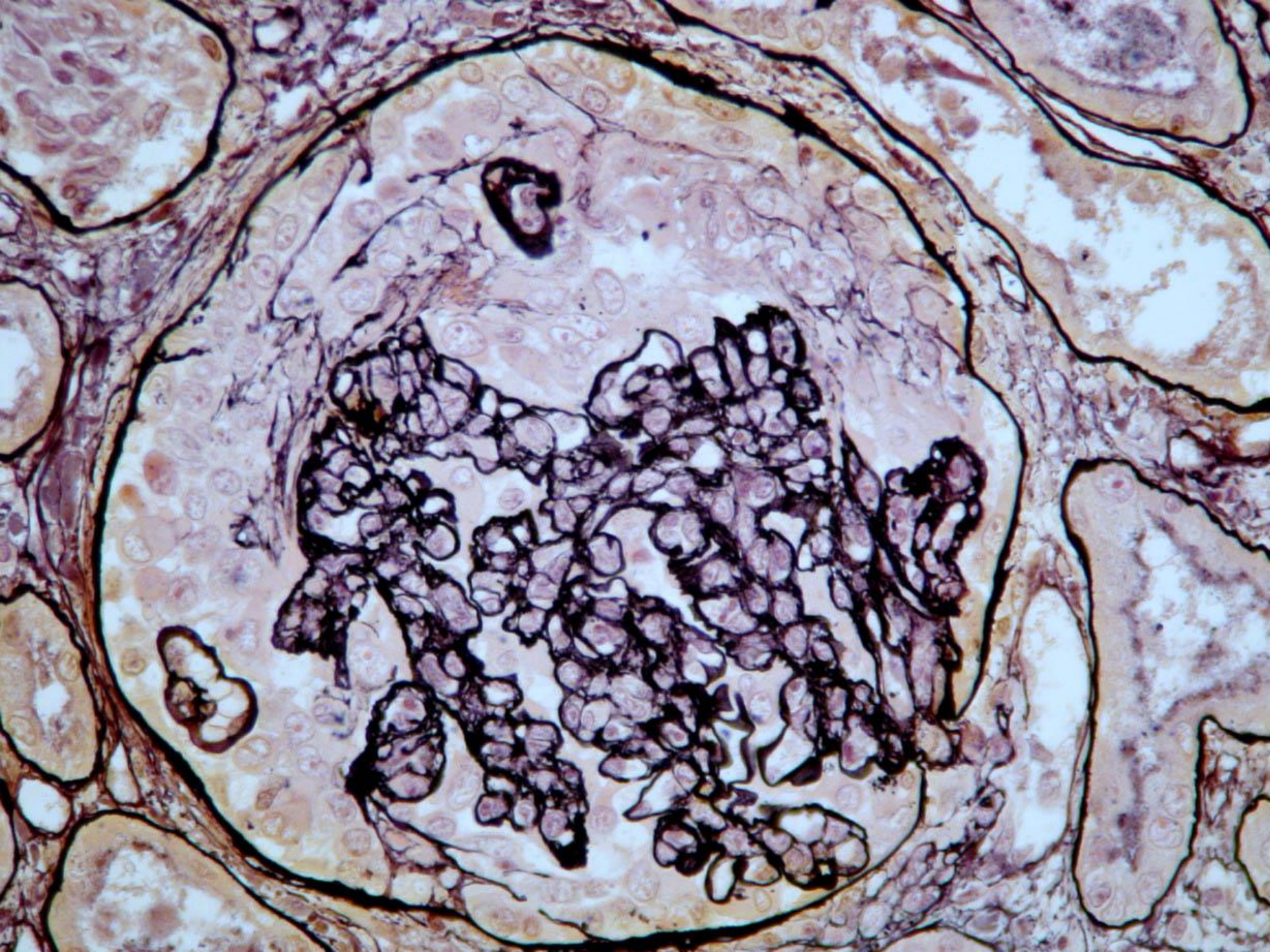


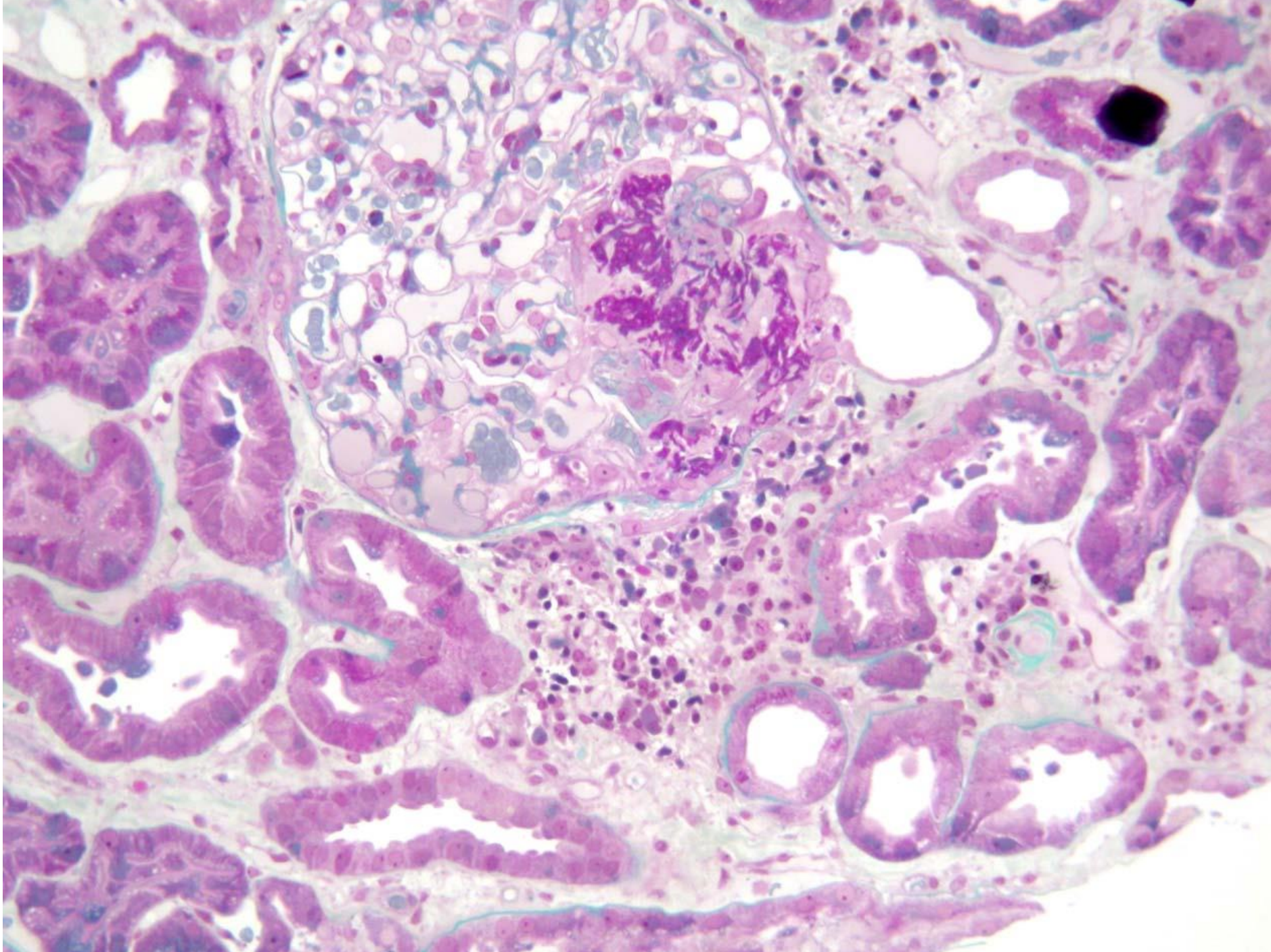


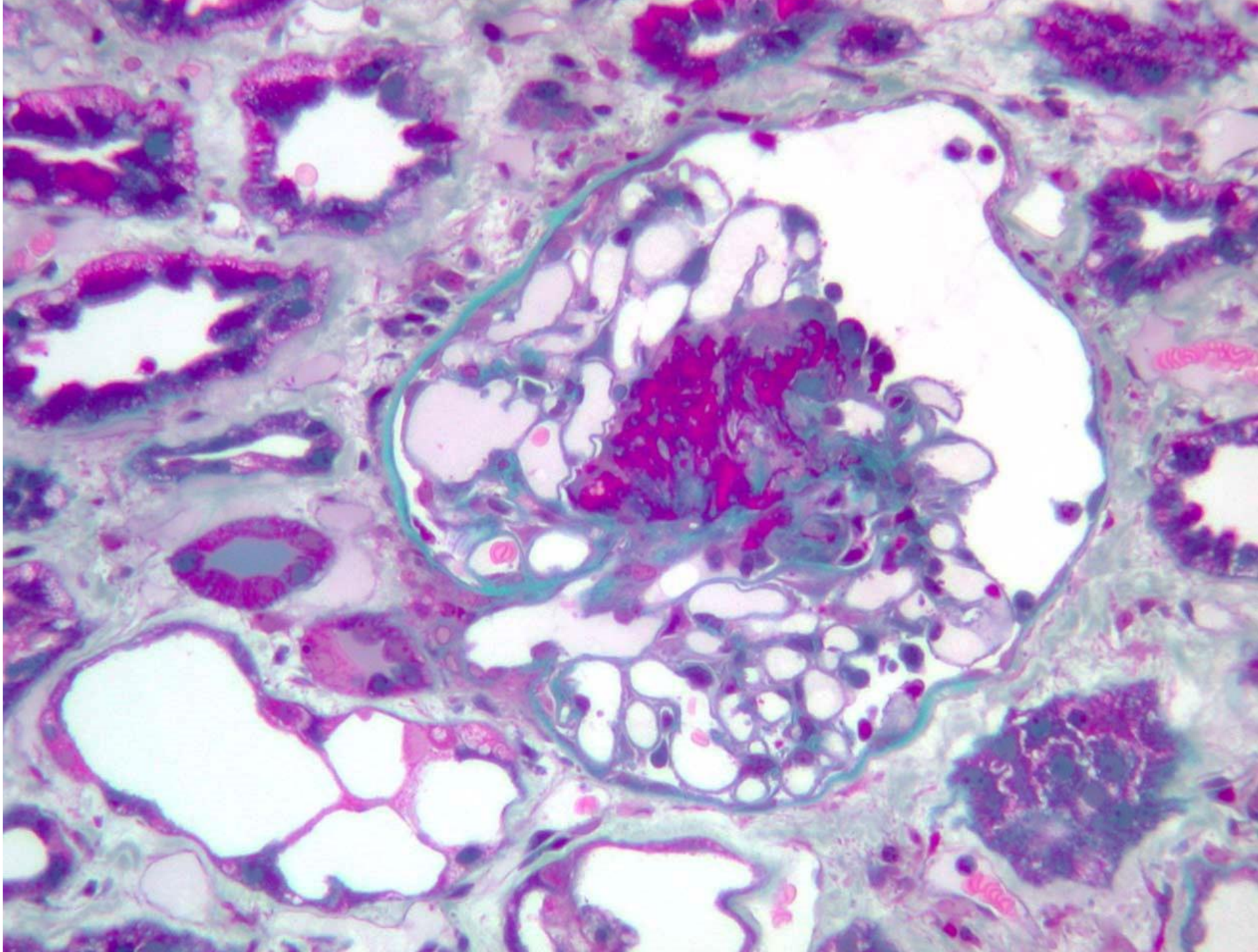


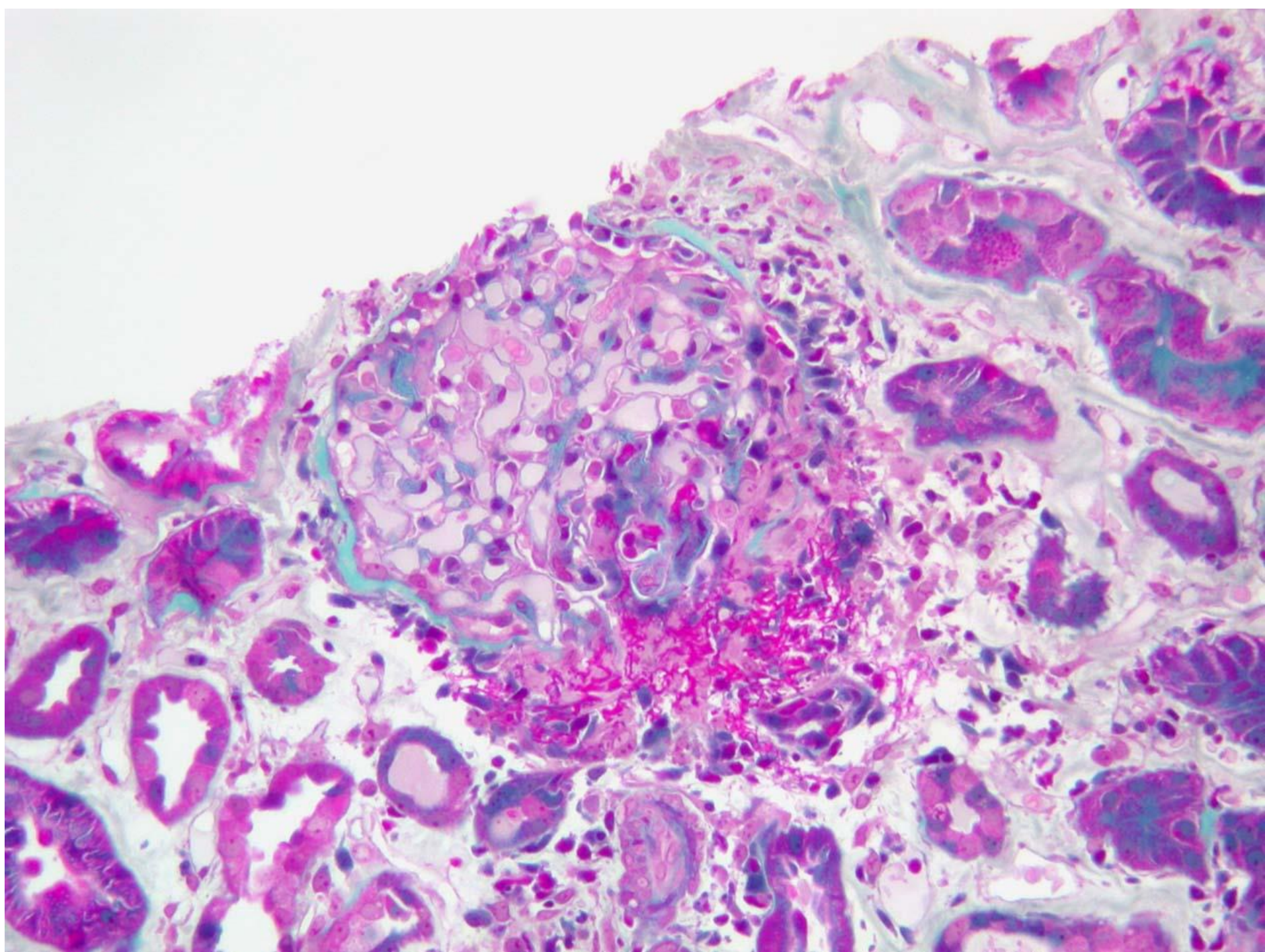
Grippe espagnole
Silice
Toxiques
Etc....

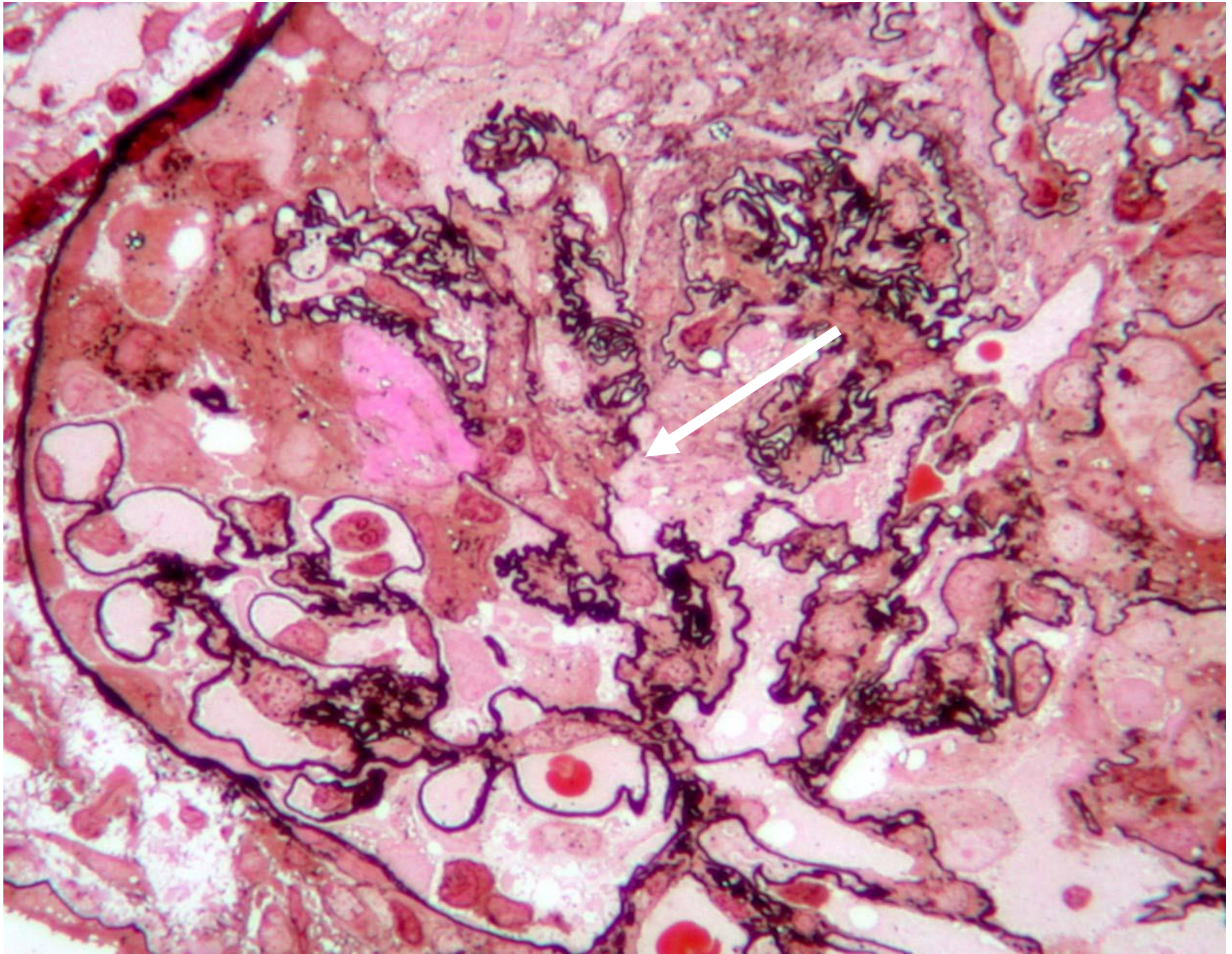


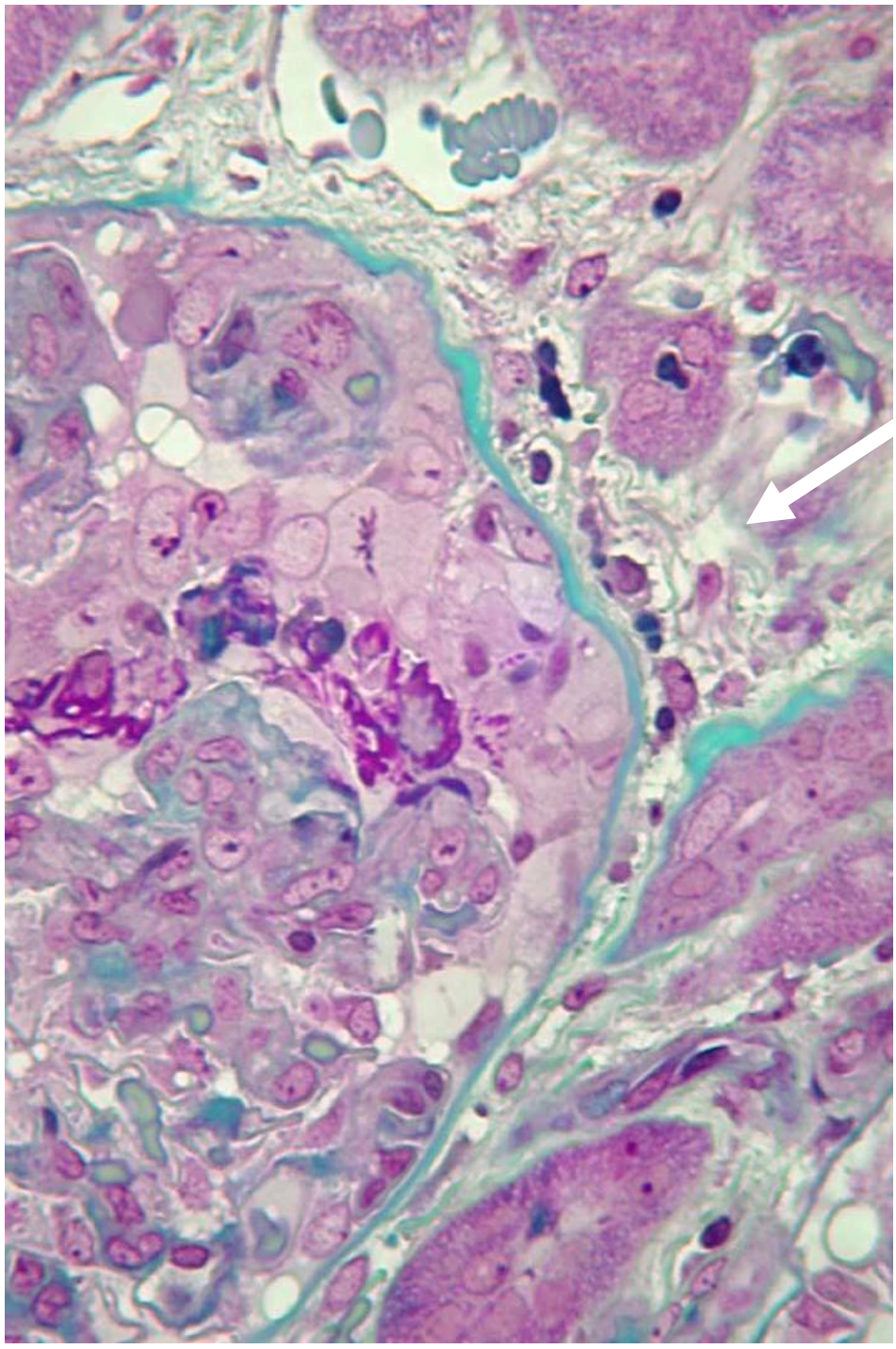
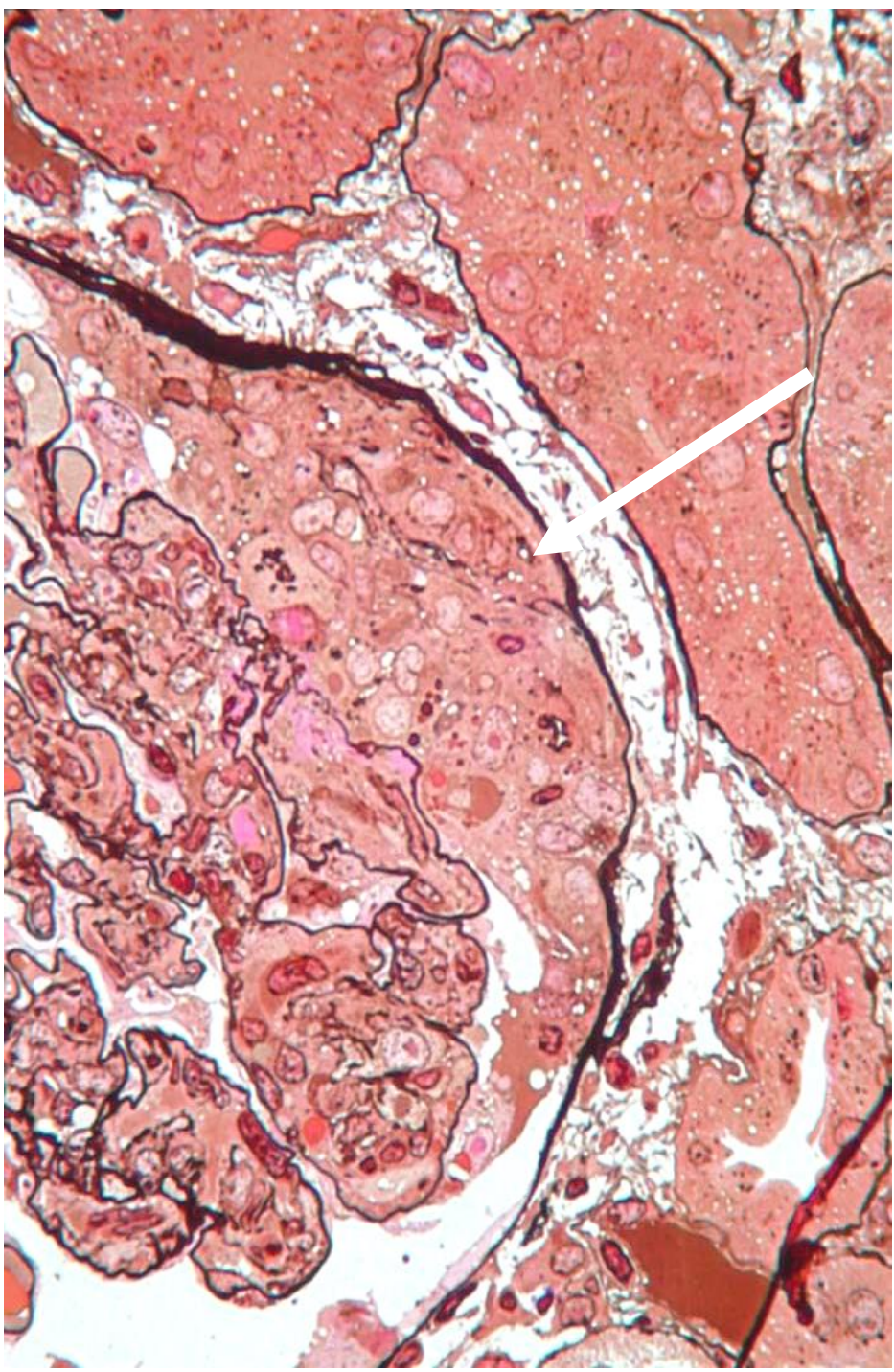


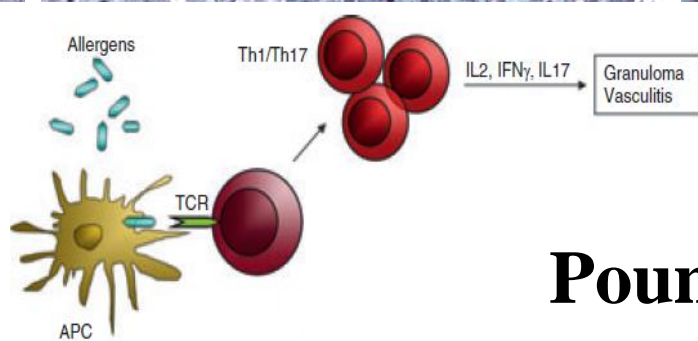
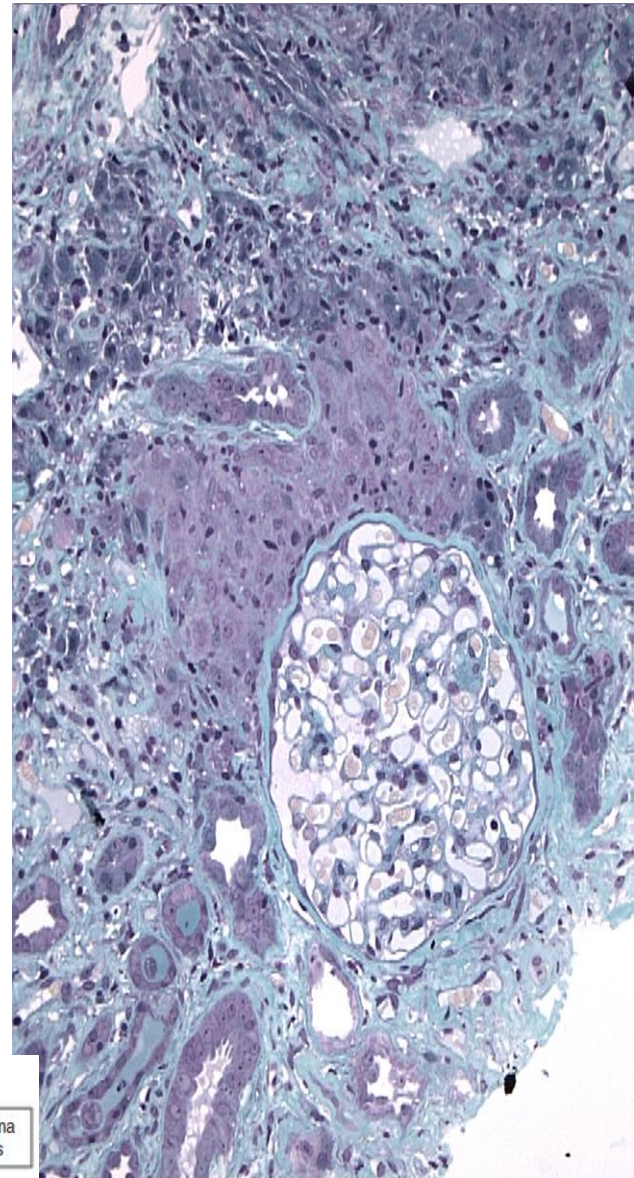
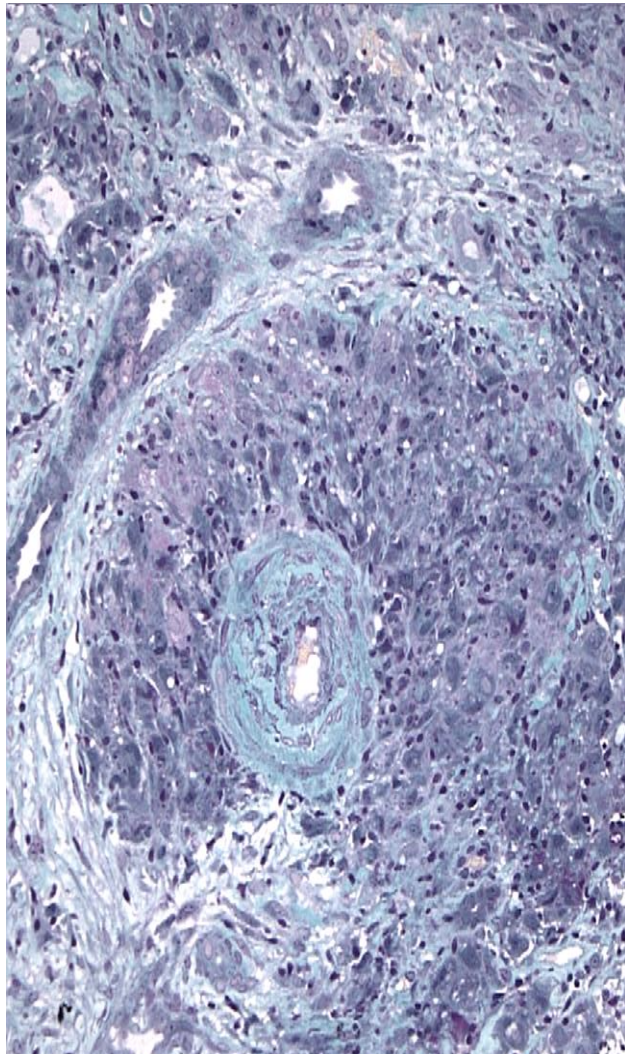
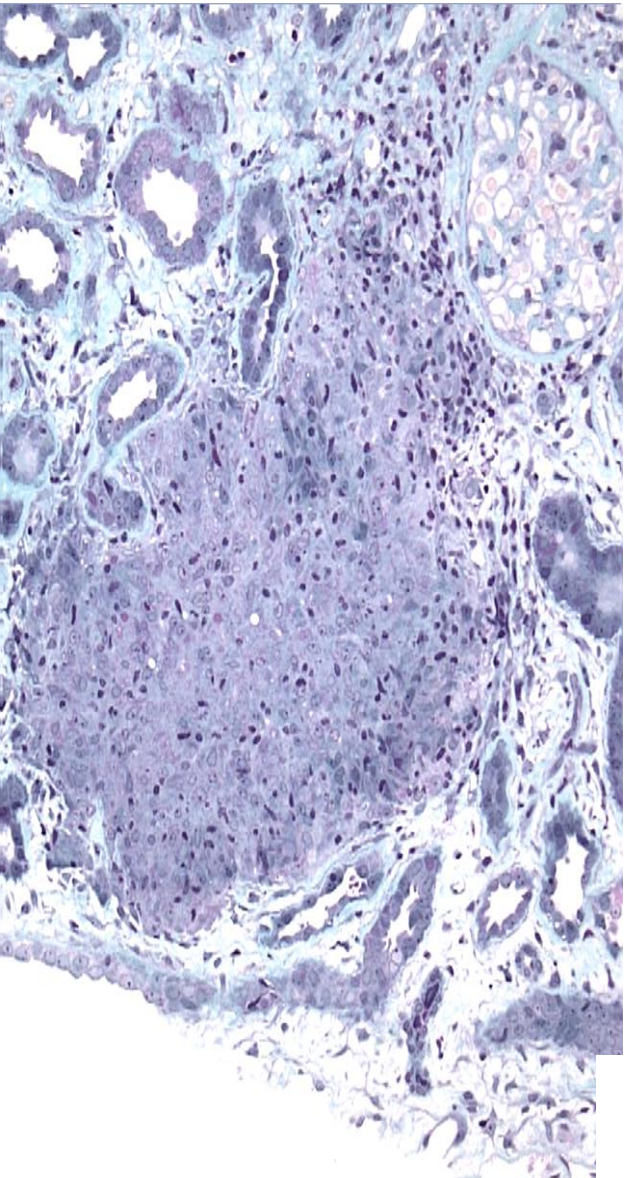




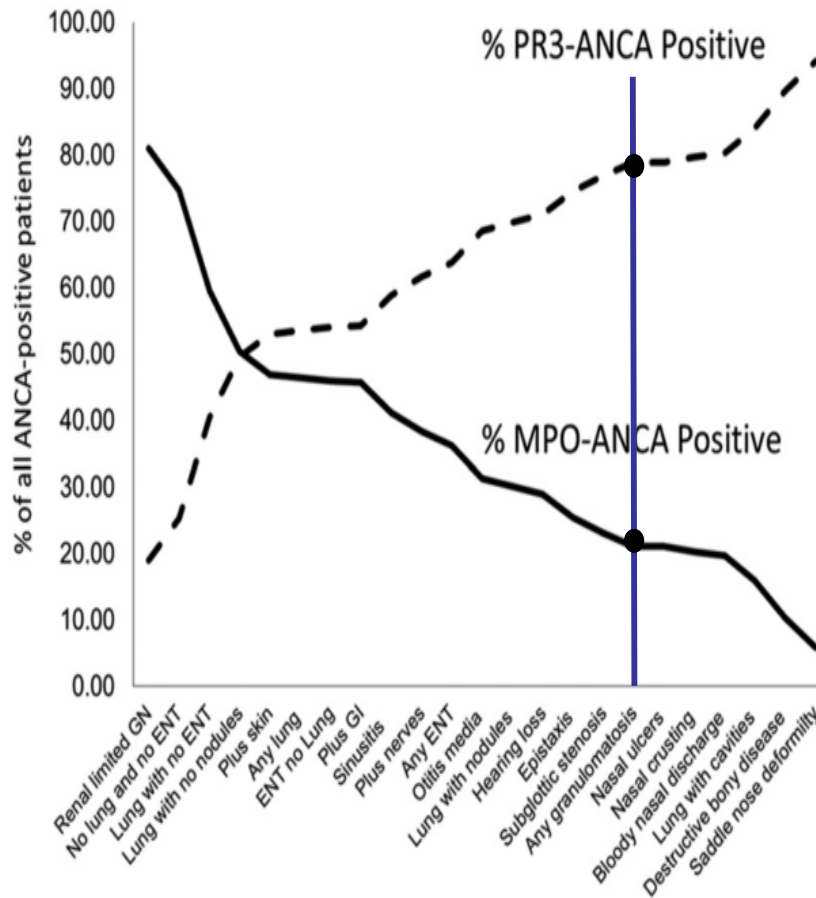








Poumons > ORL > Rein



| Lesions | MPA (N=80) | | RLV (N=19) | | WG (N=73) | | P value | | |
|----------------------------|------------|----|------------|----|-----------|----|-------------|------------|------------|
| | Mean | SD | Mean | SD | Mean | SD | MPA vs. RLV | RLV vs. WG | MPA vs. WG |
| Normal glomeruli | 21 | 22 | 27 | 25 | 40 | 34 | 0.291 | 0.137 | <0.001 |
| Fibrinoid necrosis | 21 | 25 | 21 | 26 | 23 | 26 | 0.976 | 0.656 | 0.496 |
| Crescents ^a | 47 | 28 | 49 | 33 | 42 | 32 | 0.790 | 0.365 | 0.257 |
| Cellular crescents | 45 | 28 | 55 | 27 | 45 | 29 | 0.210 | 0.205 | 0.902 |
| Fibrous crescents | 7 | 10 | 3 | 6 | 4 | 9 | 0.239 | 0.715 | 0.172 |
| Glomerulosclerosis | 30 | 29 | 23 | 25 | 16 | 23 | 0.361 | 0.265 | 0.002 |
| Periglomerular infiltrates | 10 | 16 | 16 | 20 | 9 | 12 | 0.121 | 0.033 | 0.680 |
| Granulomatous reaction | 4 | 13 | 2 | 6 | 2 | 8 | 0.566 | 0.918 | 0.379 |

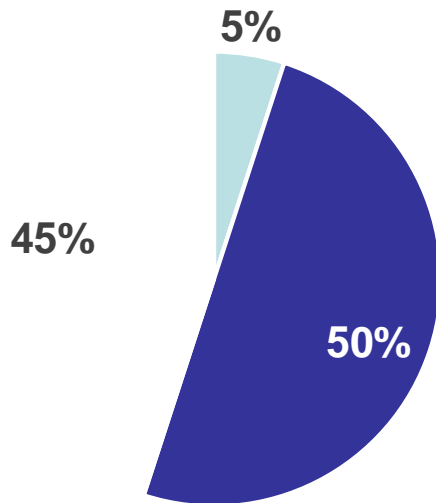
^aThe mean percentage of cellular crescents and fibrous crescents together is usually different from the mean percentage of crescents, as the former two lesions were not scored if crescents were absent

Glomérulopathie et Anti-MBG

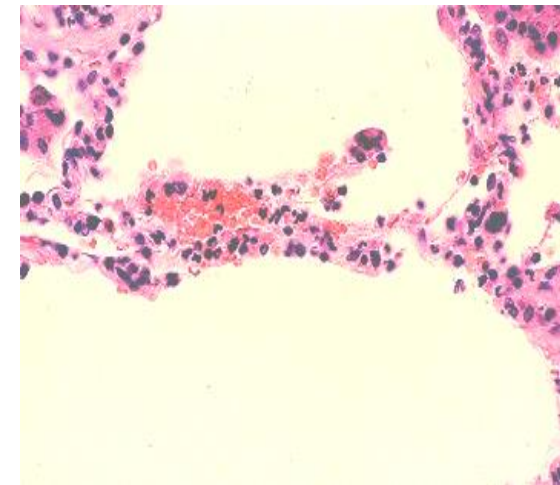
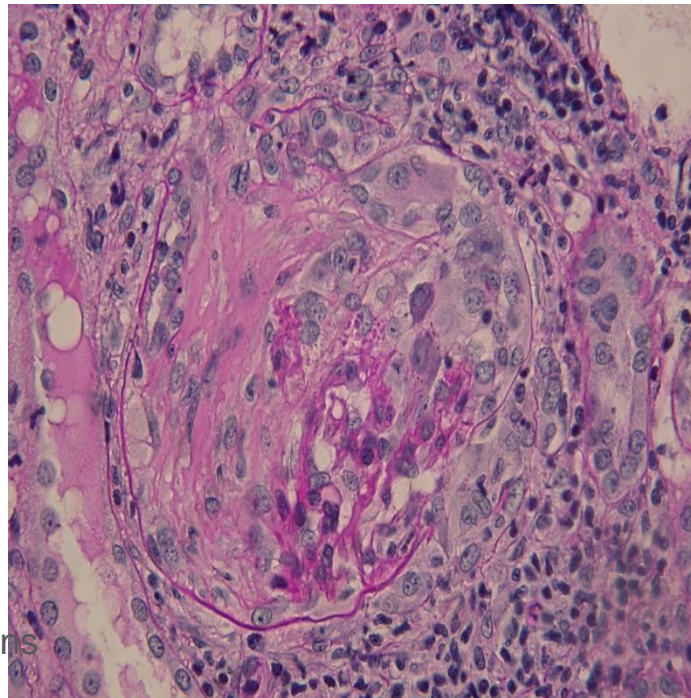
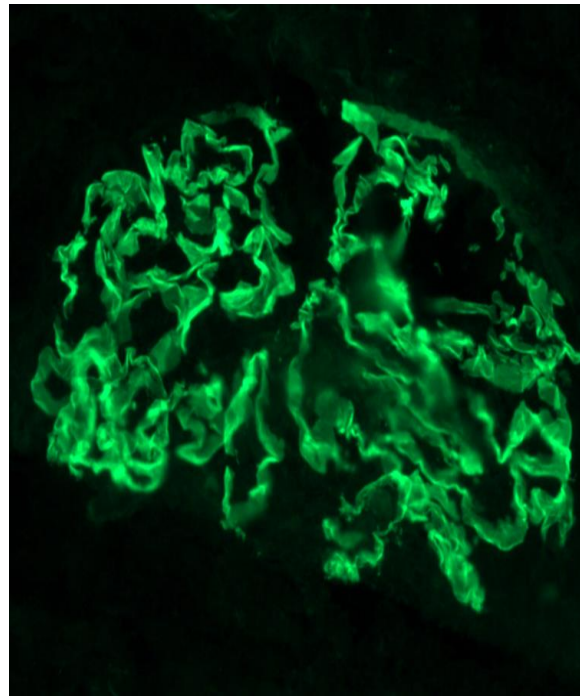


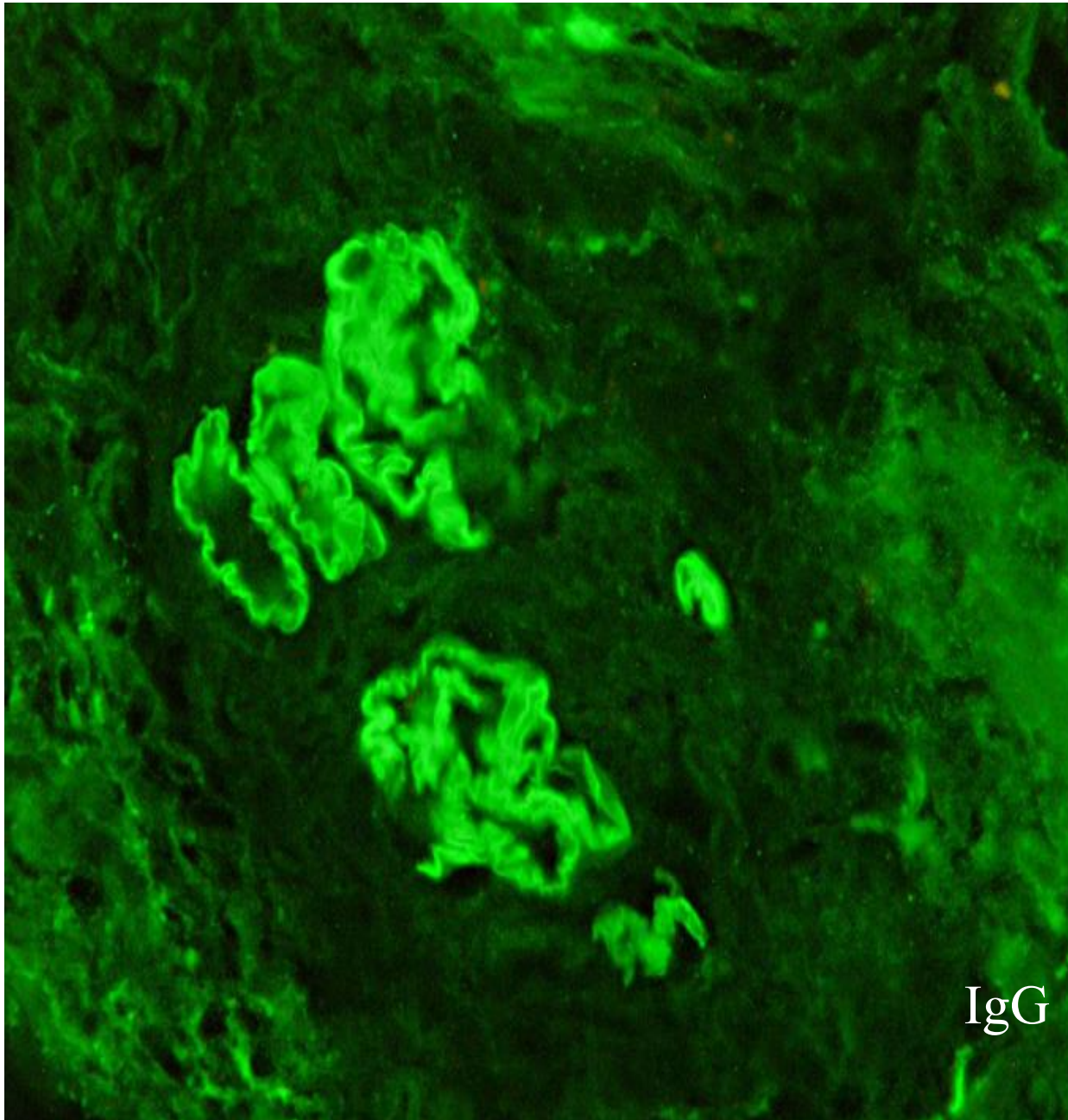
Ernest Goodpasture

Présentations



Poumons ■ Reins ■ Poumons + Reins

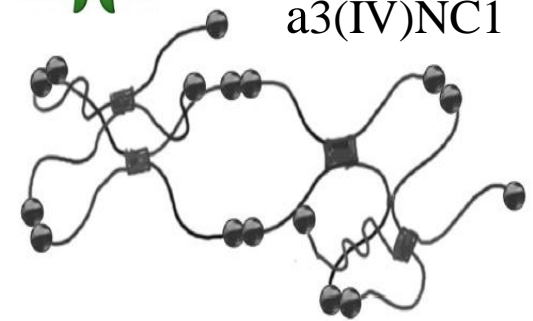




IgG



a3(IV)NC1



HLA-DRB1*1501 and DRB1*1502

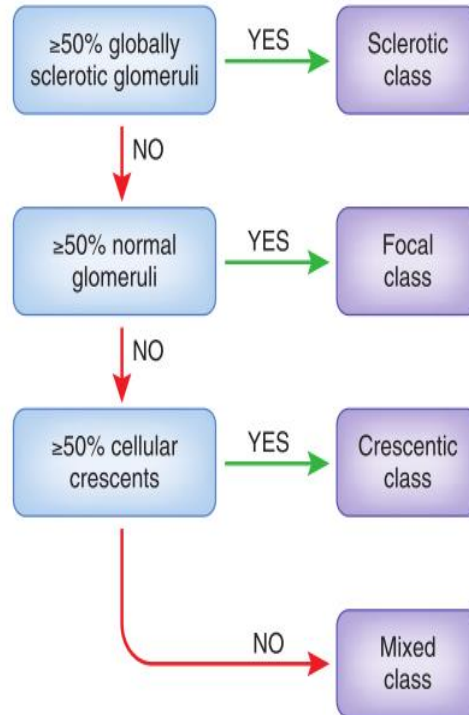
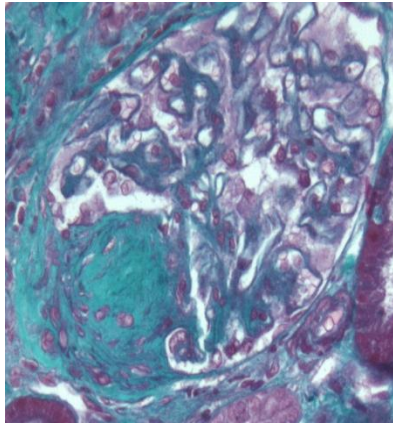
Environnement:

- Virus respiratoires (I. A2).
- Solvants et produits caustiques (Decapfour);
- Cocaïne.
- Alemtuzumab (anti-CD52).

Table 2. Classification schema for ANCA-associated glomerulonephritis

| Class | Inclusion Criteria ^a |
|------------|---|
| Focal | ≥50% normal glomeruli |
| Crescentic | ≥50% glomeruli with cellular crescents |
| Mixed | <50% normal, <50% crescentic, <50% globally sclerotic glomeruli |
| Sclerotic | ≥50% globally sclerotic glomeruli |

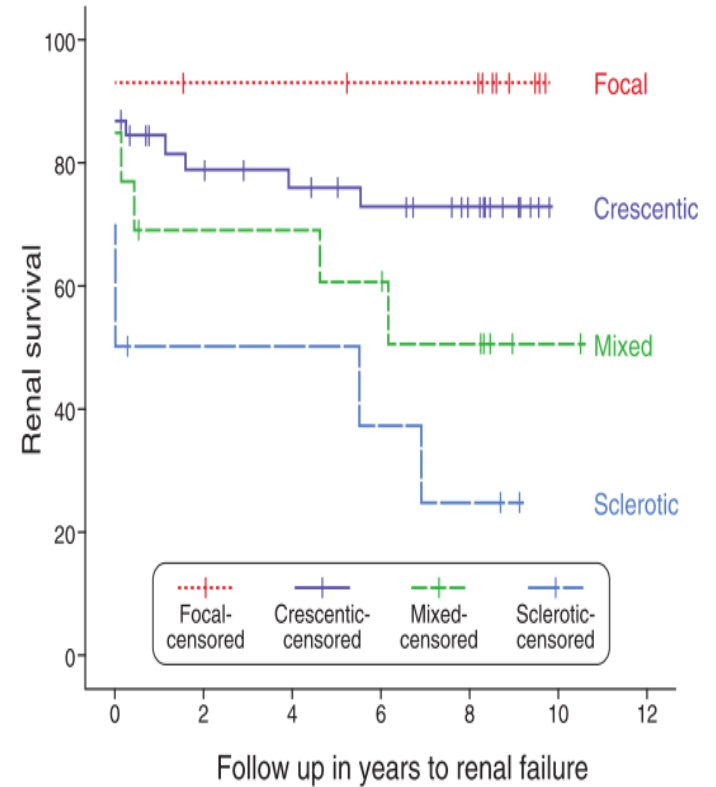
^aPauci-immune staining pattern on immunofluorescence microscopy (IM) and ≥1 glomerulus with necrotizing or crescentic glomerulonephritis on light microscopy (LM) are required for inclusion in all four classes. See Figure 1 for hierarchical structure.



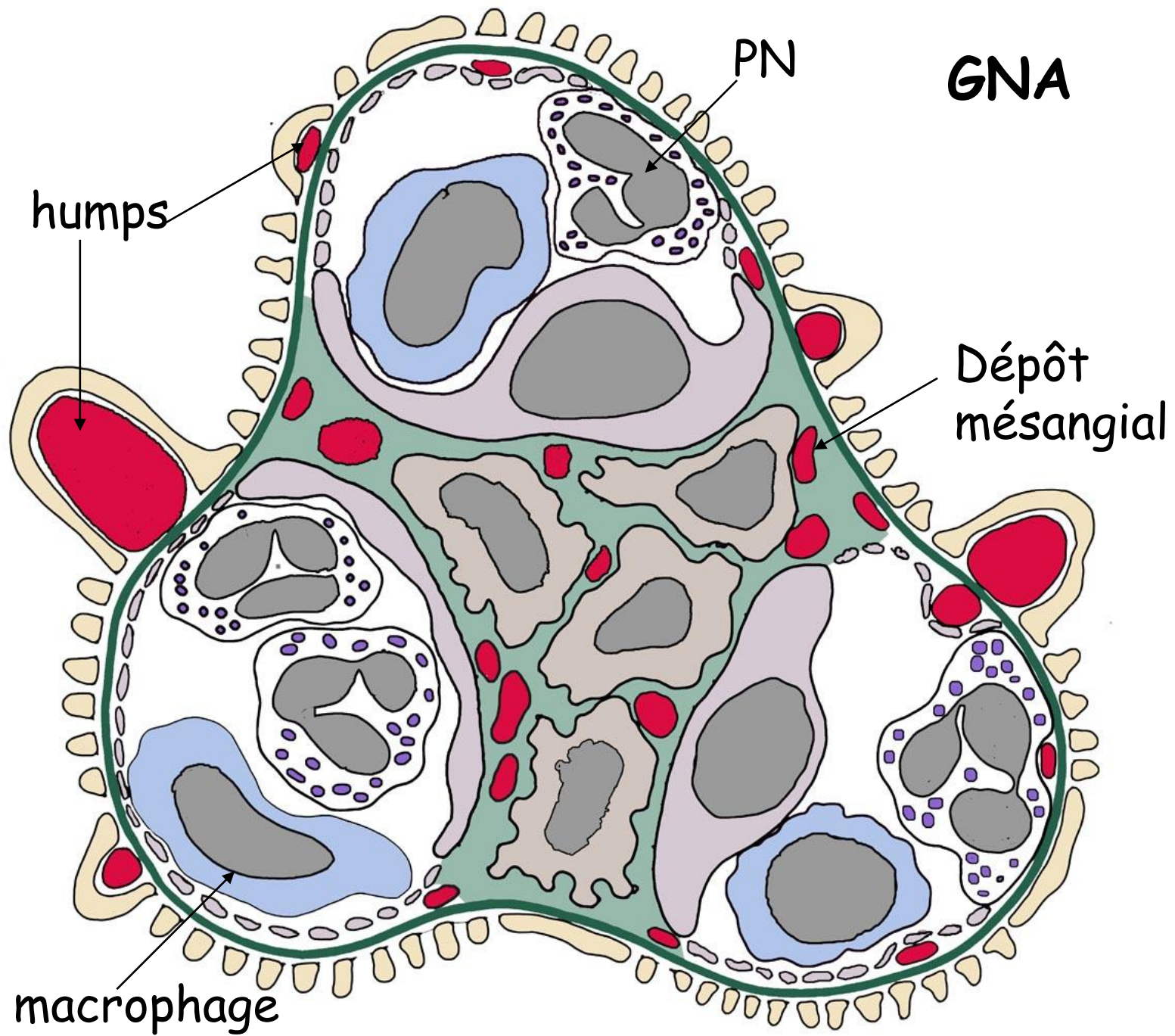
ATTENTION!

Applicable surtout si DFG > 15 ml/min (Lee T et al, cJASN 2014)

Berden AE, JASN 2010.



Les glomérulopathies aigues post-infectieuses



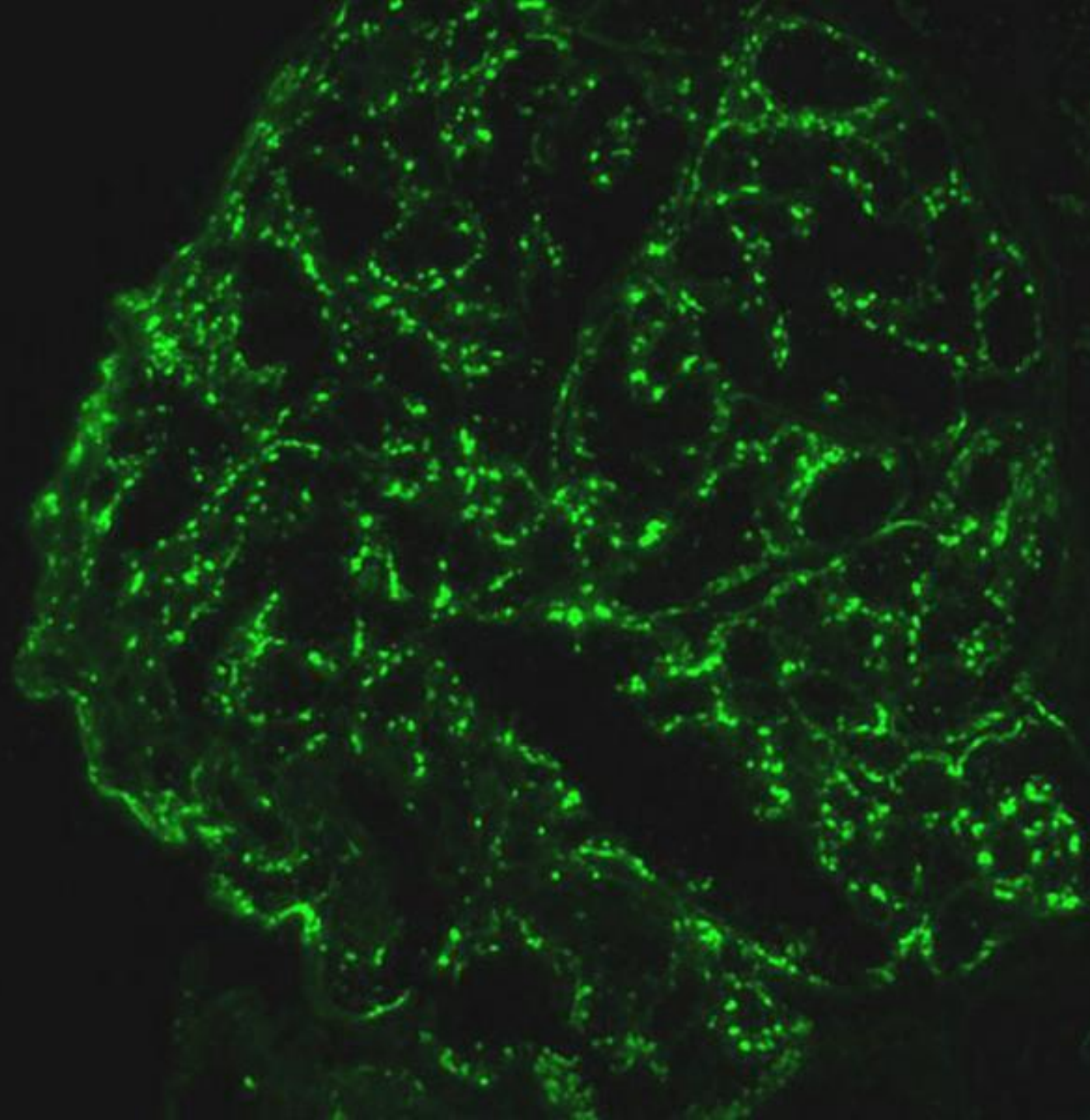
humps

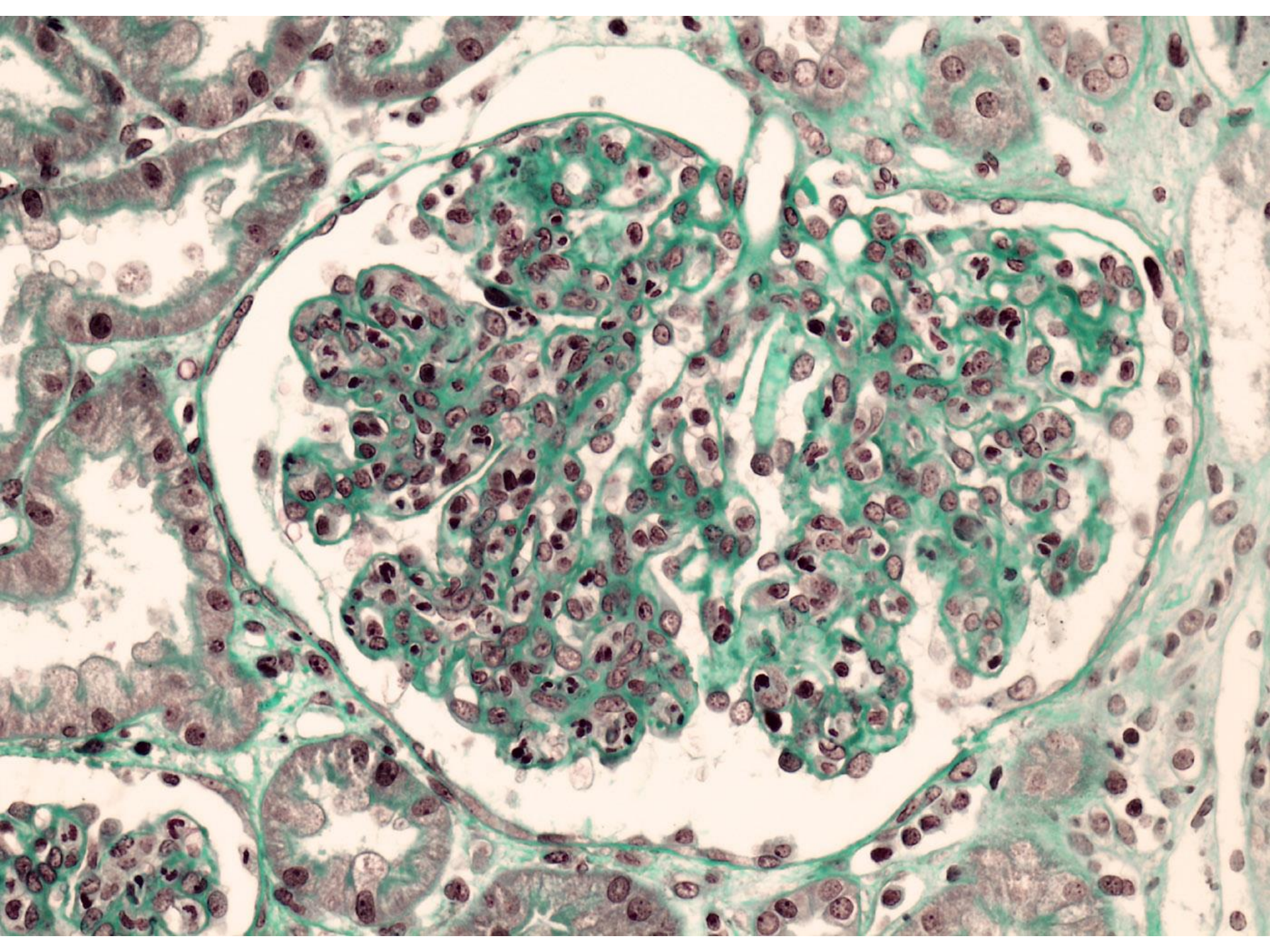
PN

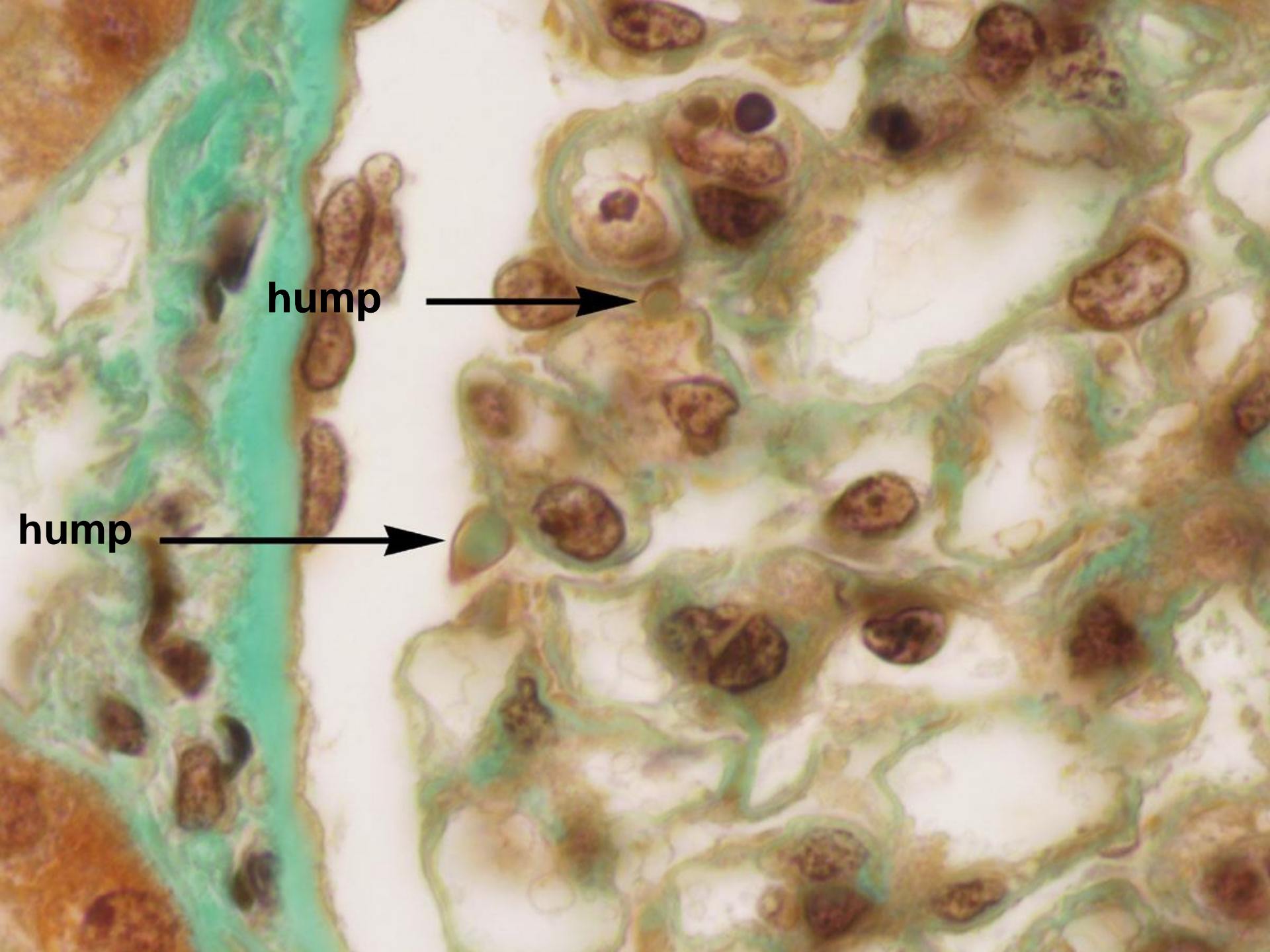
GNA

Dépôt
mésangial

macrophage



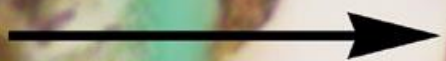


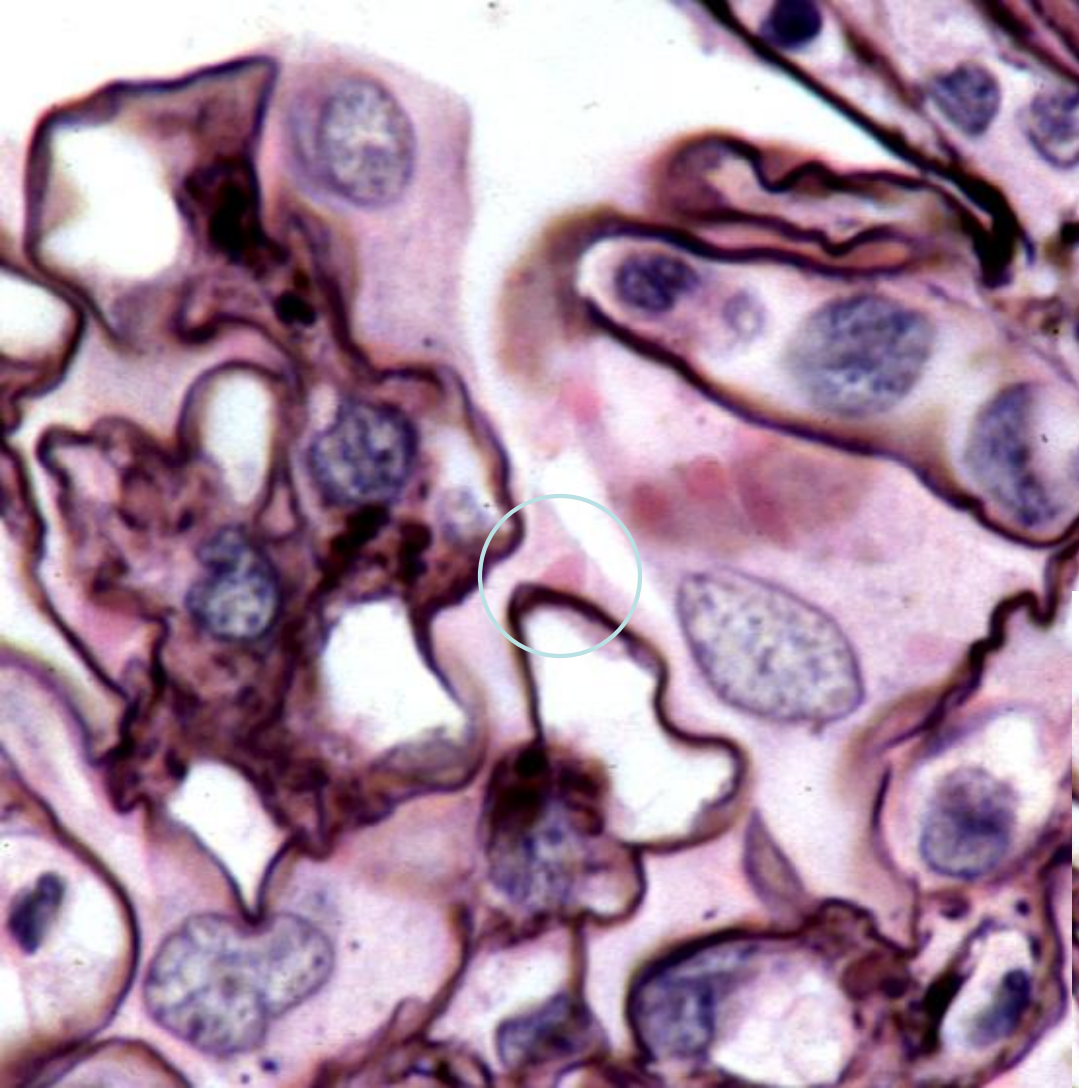


hump



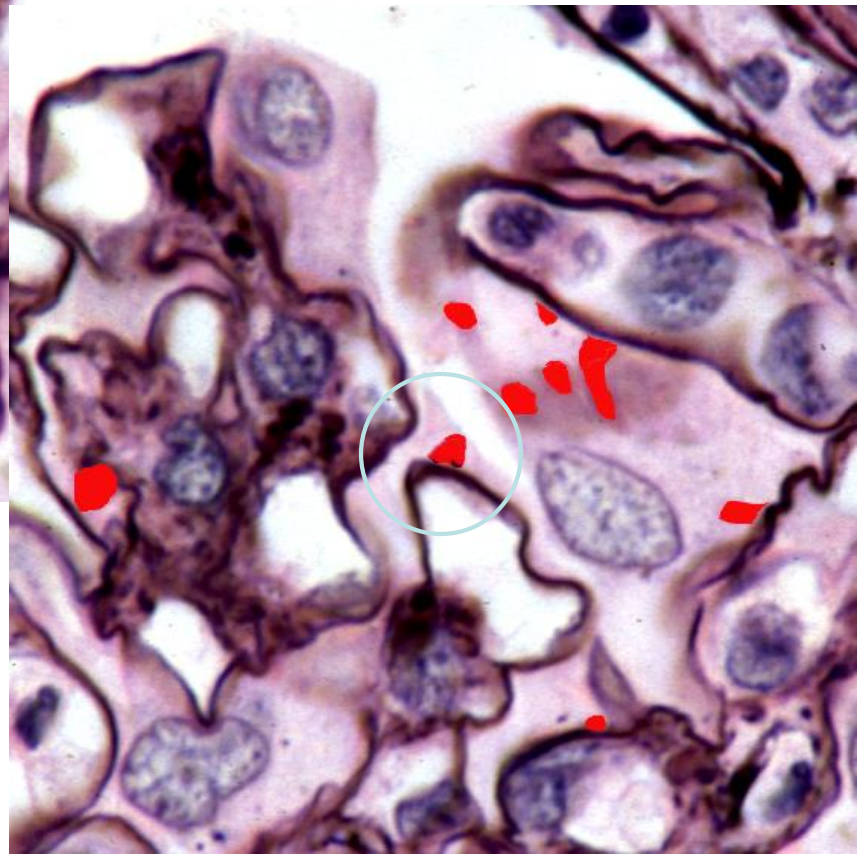
hump





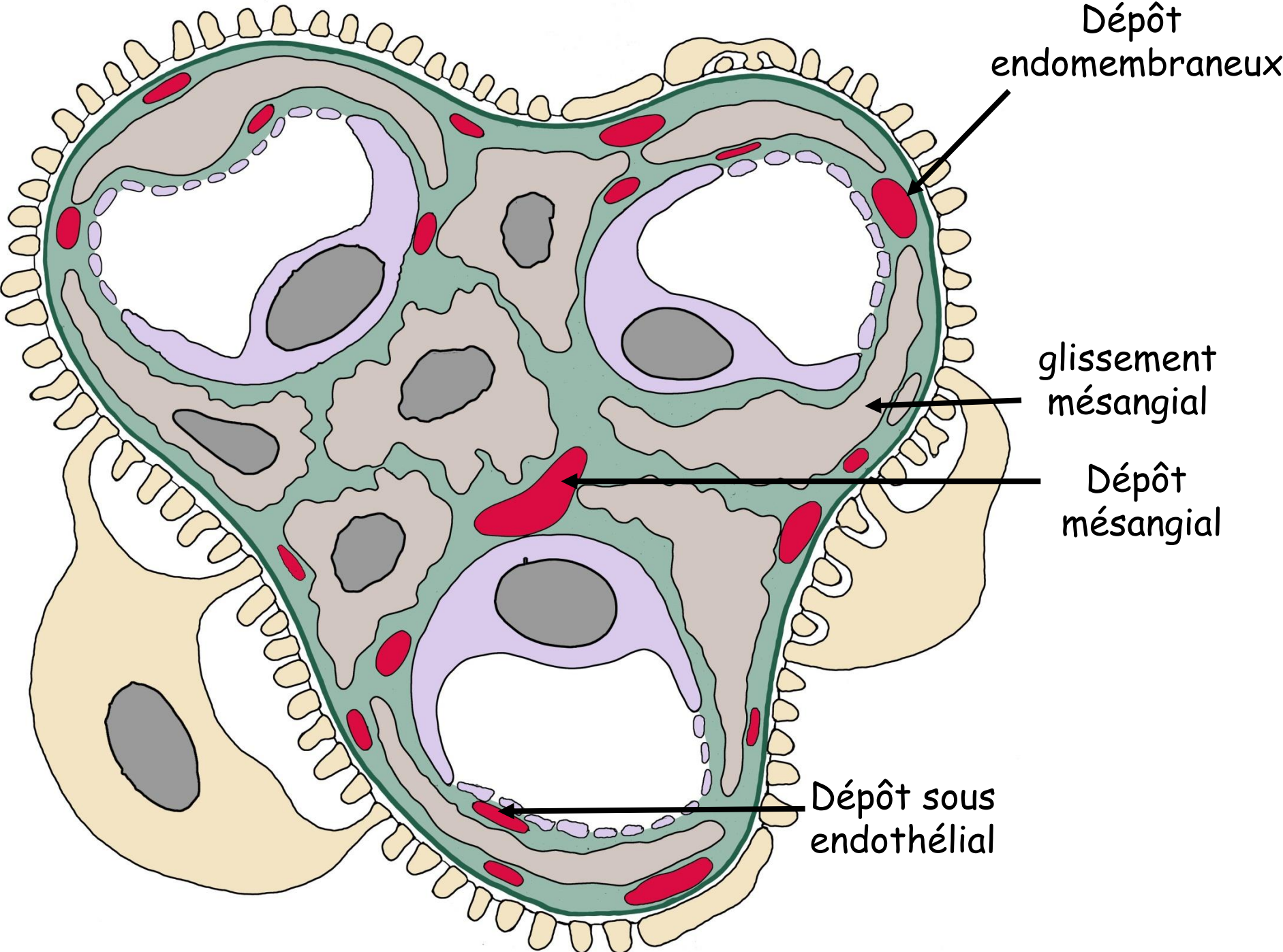
Humps dans une GNA

Argentation + H&E



Les humps ont été soulignés

**Les glomérulopathies
proliférantes membrano-
prolifératives**



Dépôt endomembraneux

glissement mésangial

Dépôt mésangial

Dépôt sous endothélial

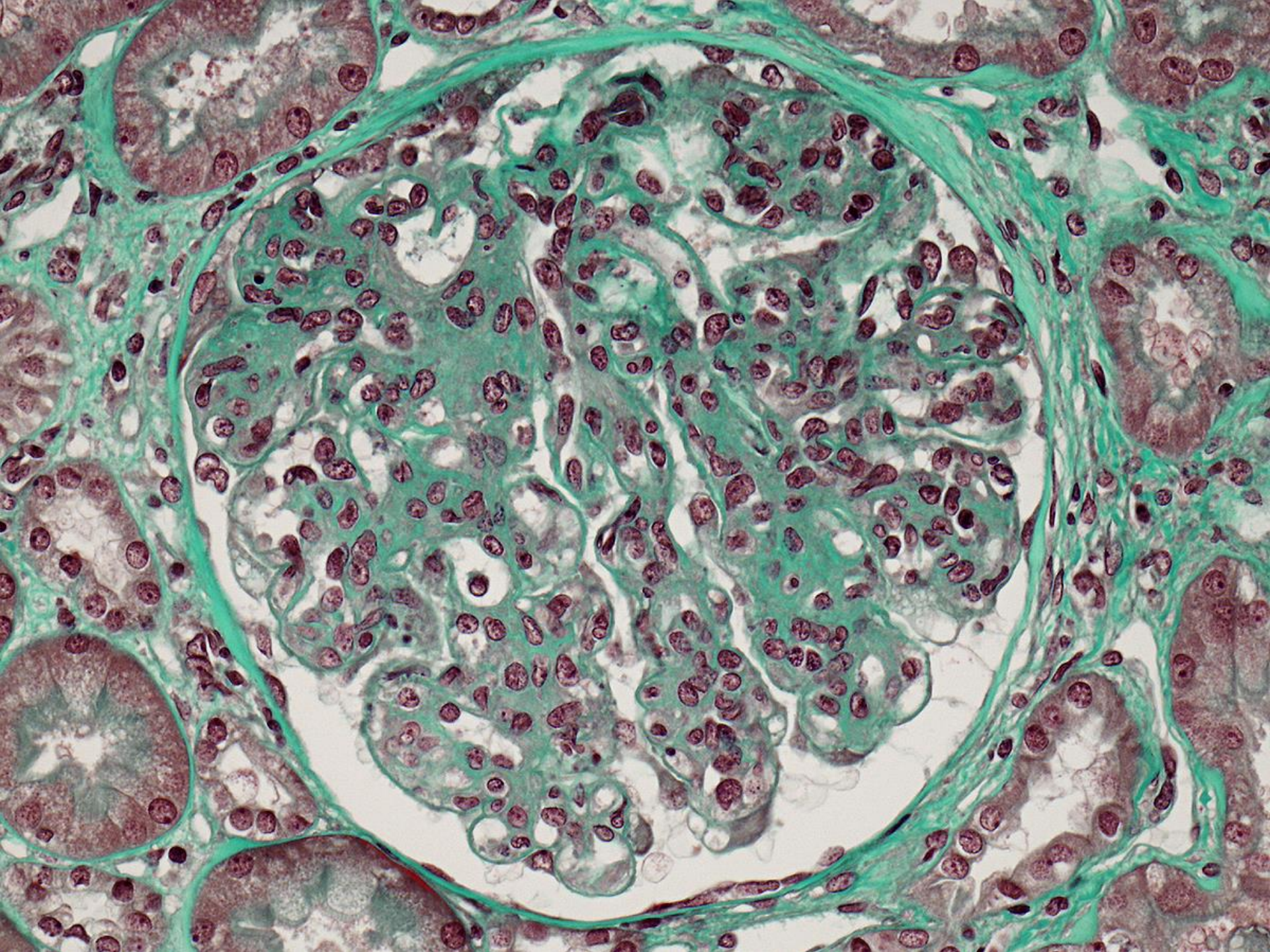
GN membrano prolifératives

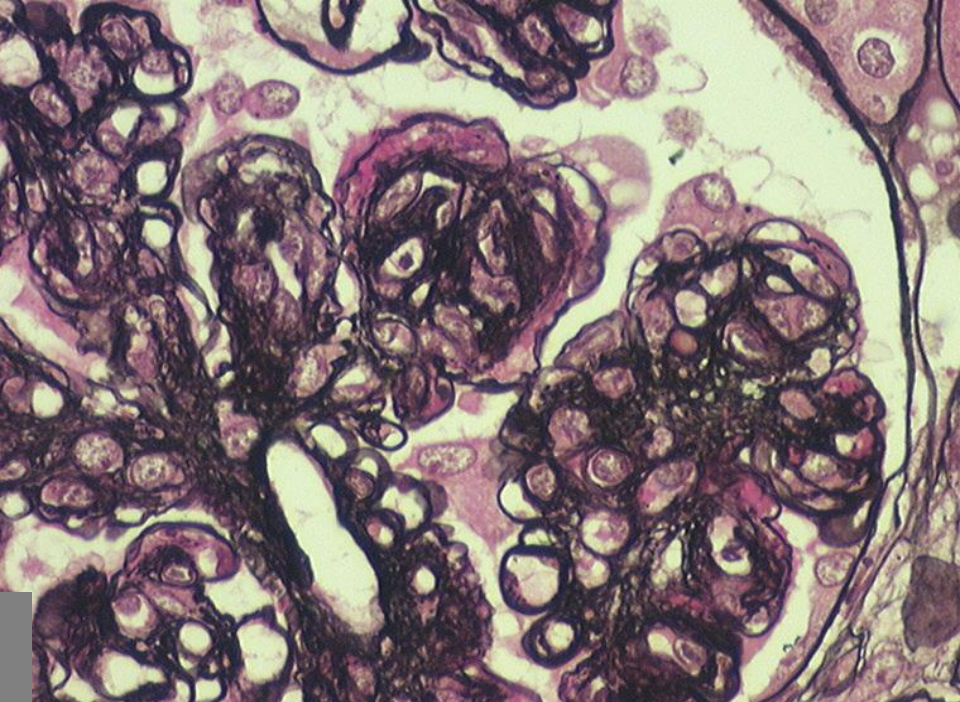
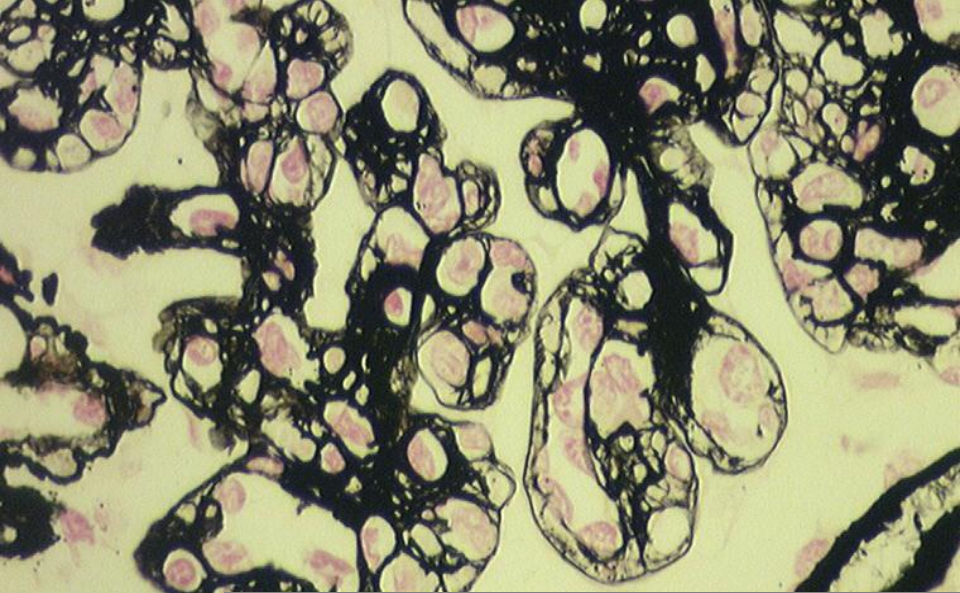
**GN membrano-prolifératives de type I
(à dépôts sous endothéliaux-endo)**

**GN membrano-prolifératives de type II
(à dépôts denses dans les basales, intra-mb)**

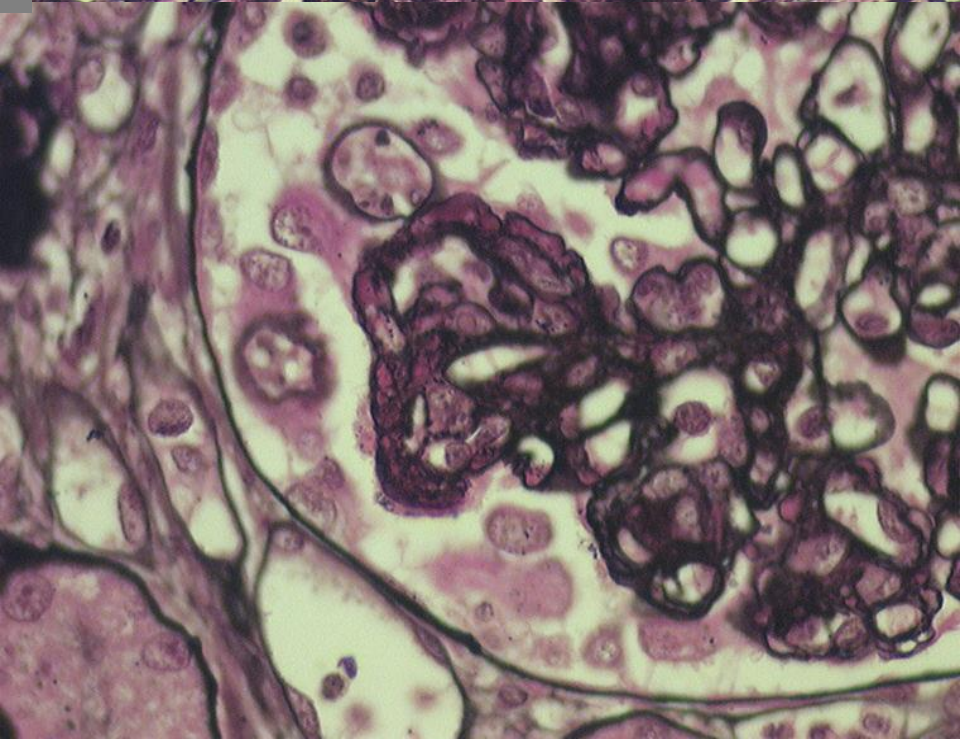
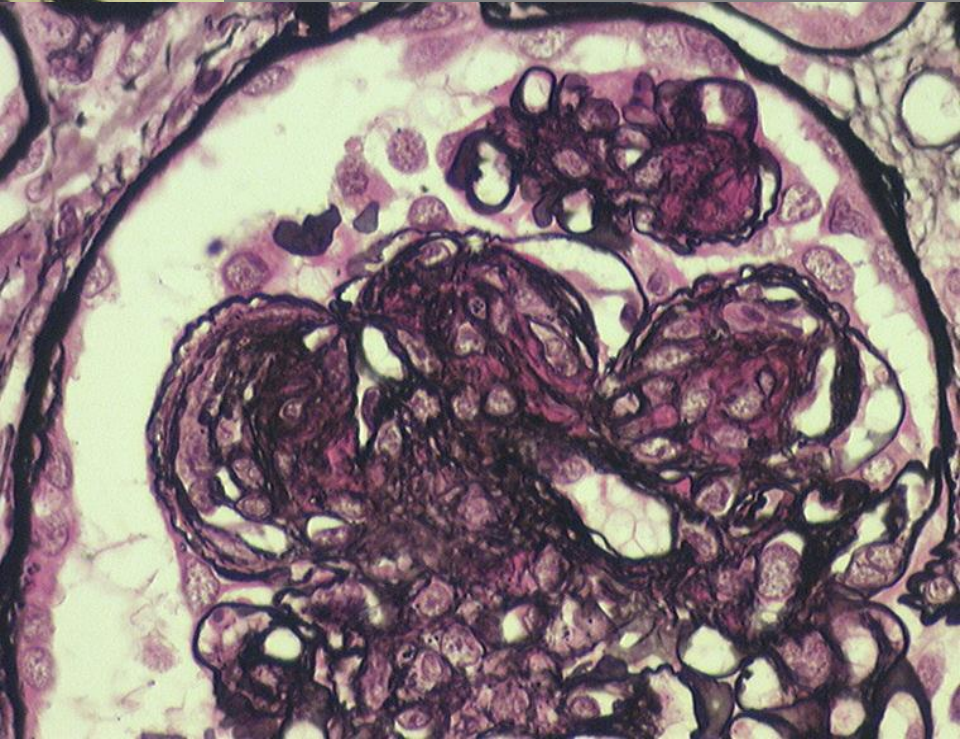
**GN membrano-prolifératives à dépôts isolés de C3
(type III, endo-extra)**

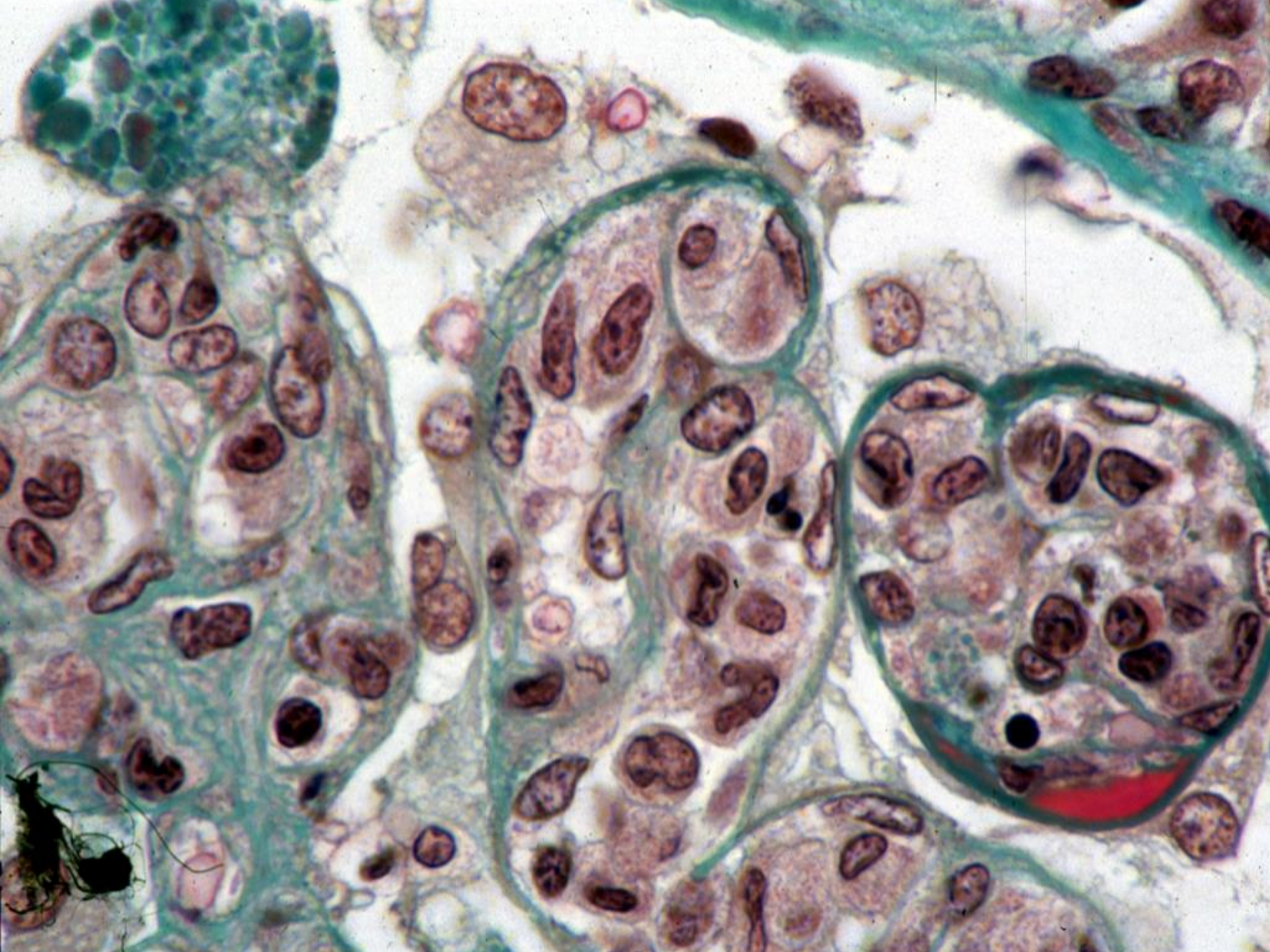
GN membrano proliferatives de type 1



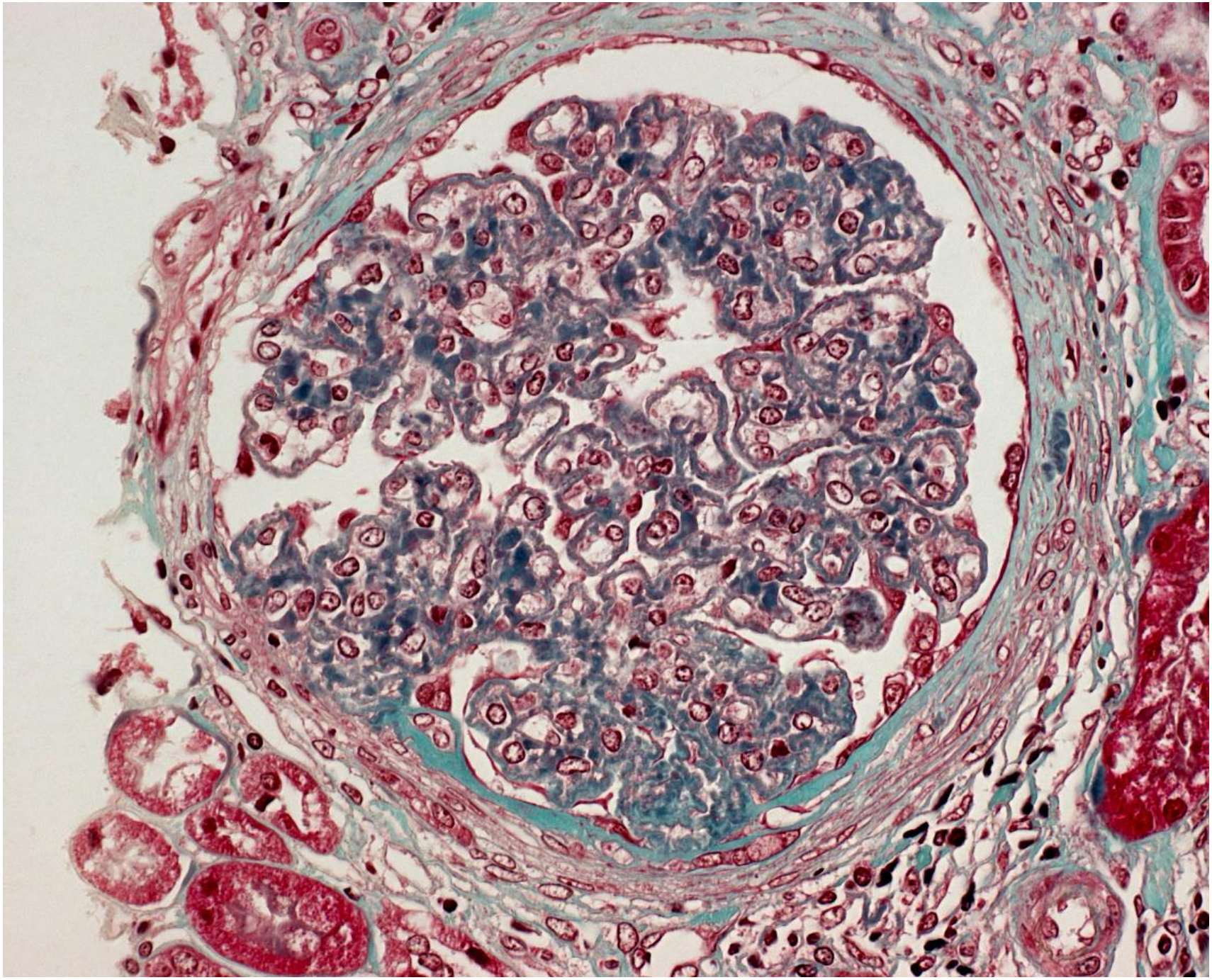


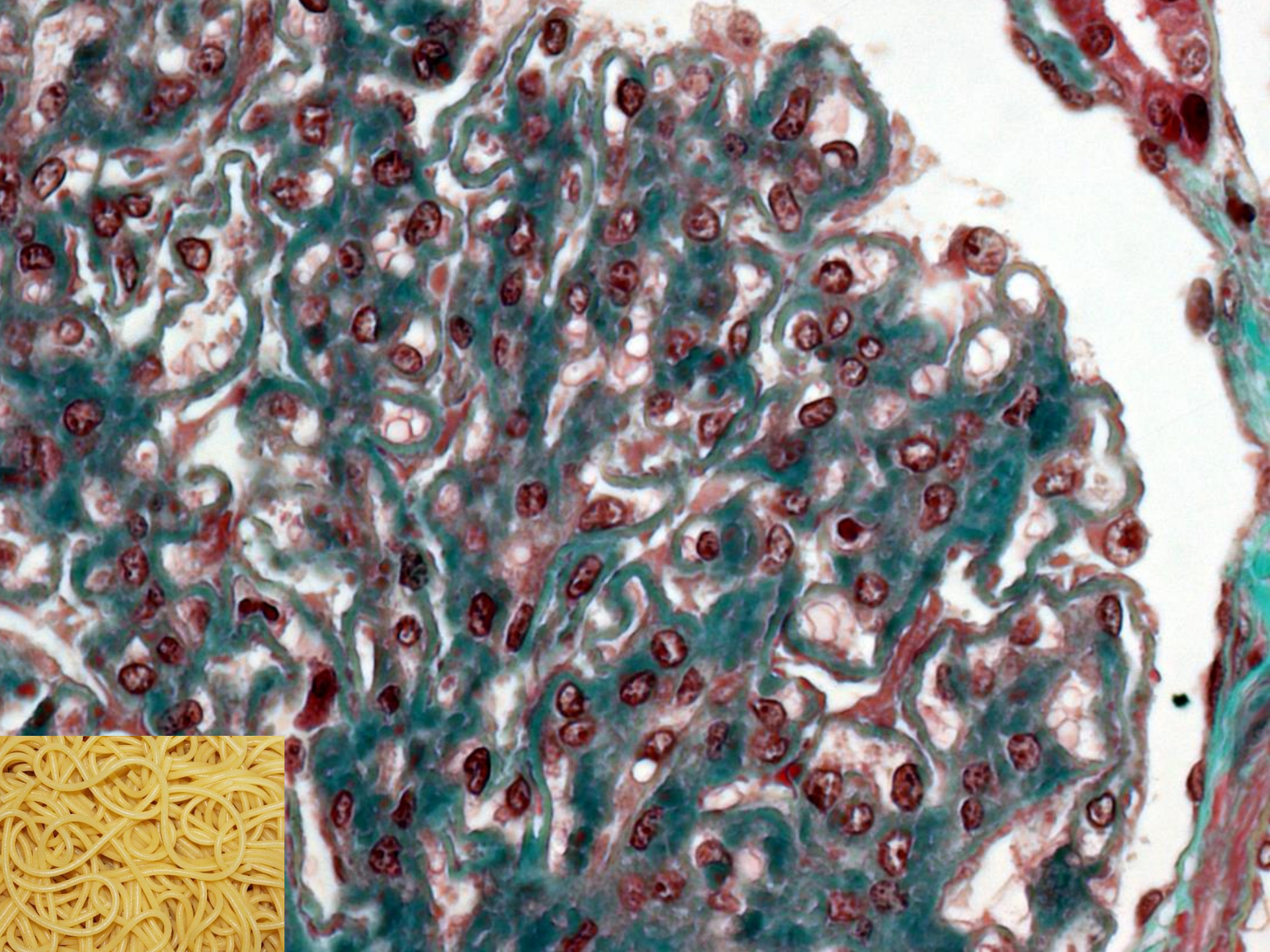
Double contours





**GN membrano-prolifératives
de type 2 (dépôts denses)**

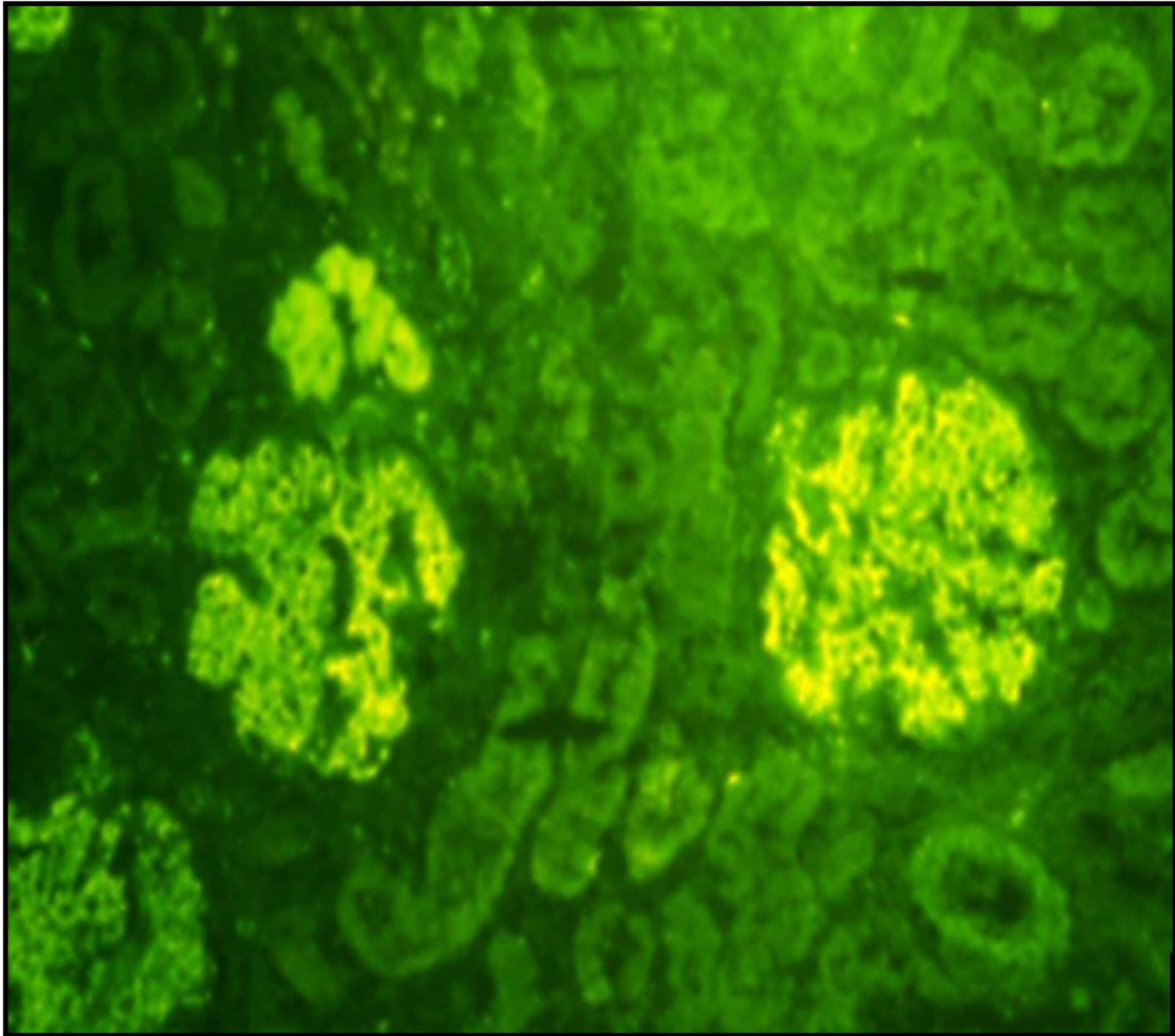




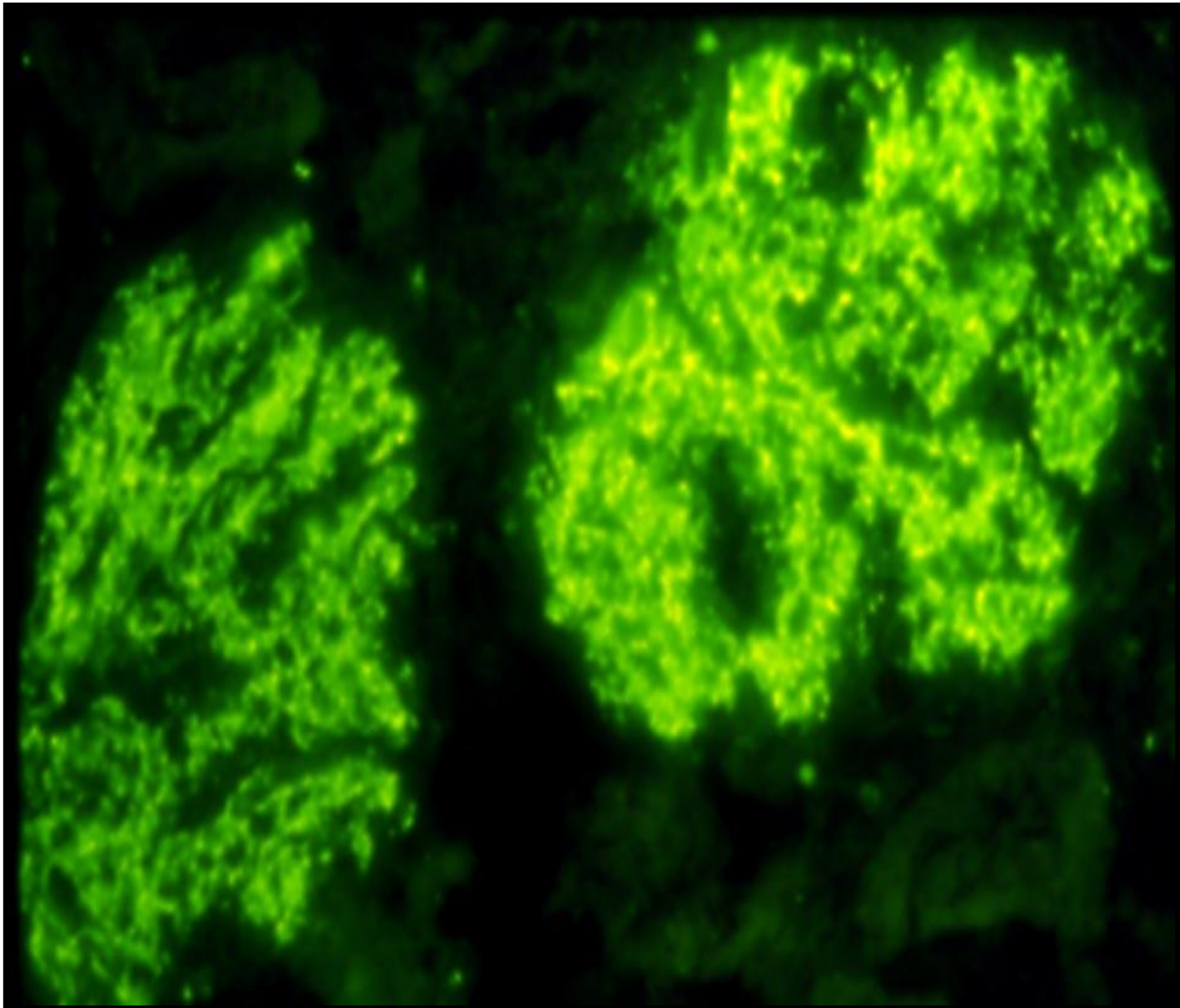
GN membrano-prolifératives de type 3 (dépôts mixtes)

**Rôle du complément (GN à
C3).....**

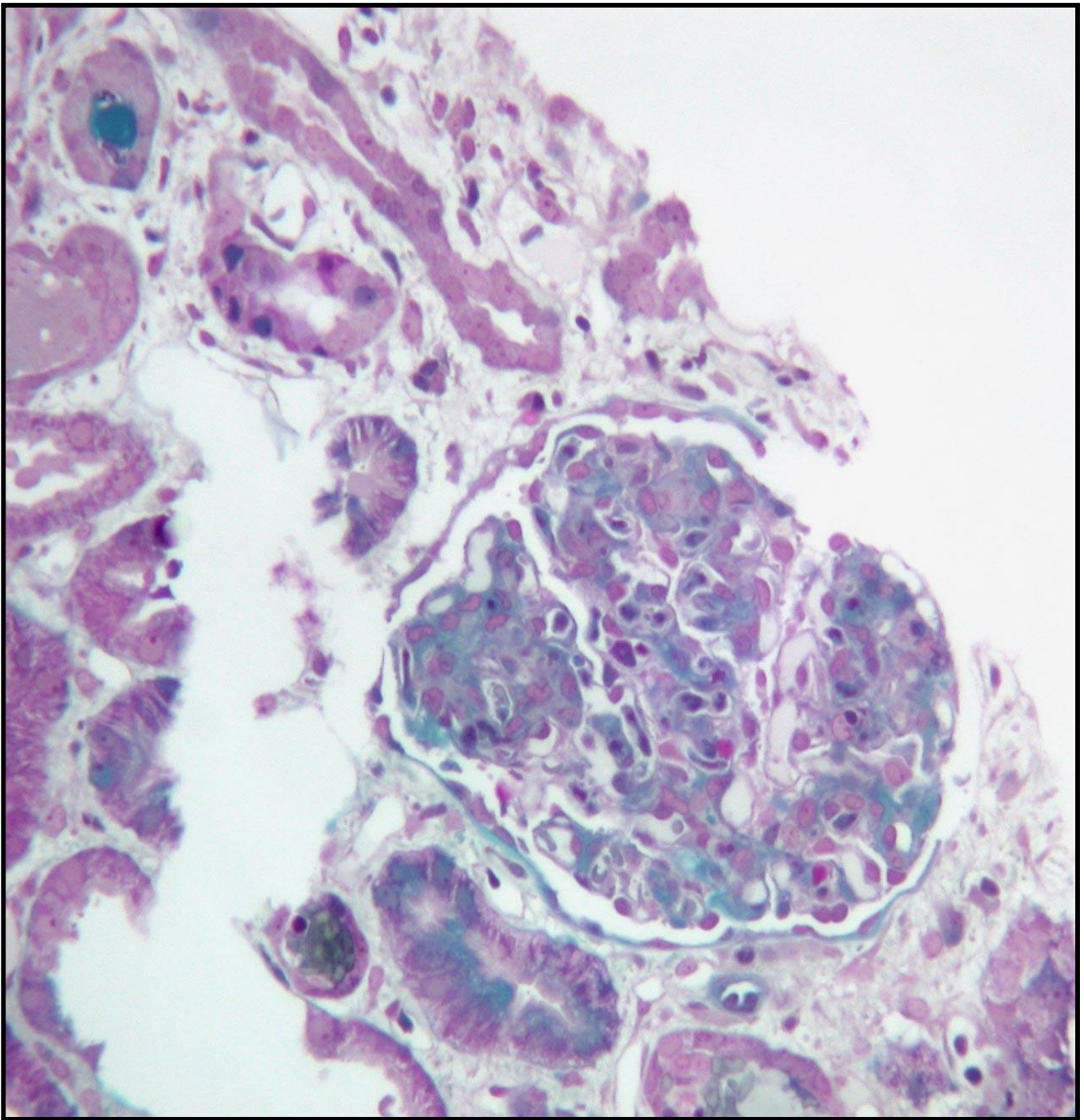
B. Nicolas

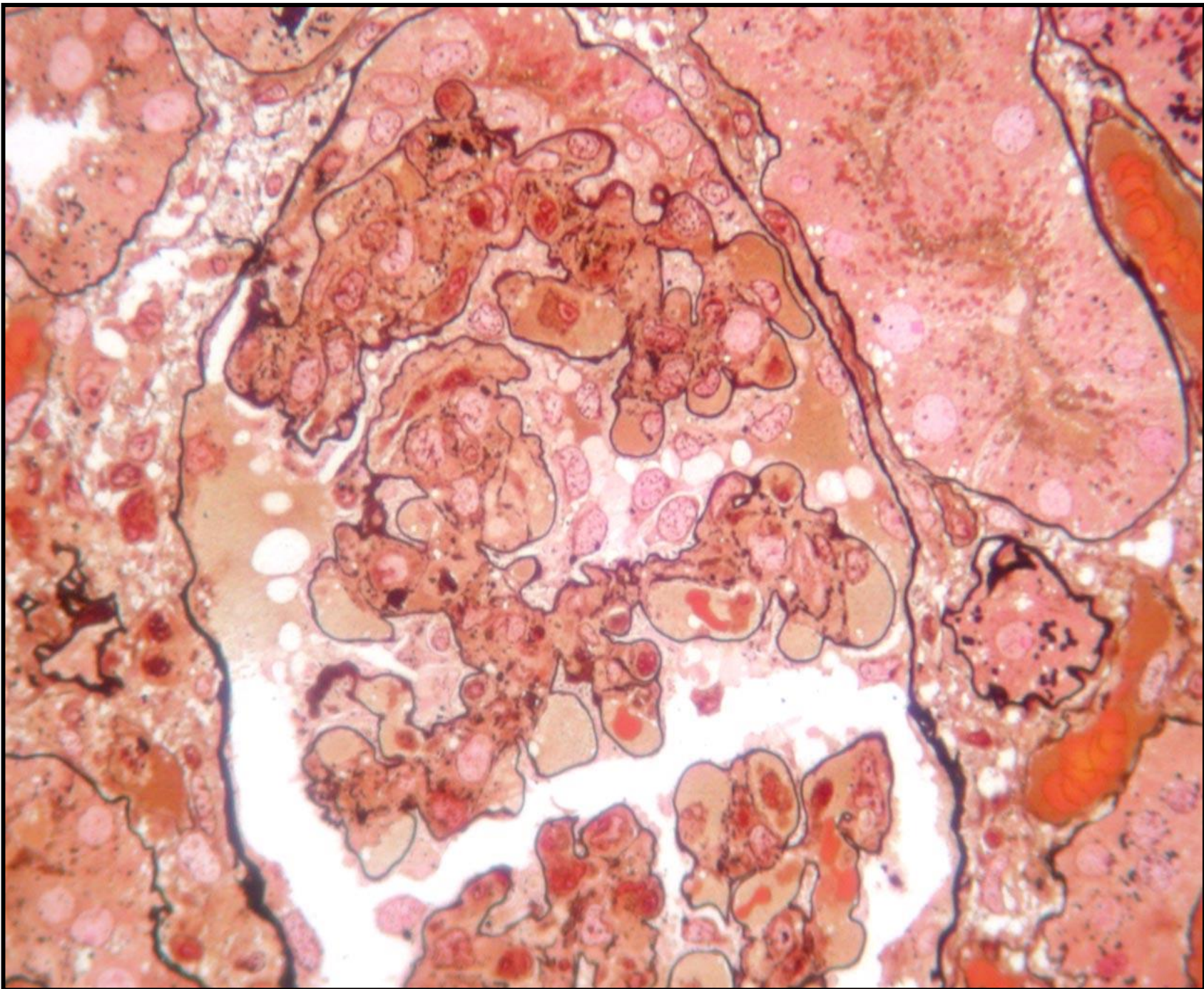


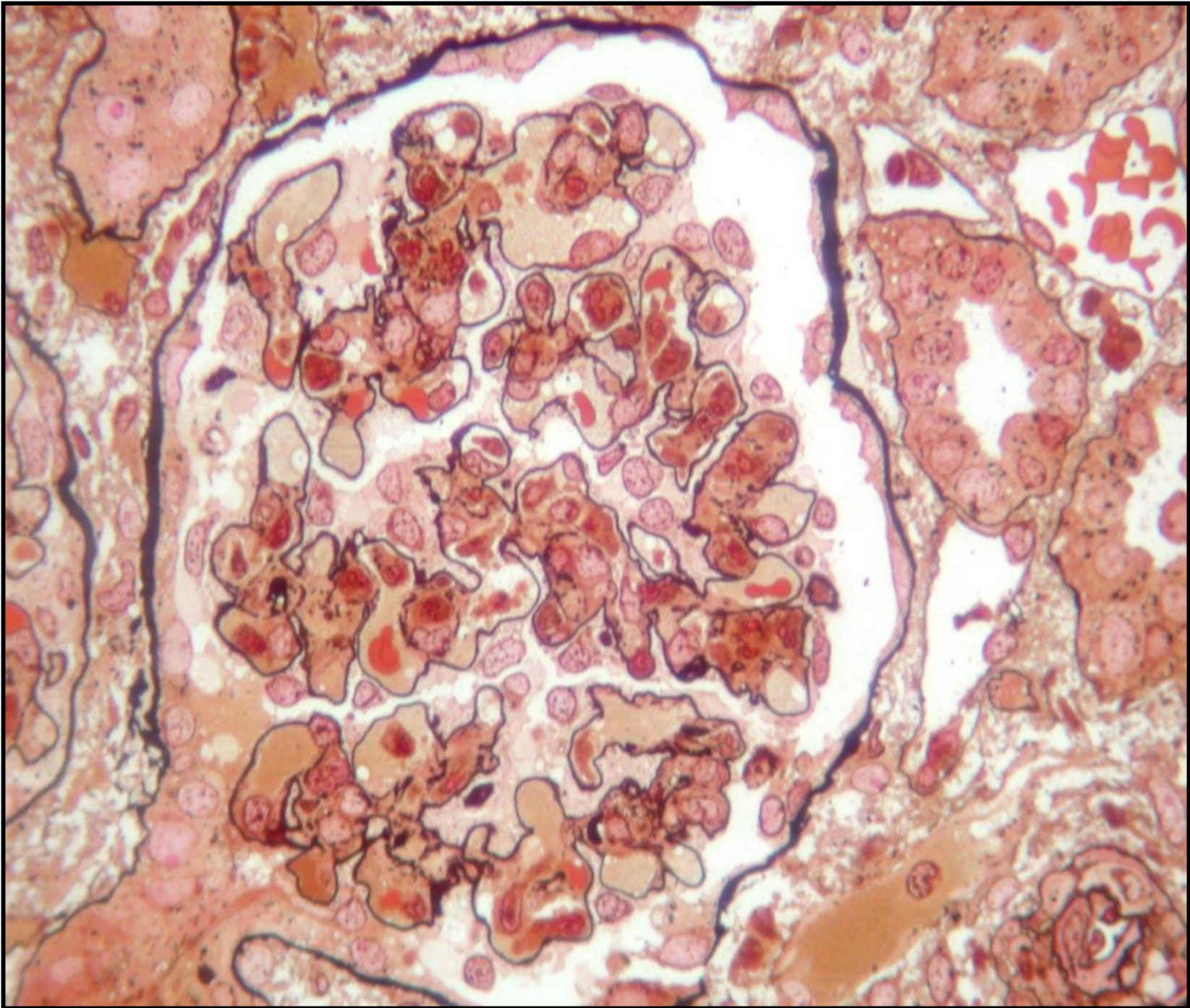
C3

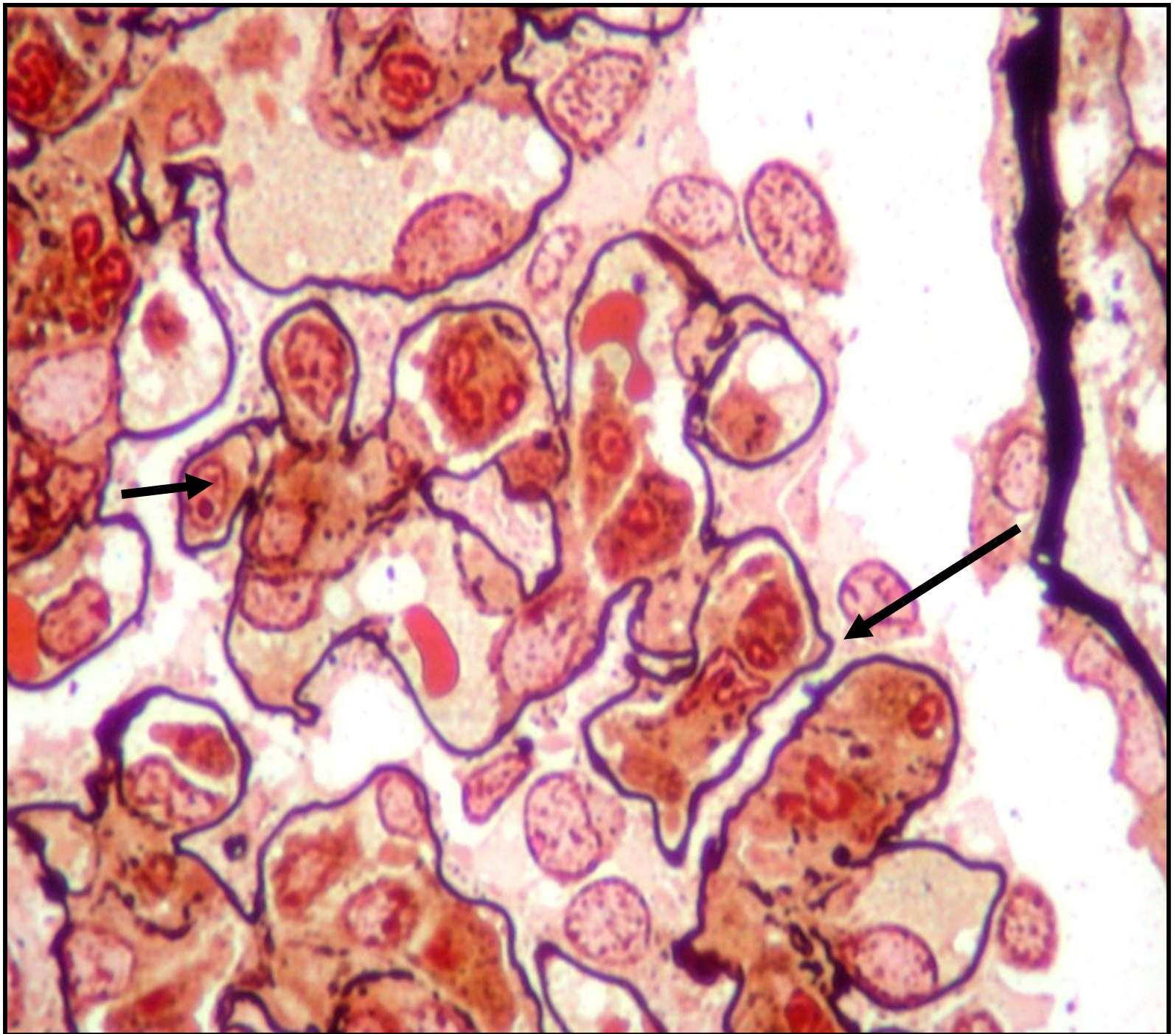


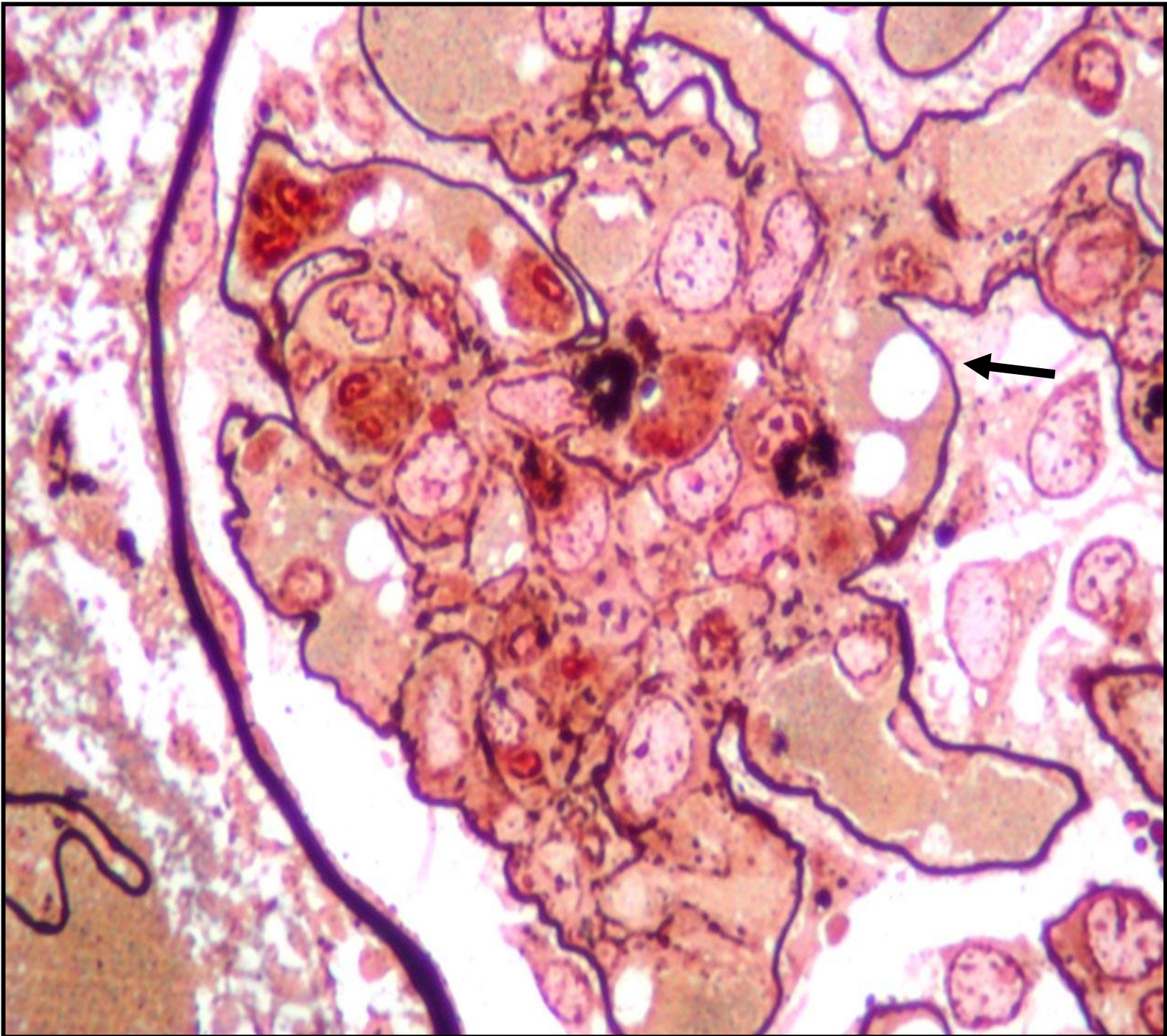
C3



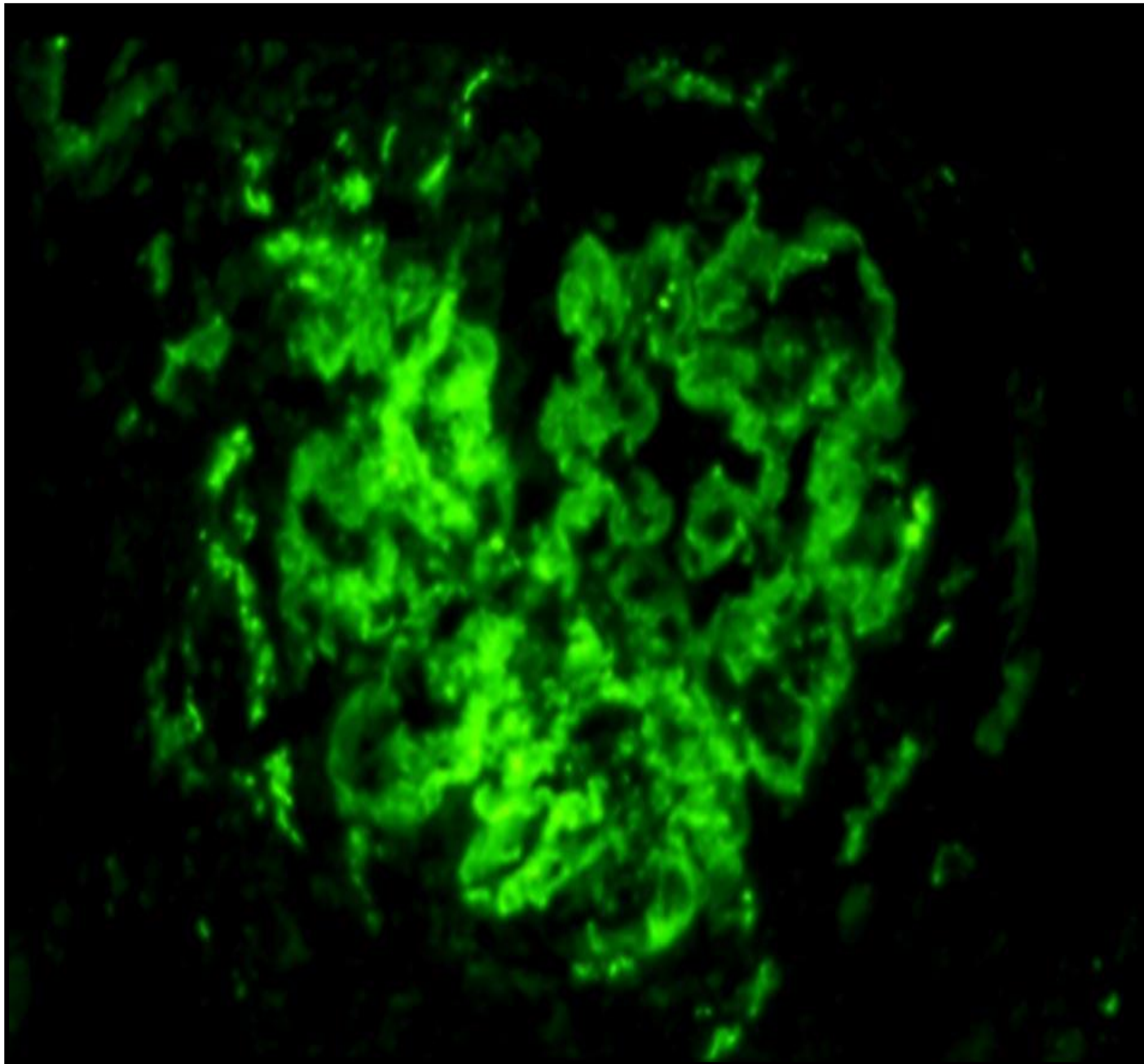




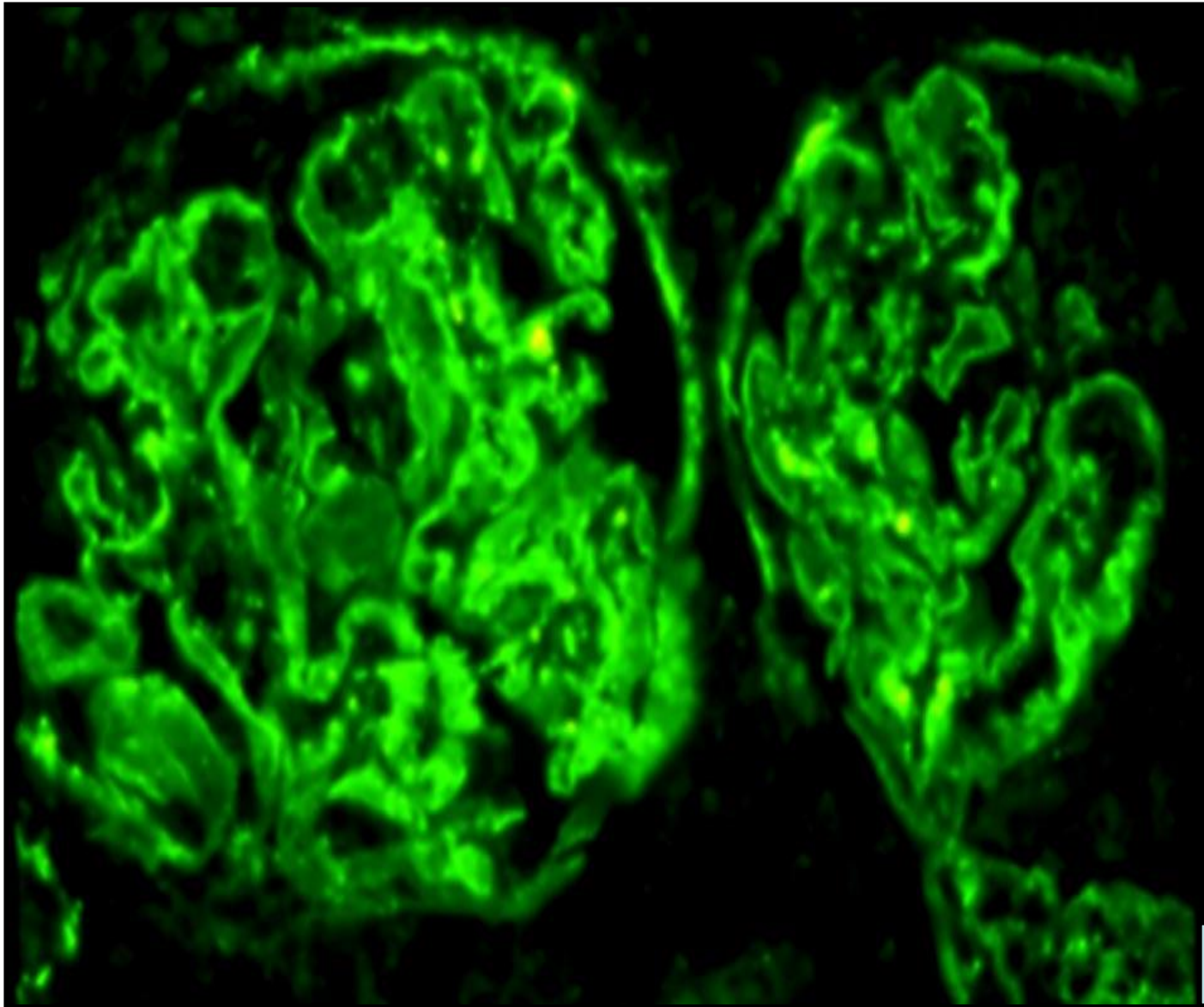




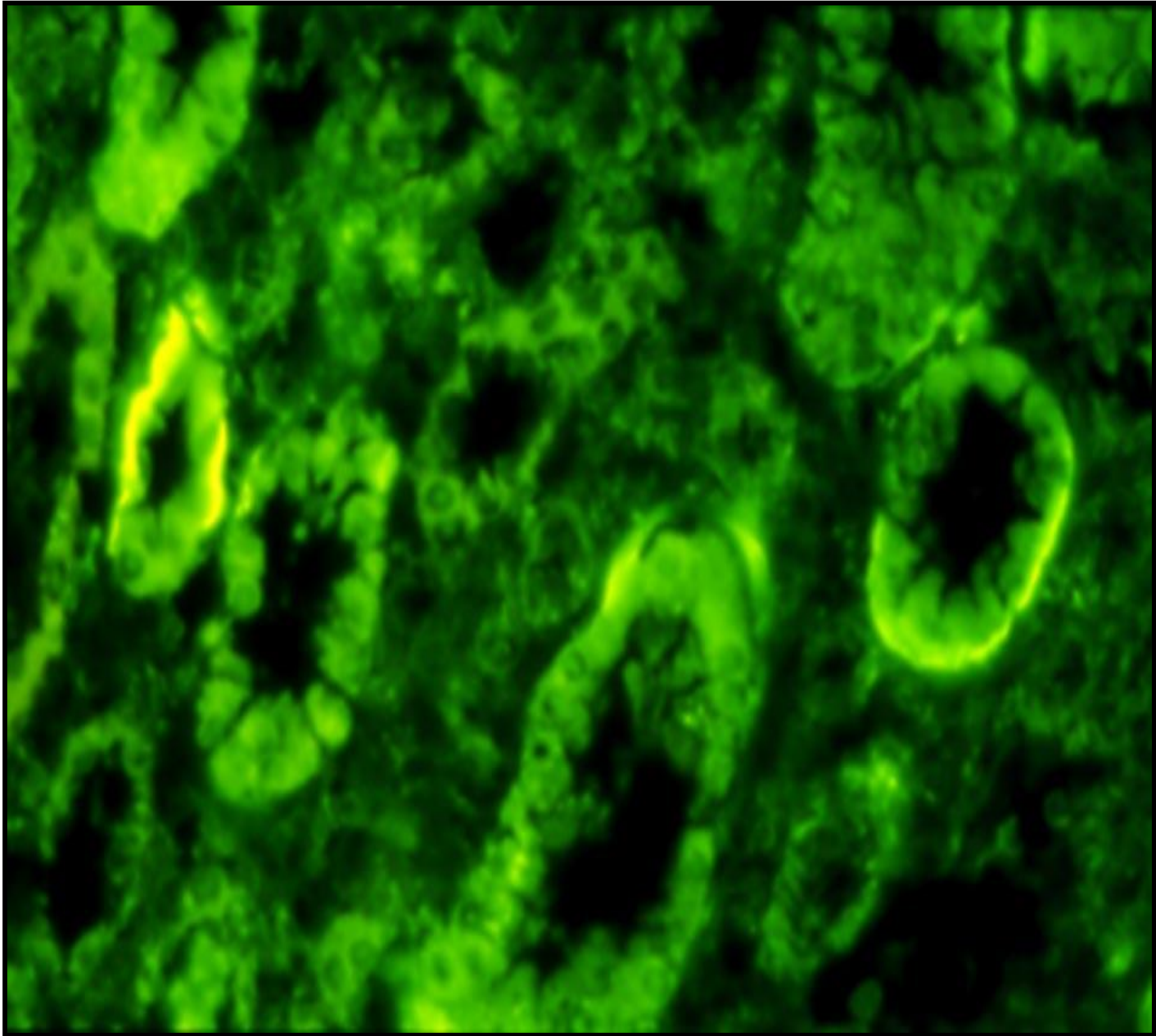
M. Benoit



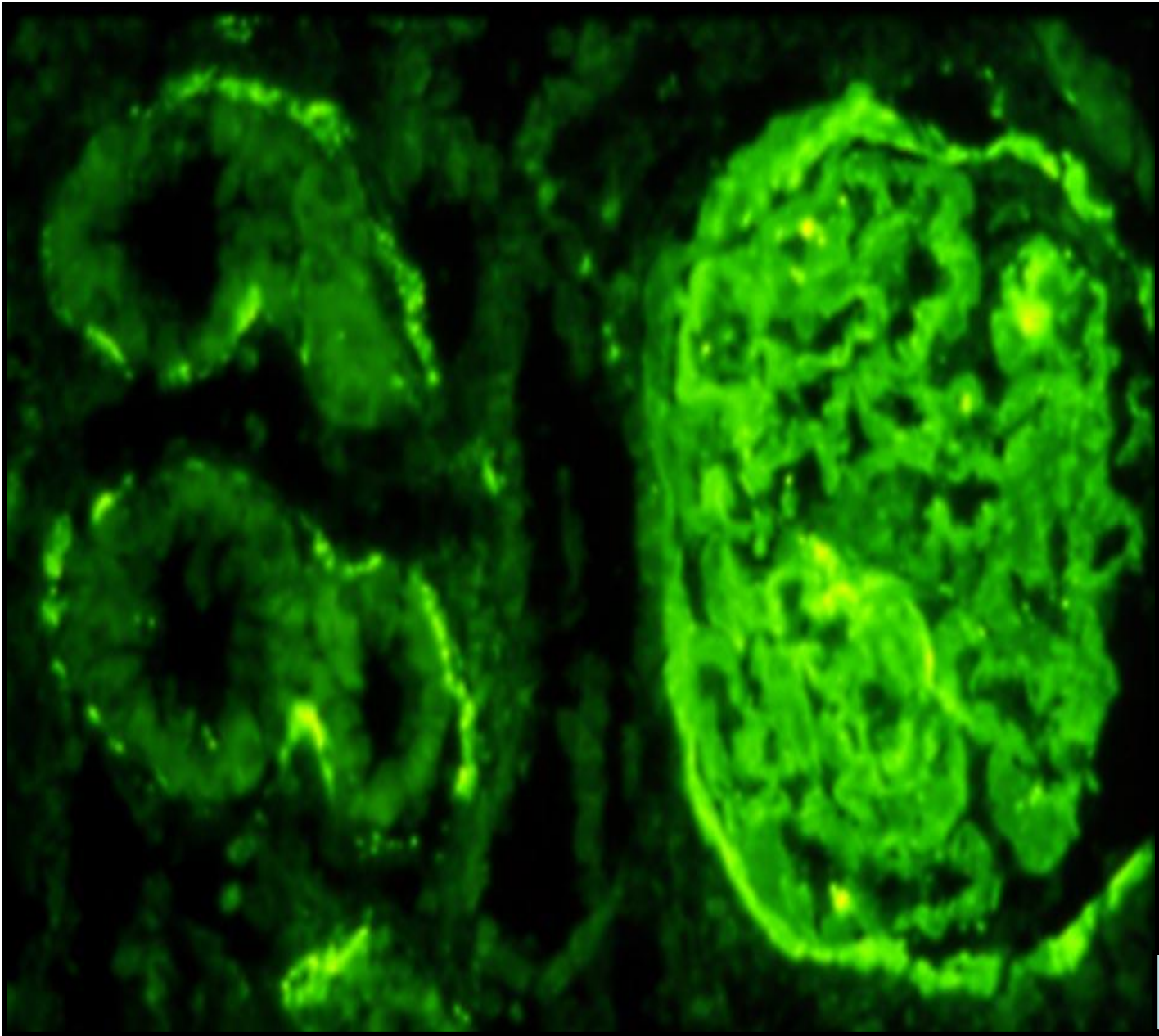
C3



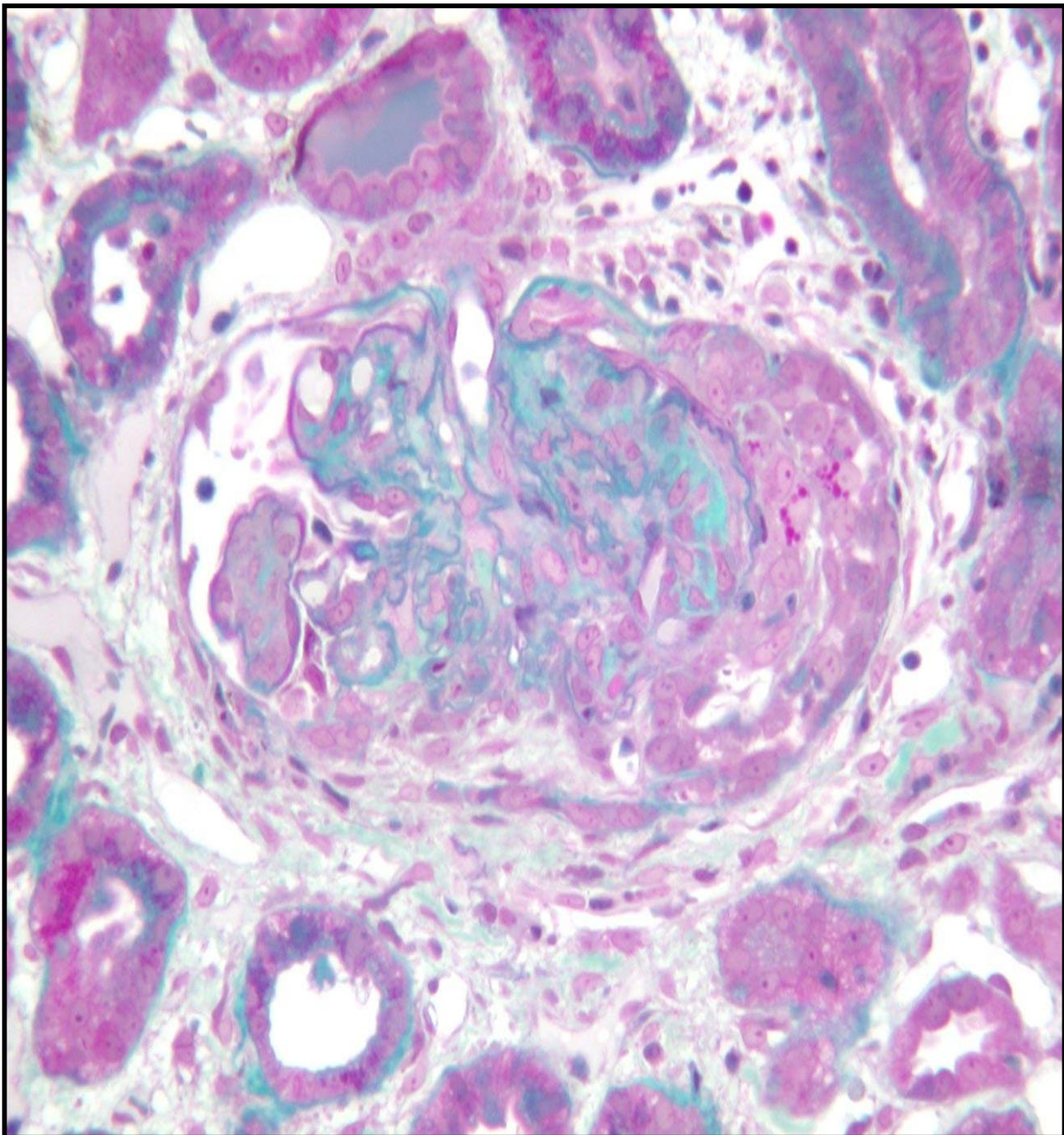
C3

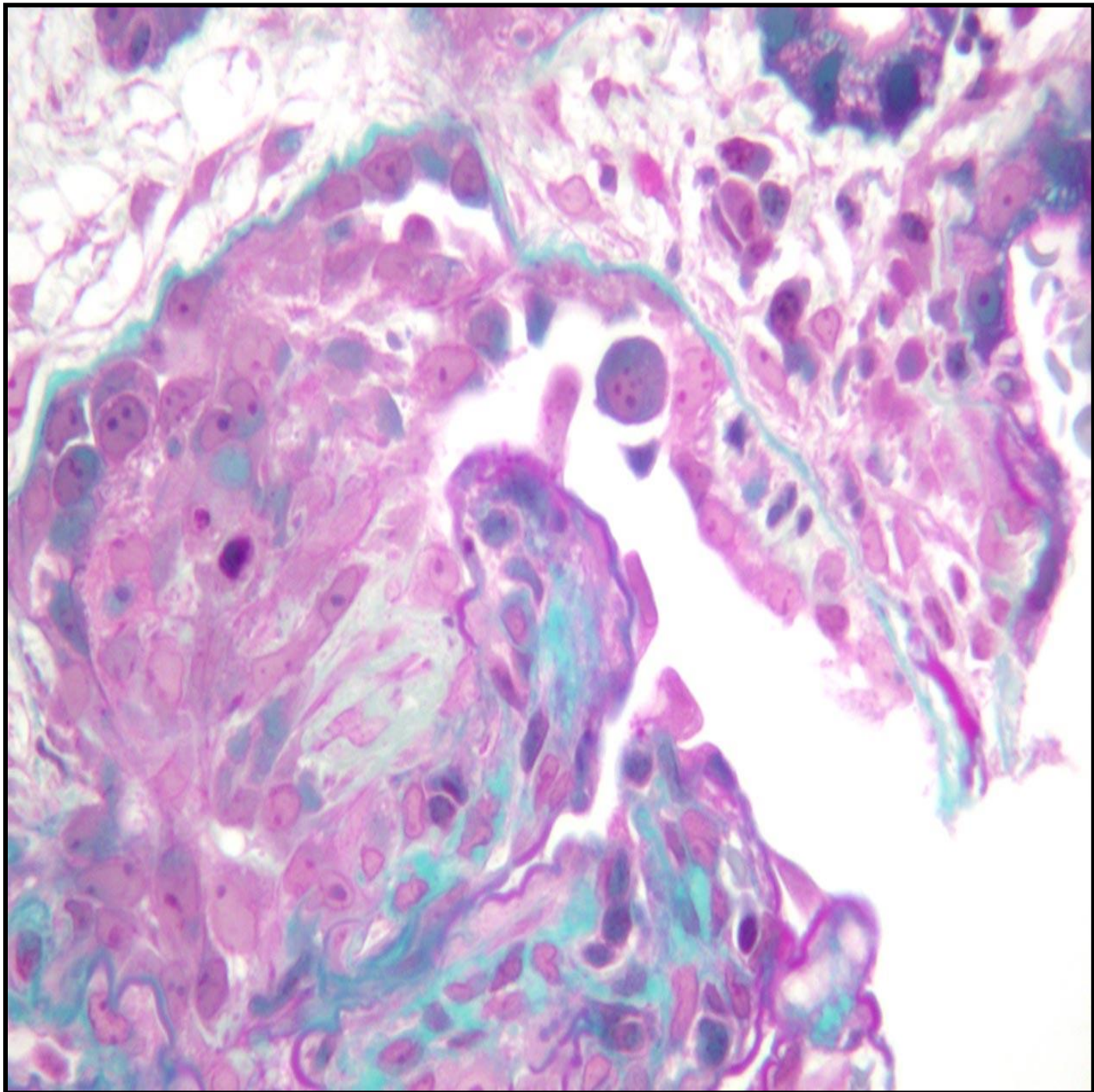


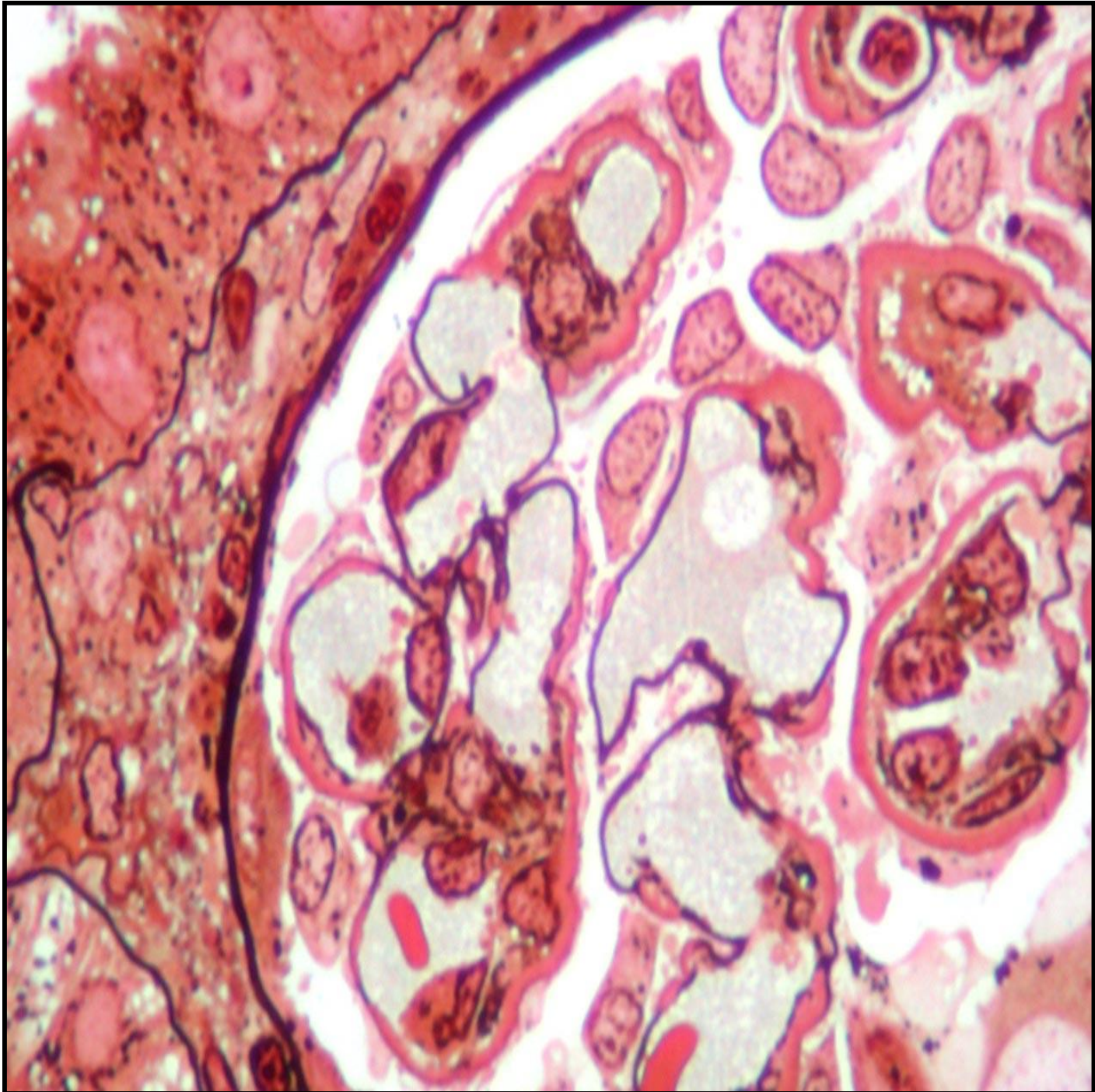
C3

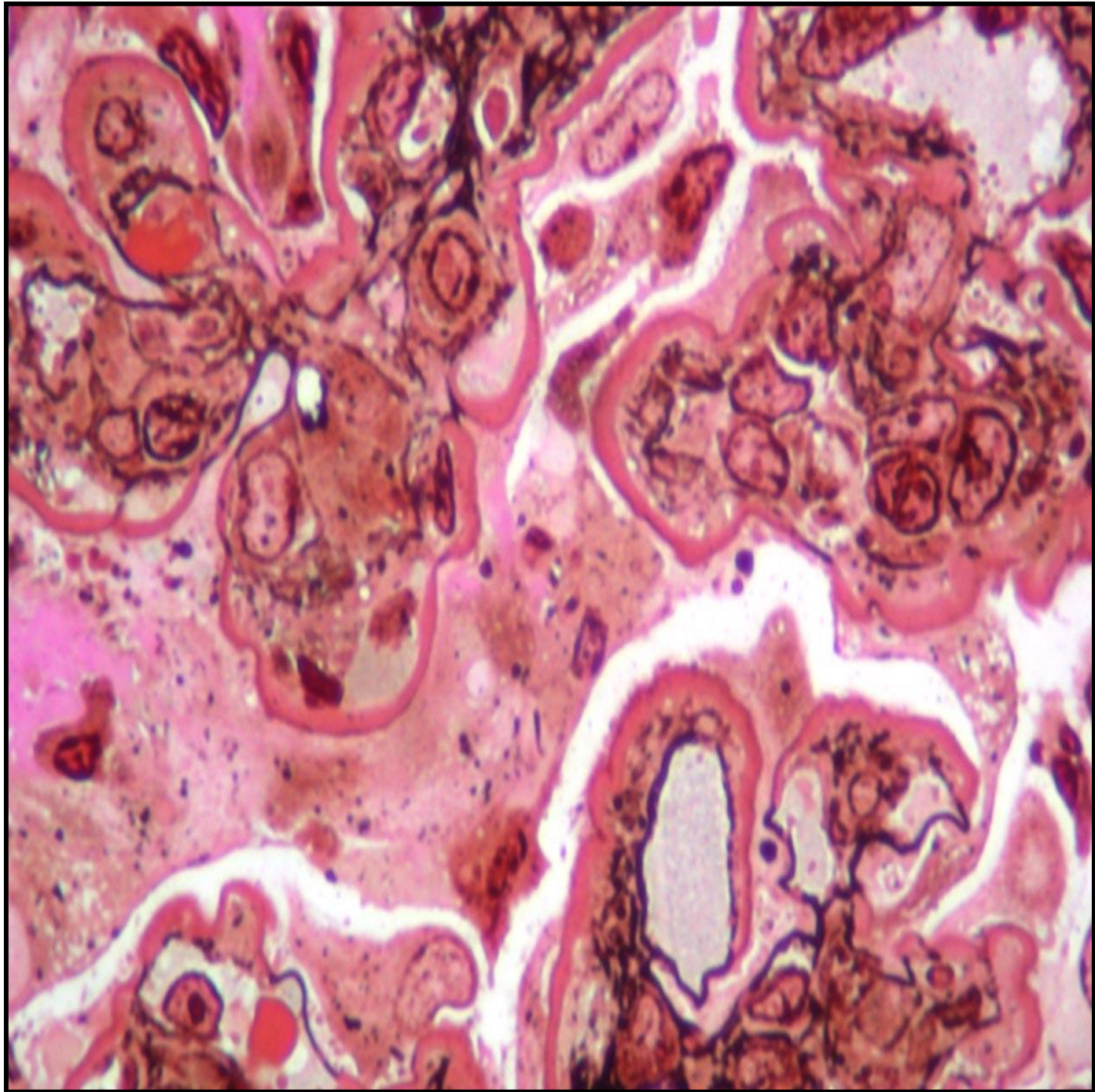


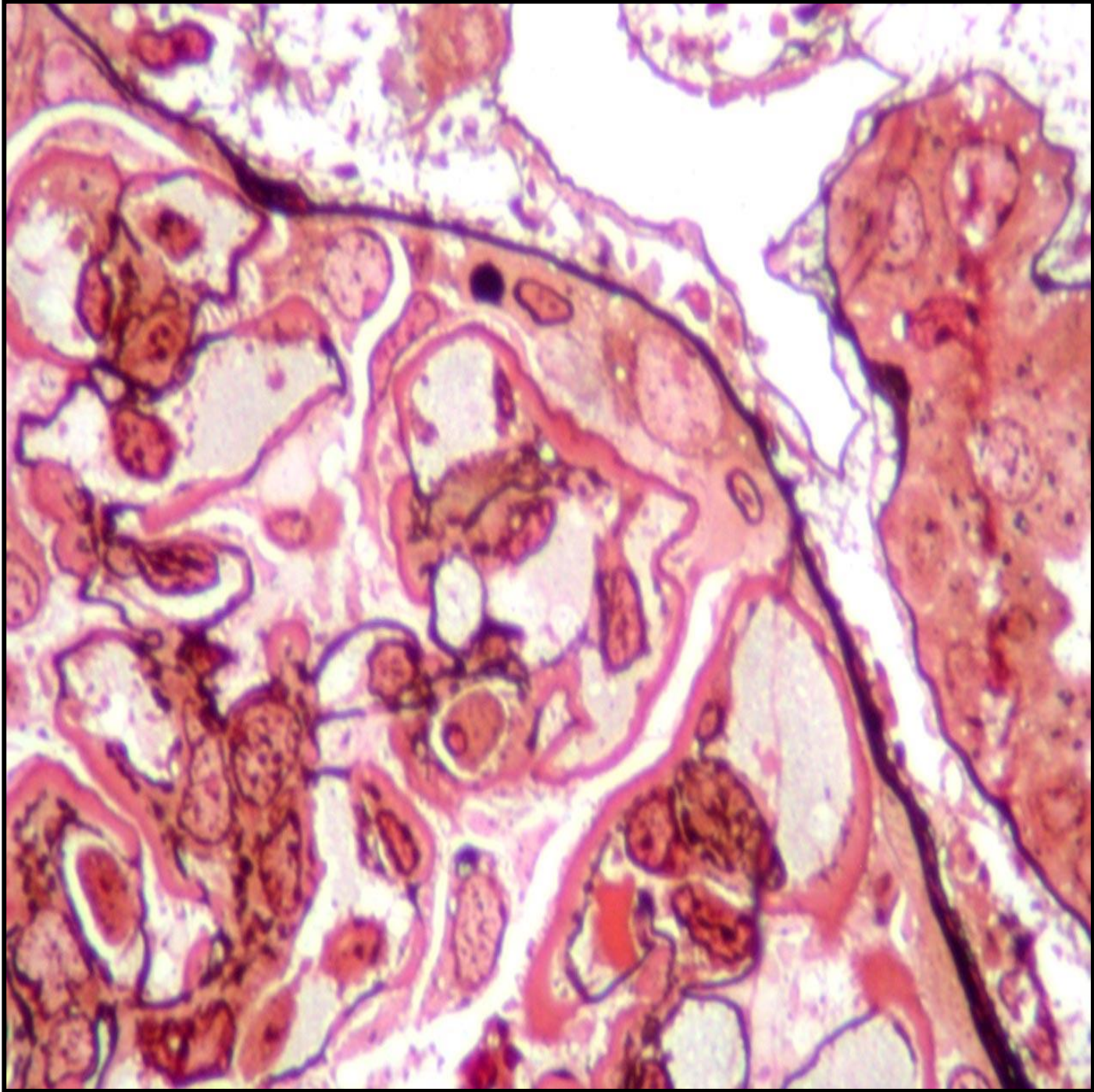
C3

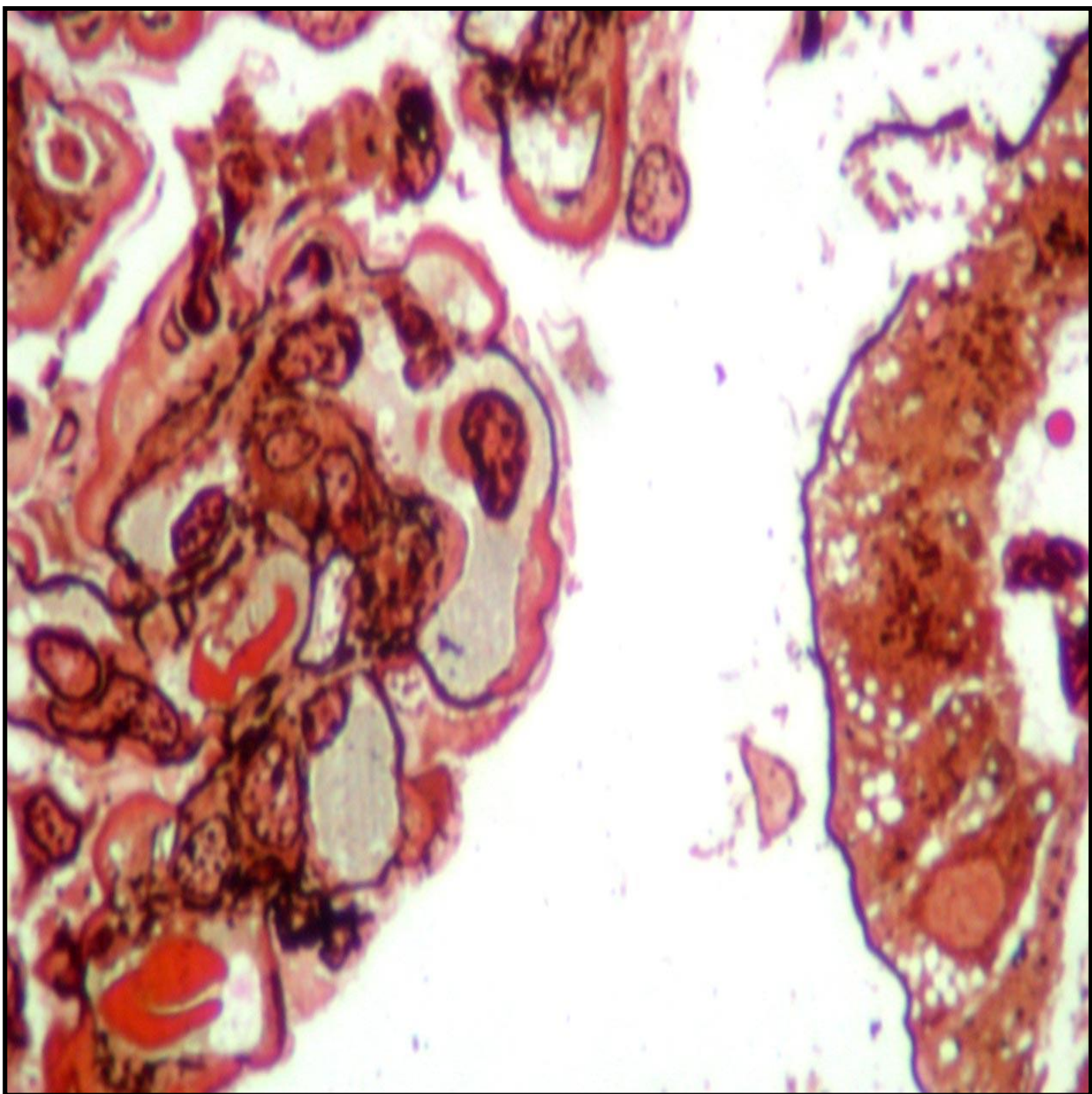




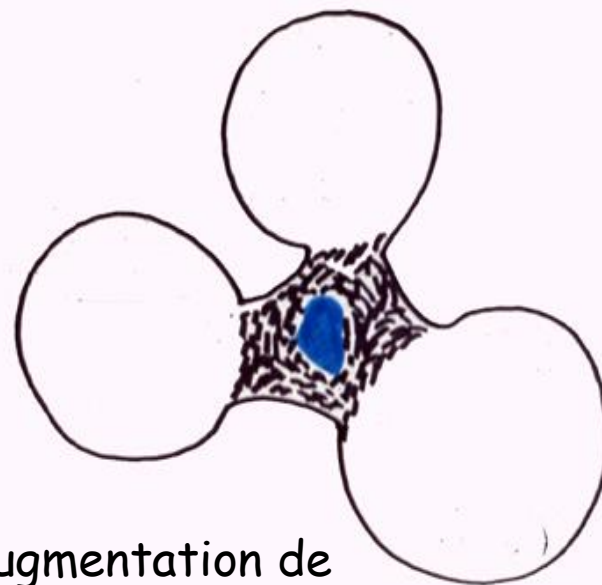
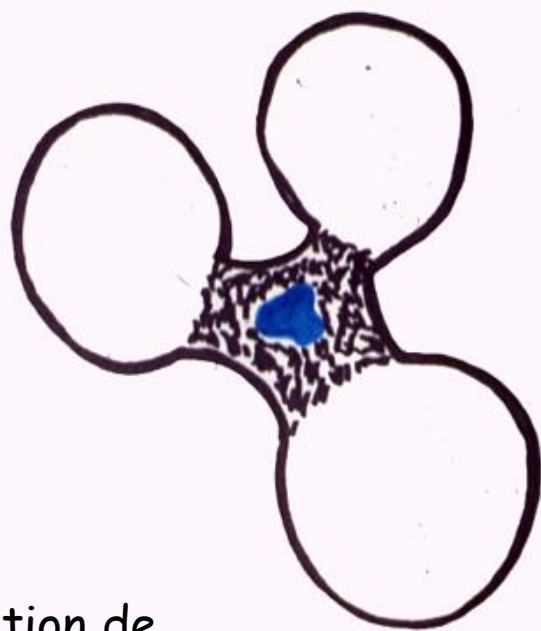




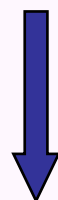




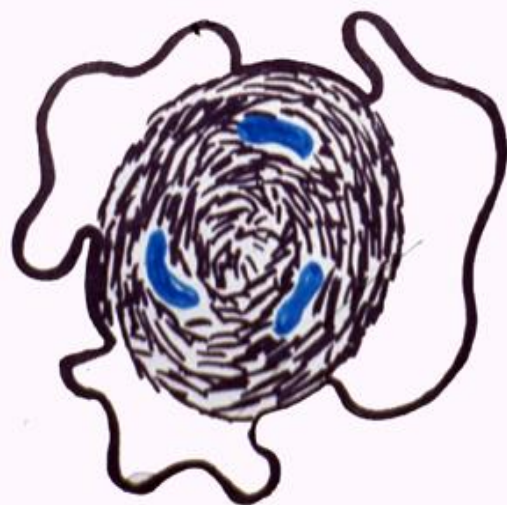
Les glomérulopathies diabétiques



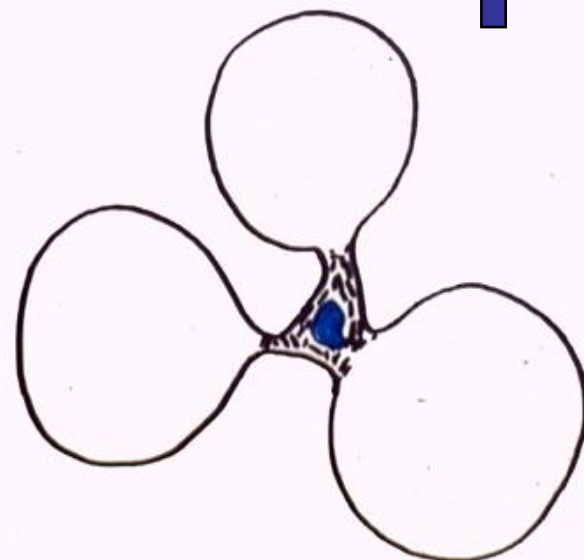
Augmentation de la matrice mésangiale



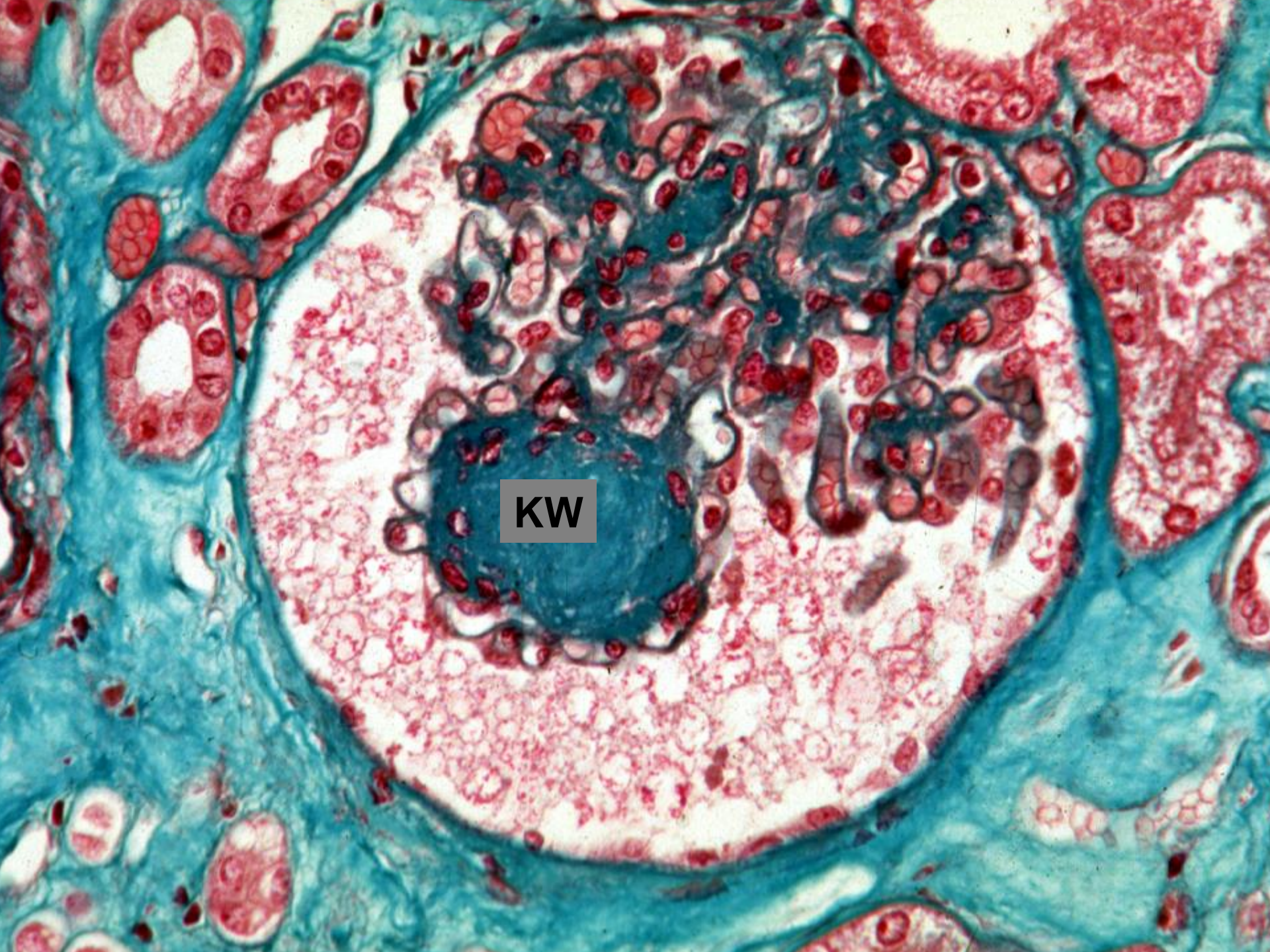
Augmentation de l'épaisseur des MBG



Nodule de KW



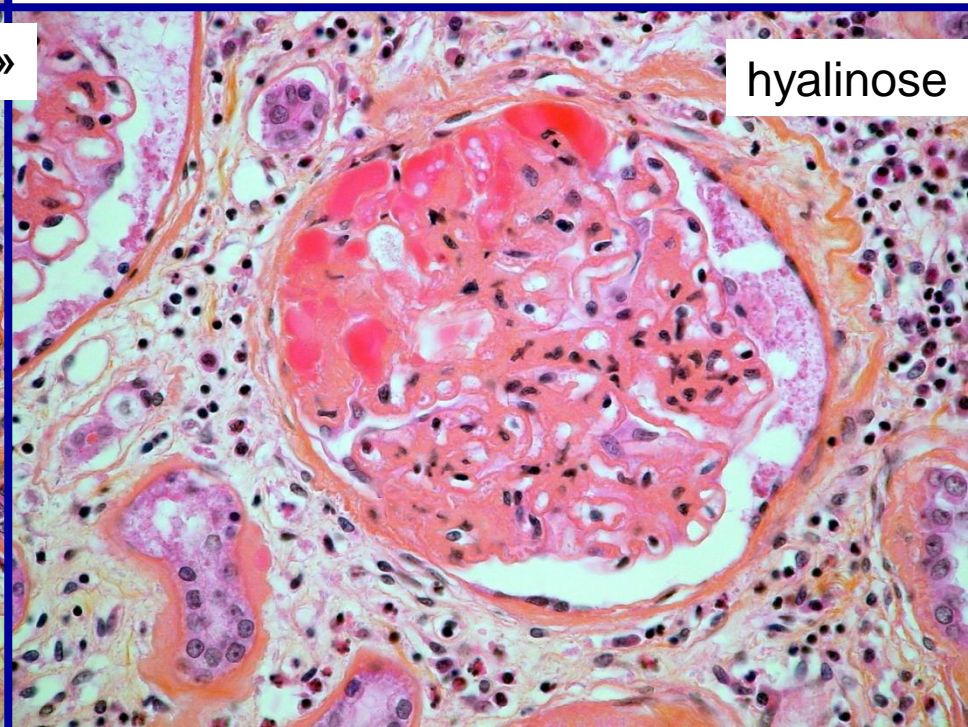
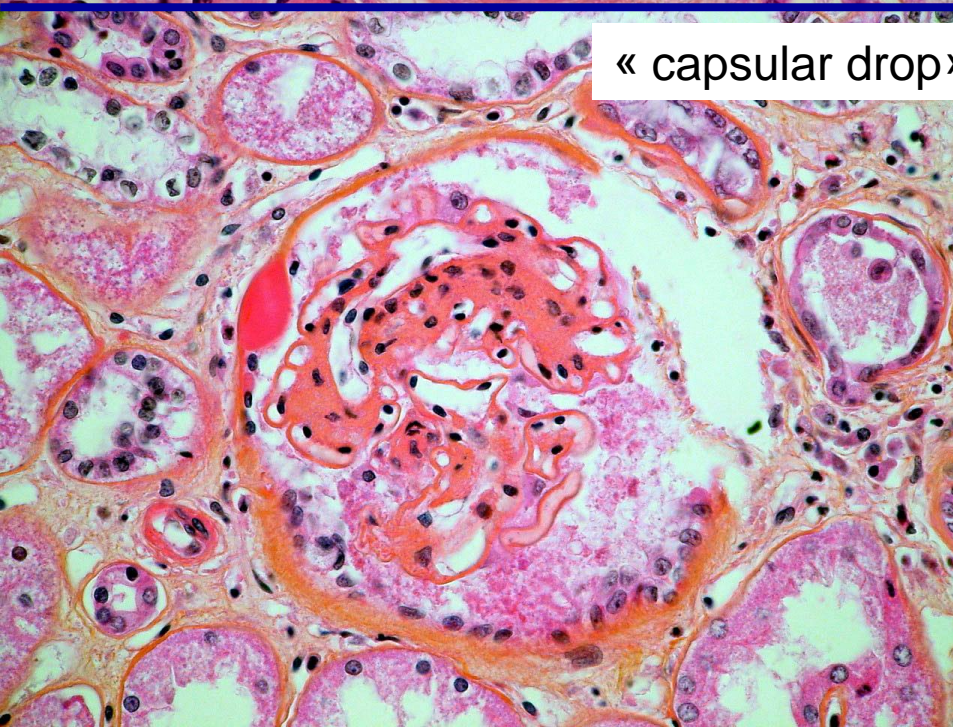
normal



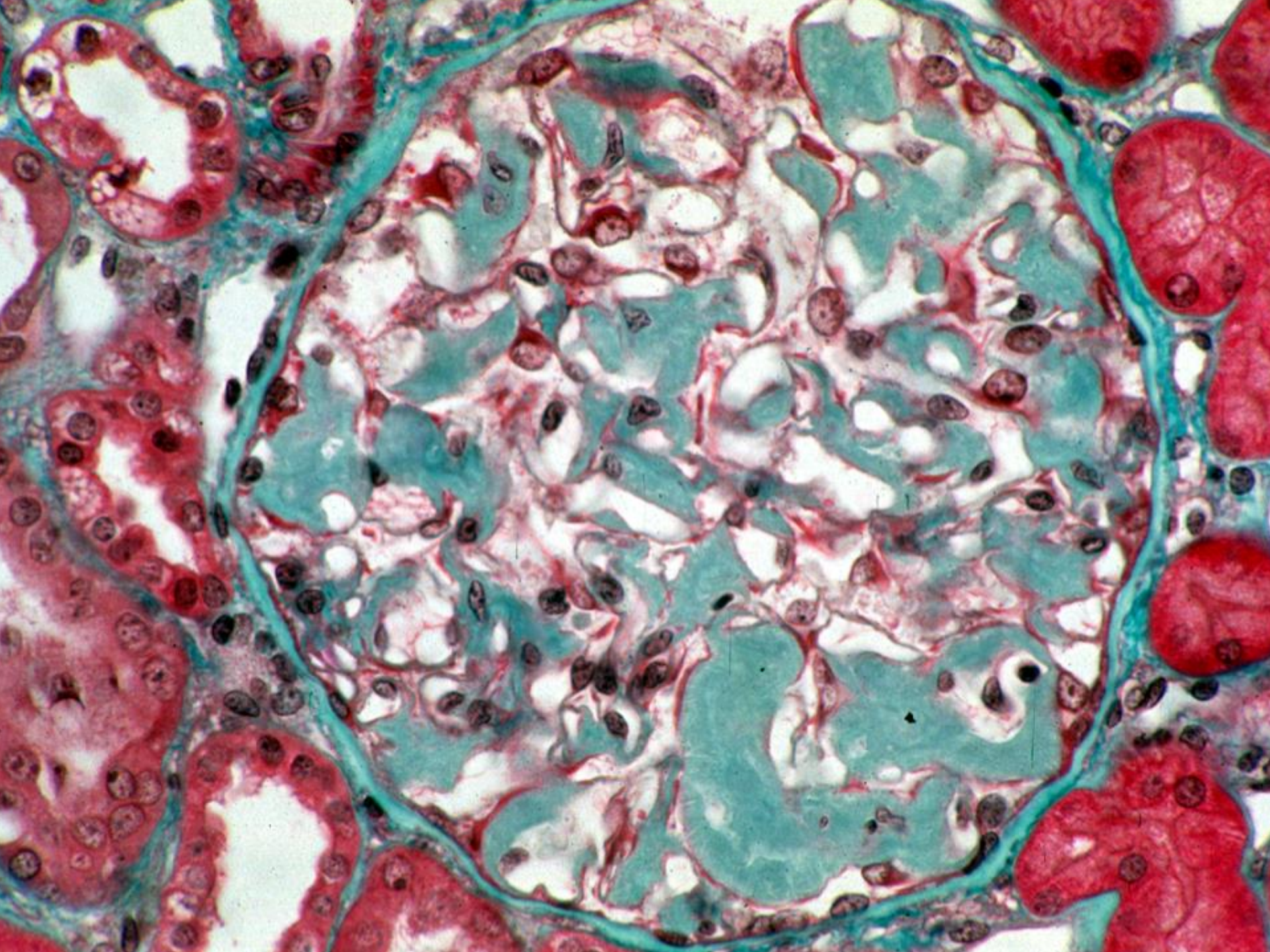
KW

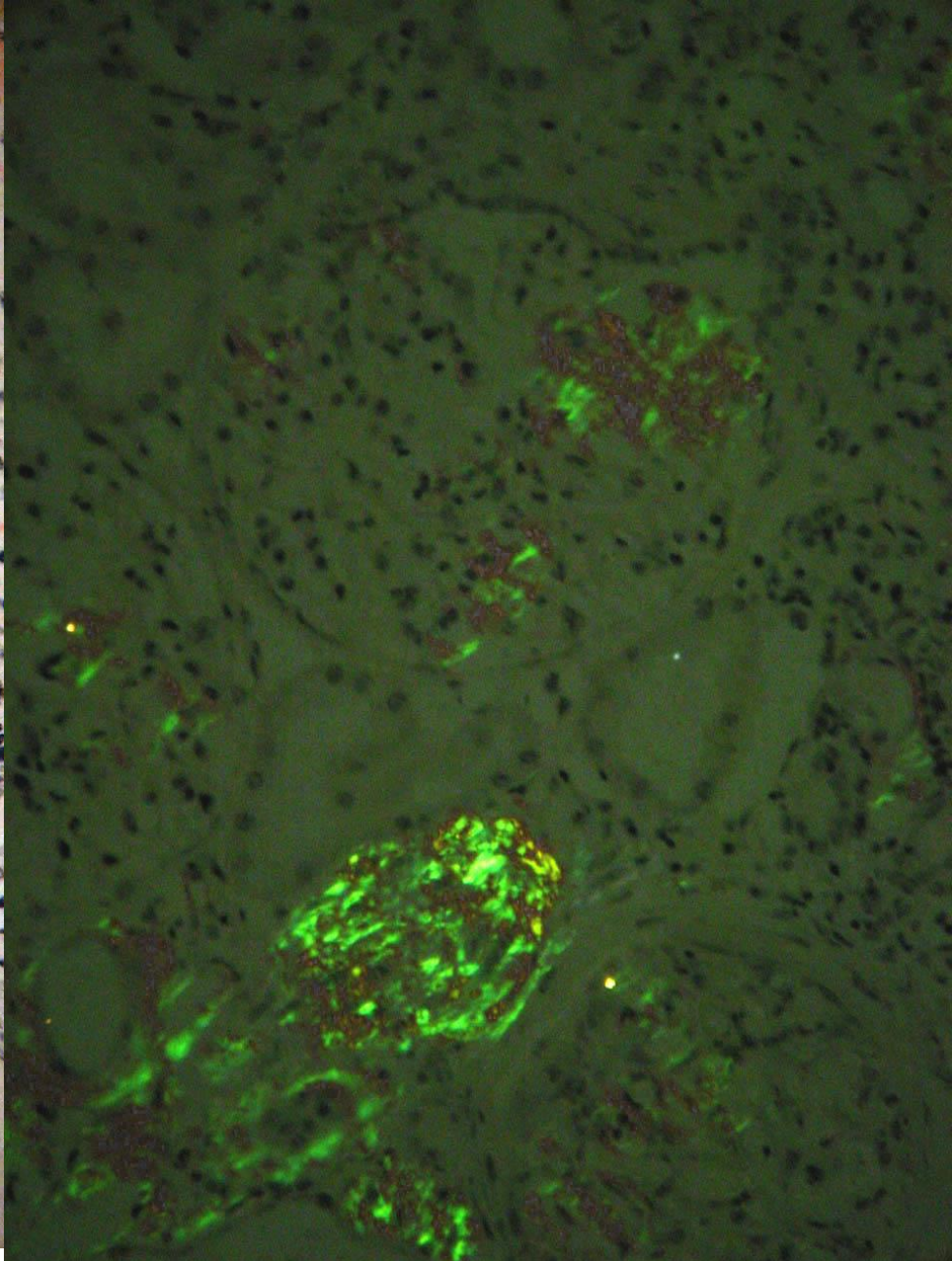
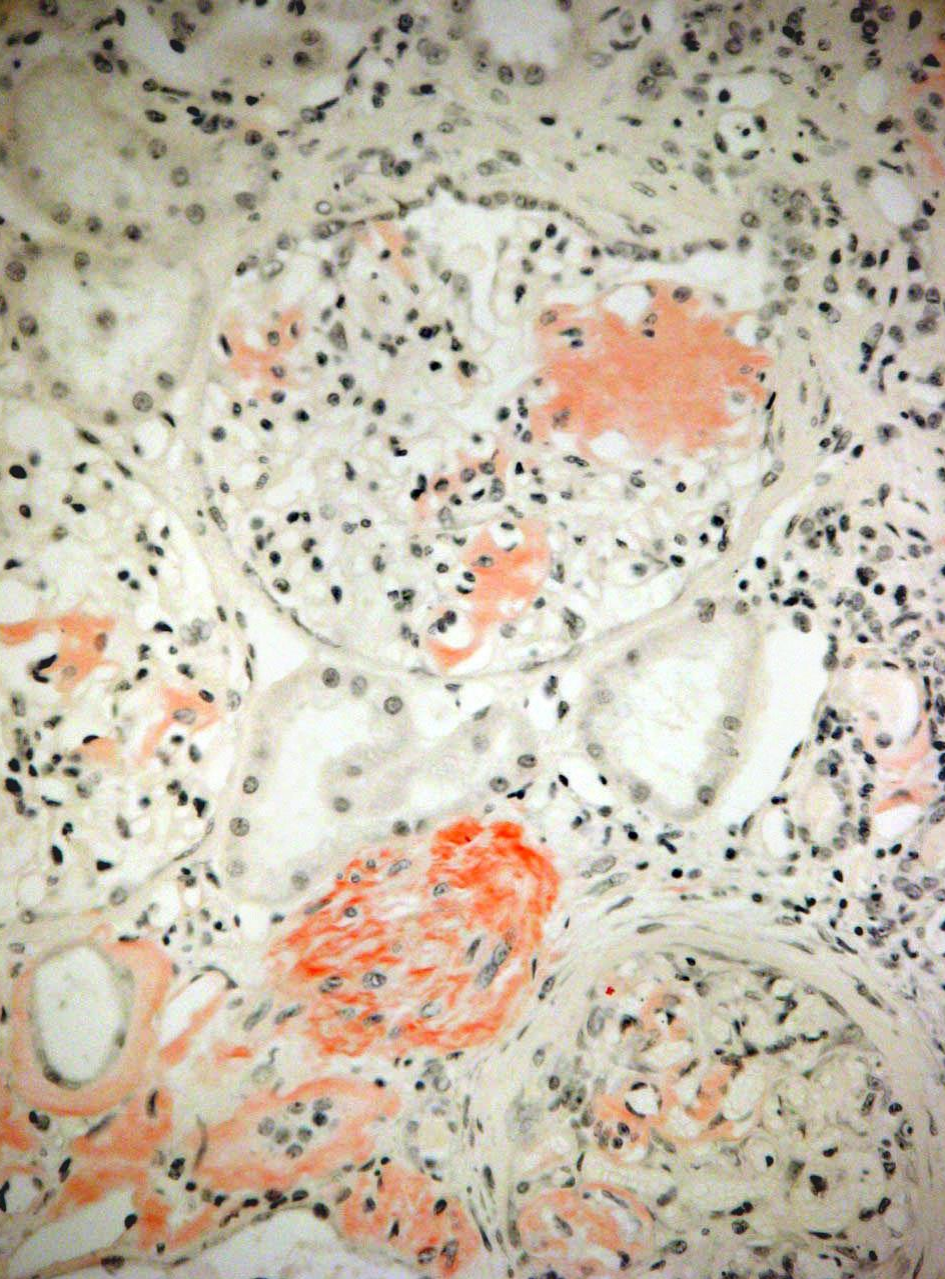


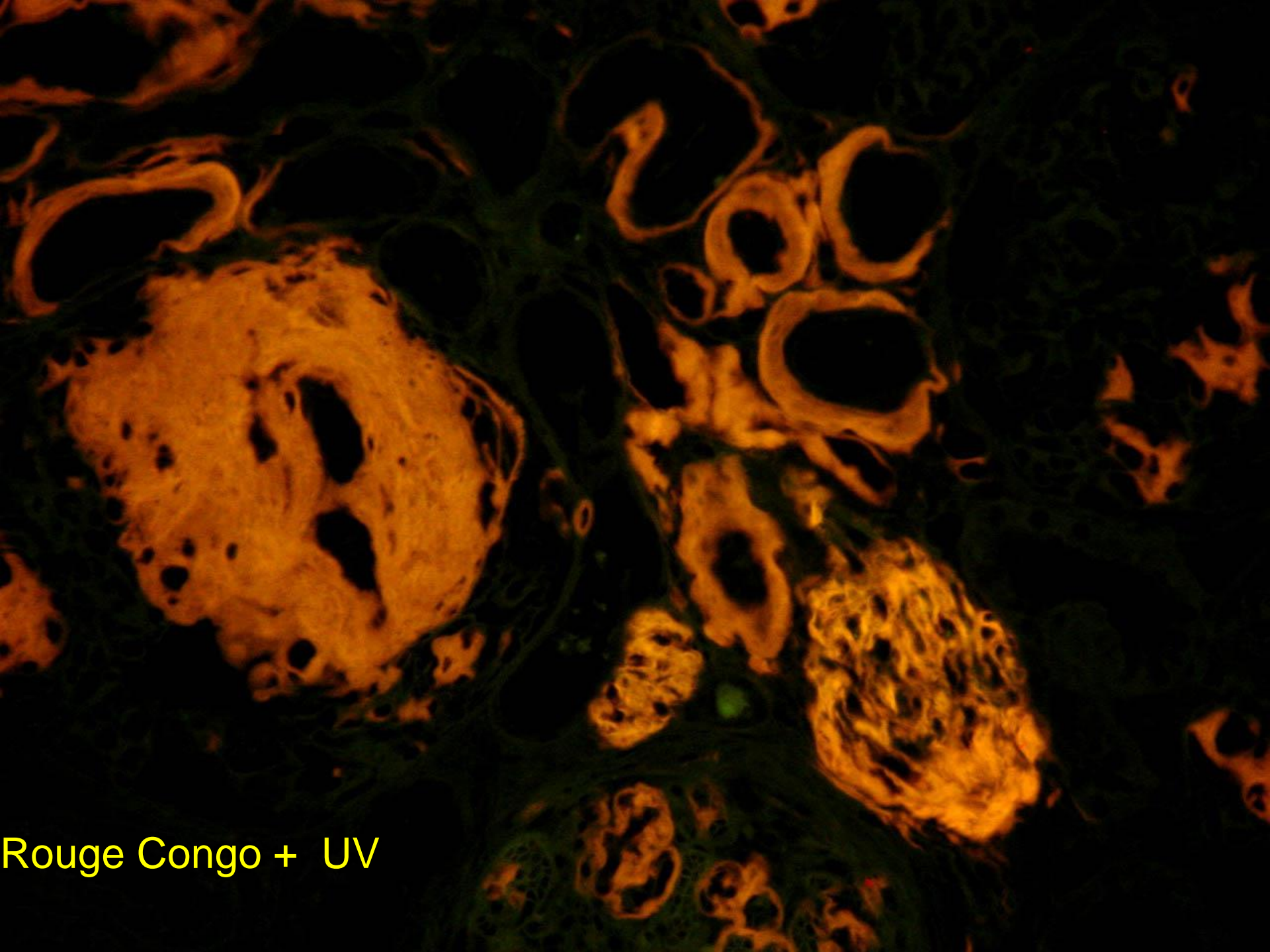
Diabète



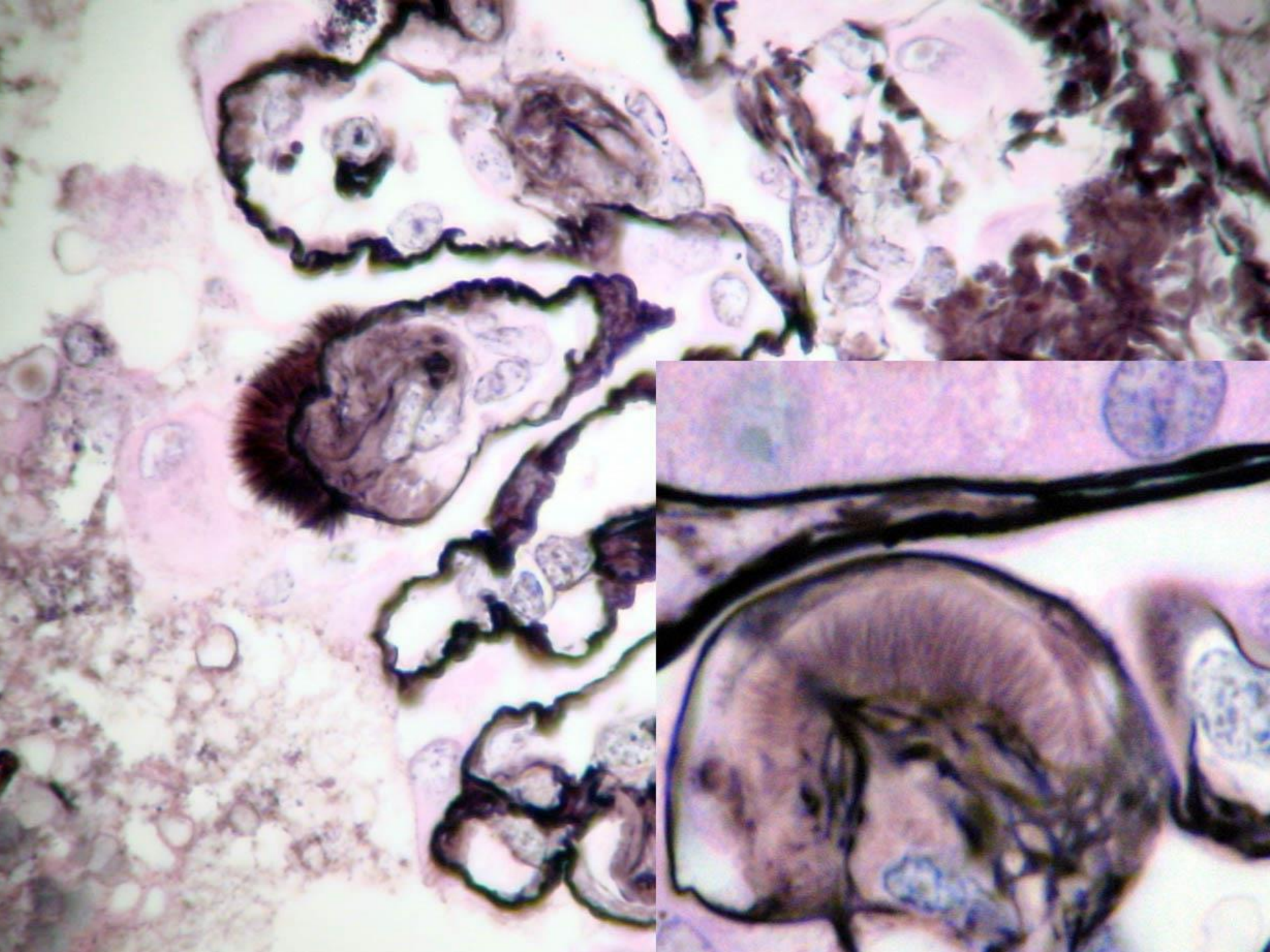
Les glomérulopathies amyloïdes



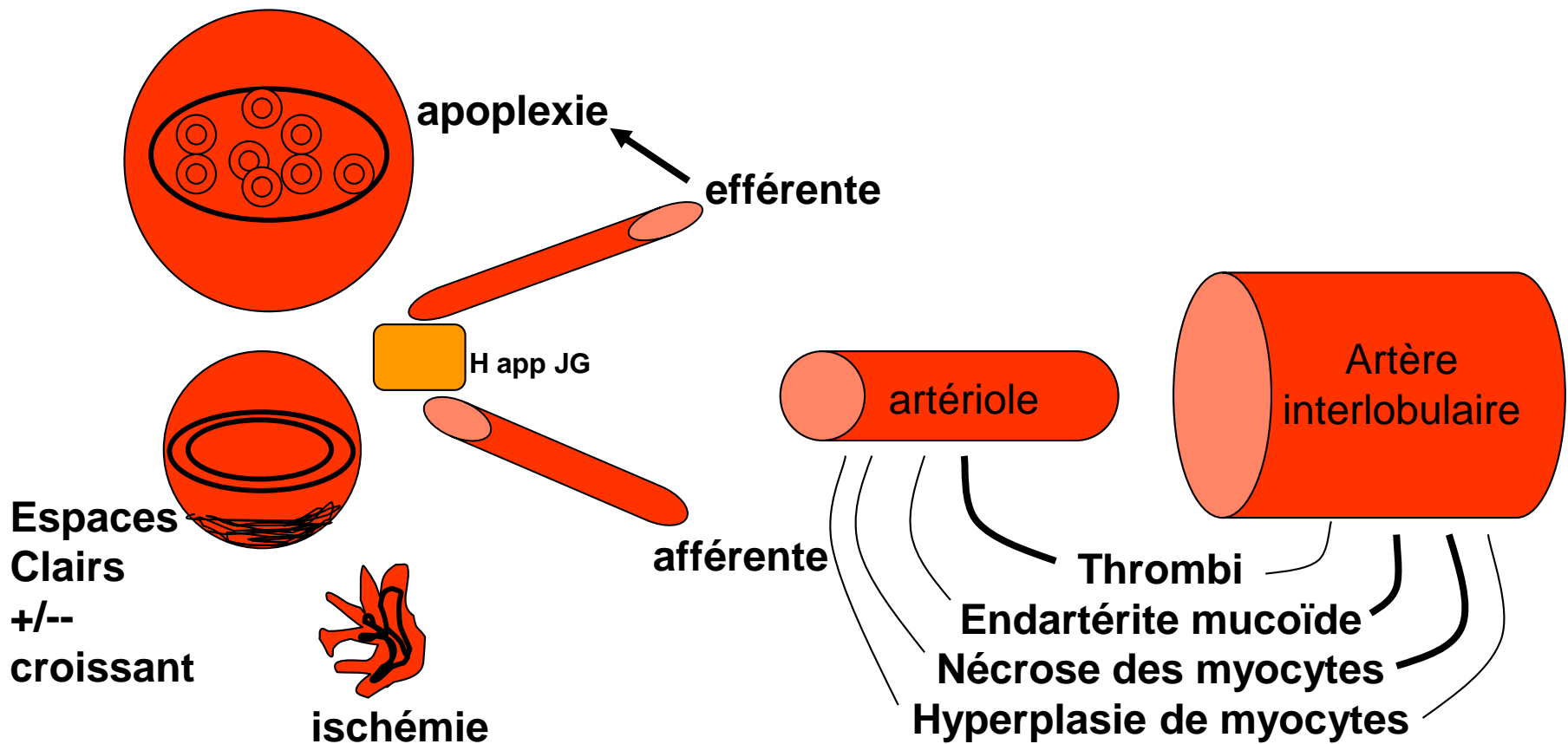




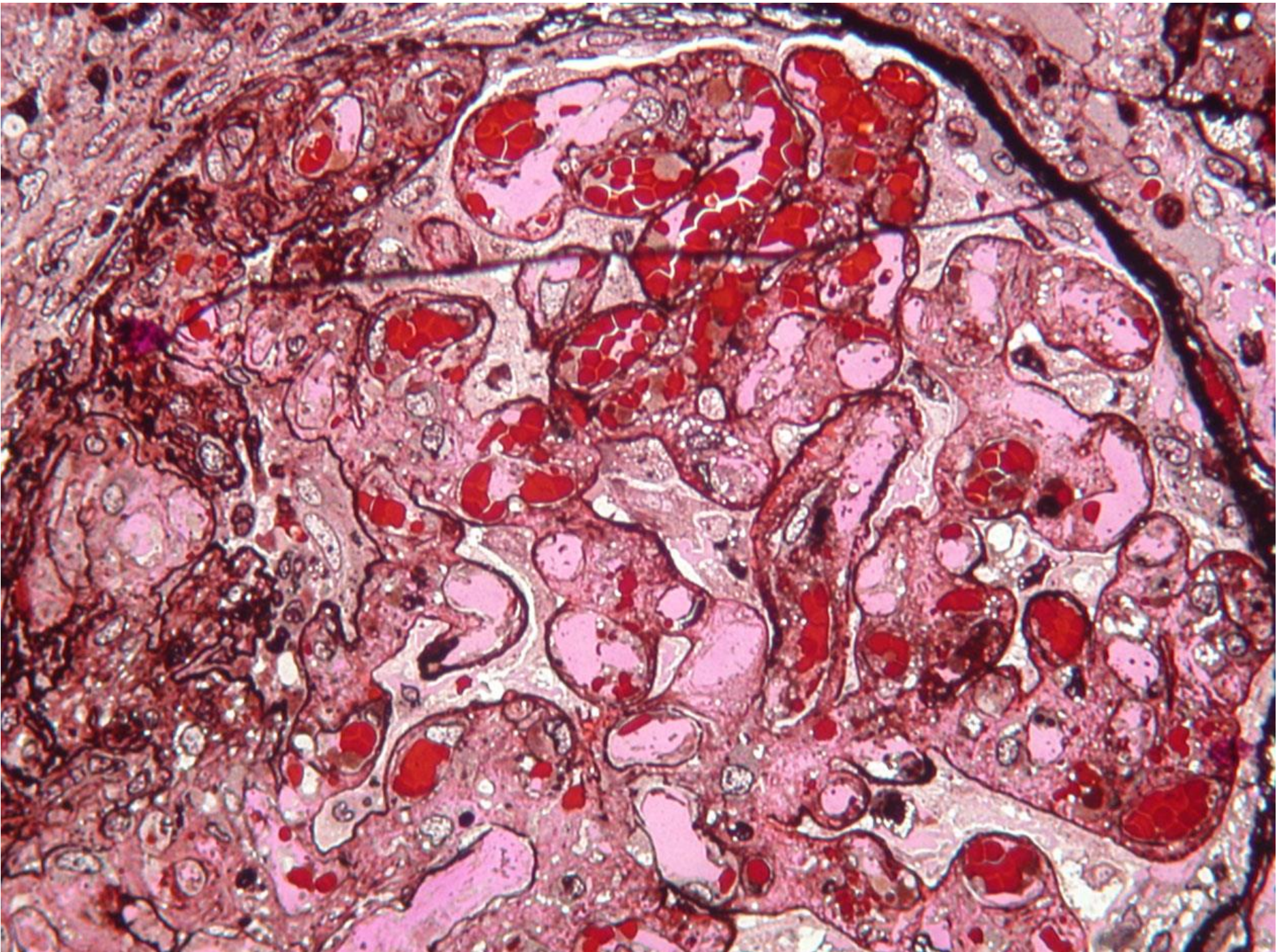
Rouge Congo + UV



Microangiopathies

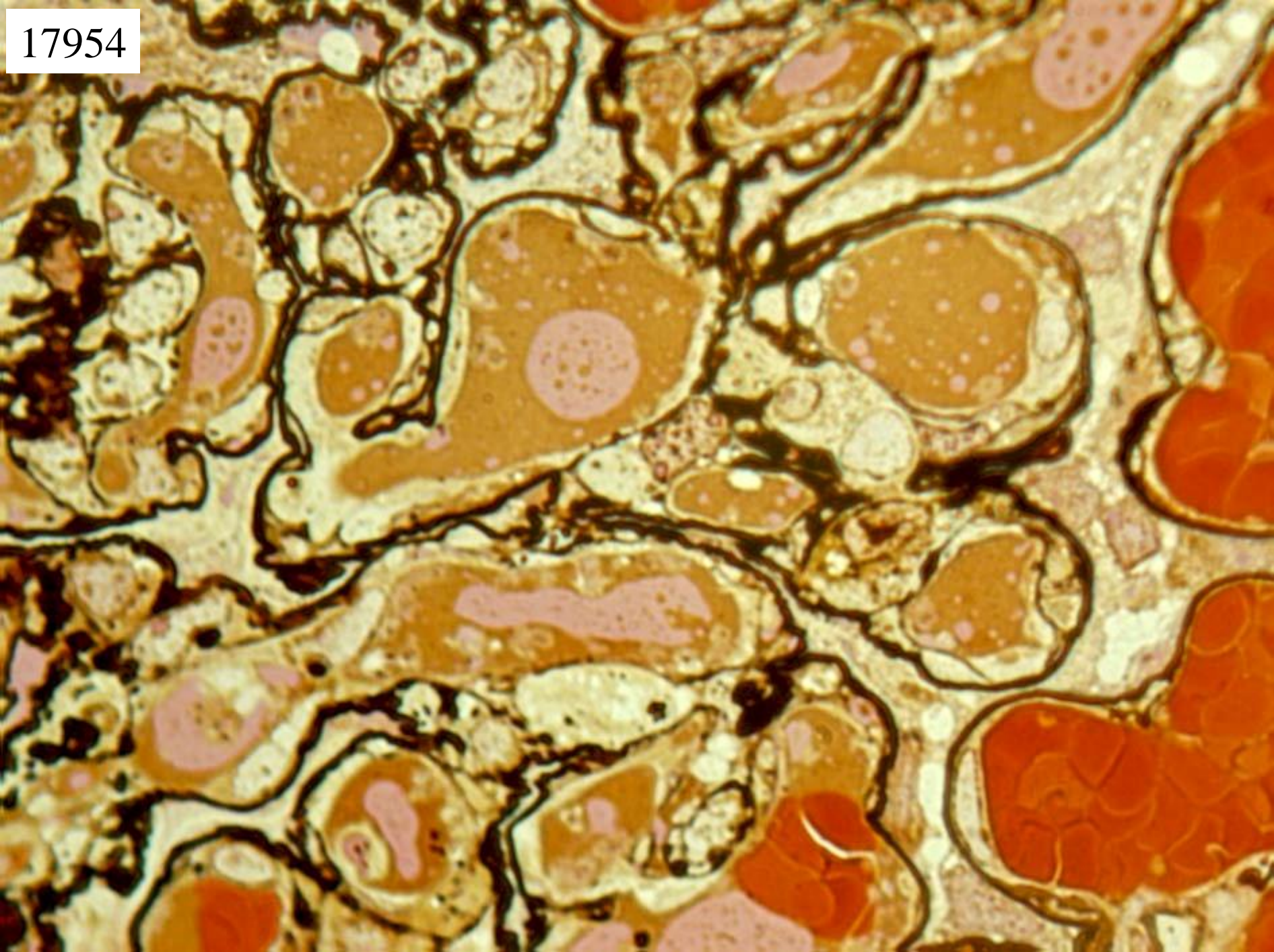


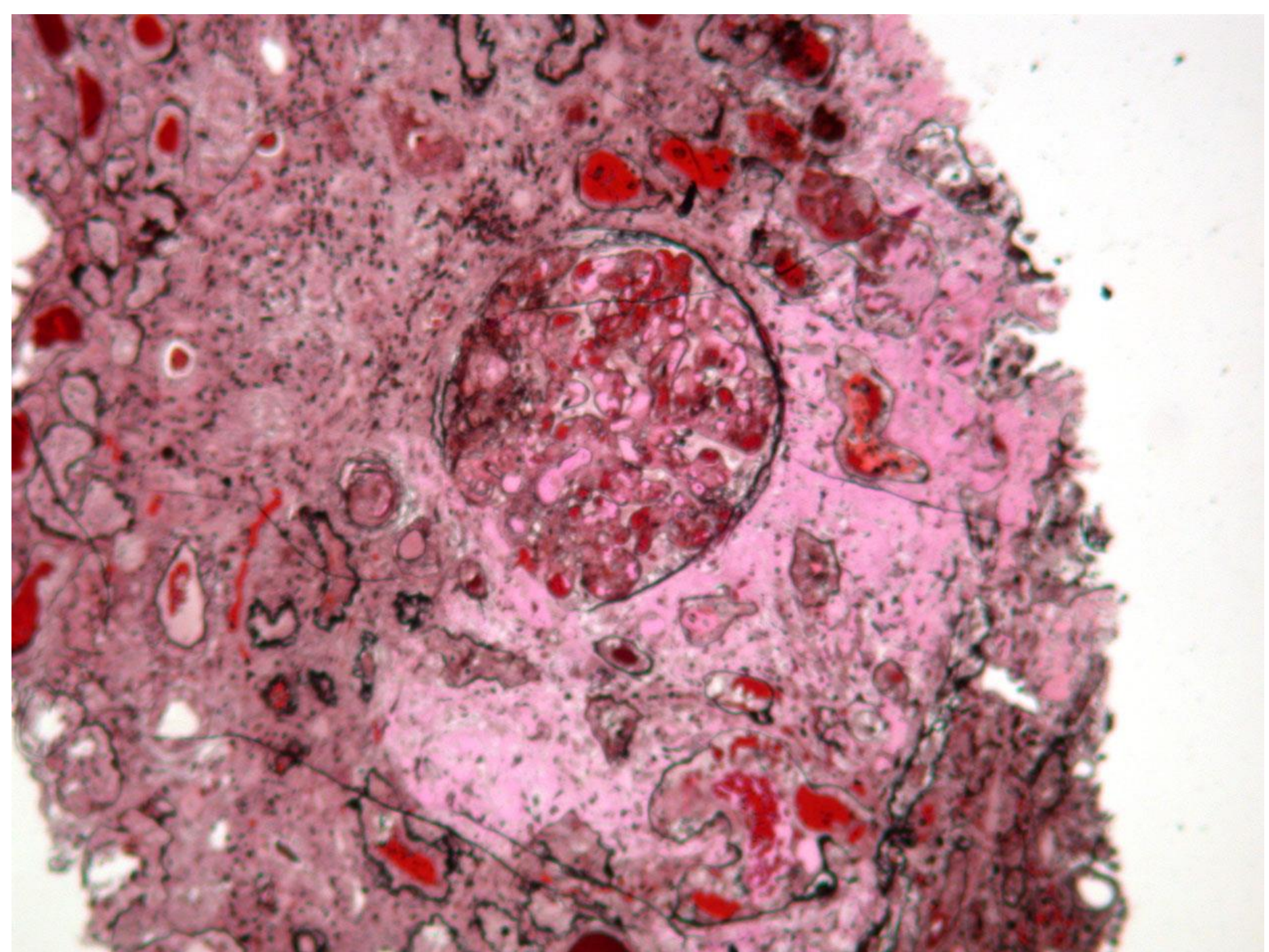
- **Forme artériolo-glomérulaire +/- mésangiolyse**
- **Forme artérielle +/- ischémie**
- **Nécrose corticale (30% enfants- 10% des adultes)**

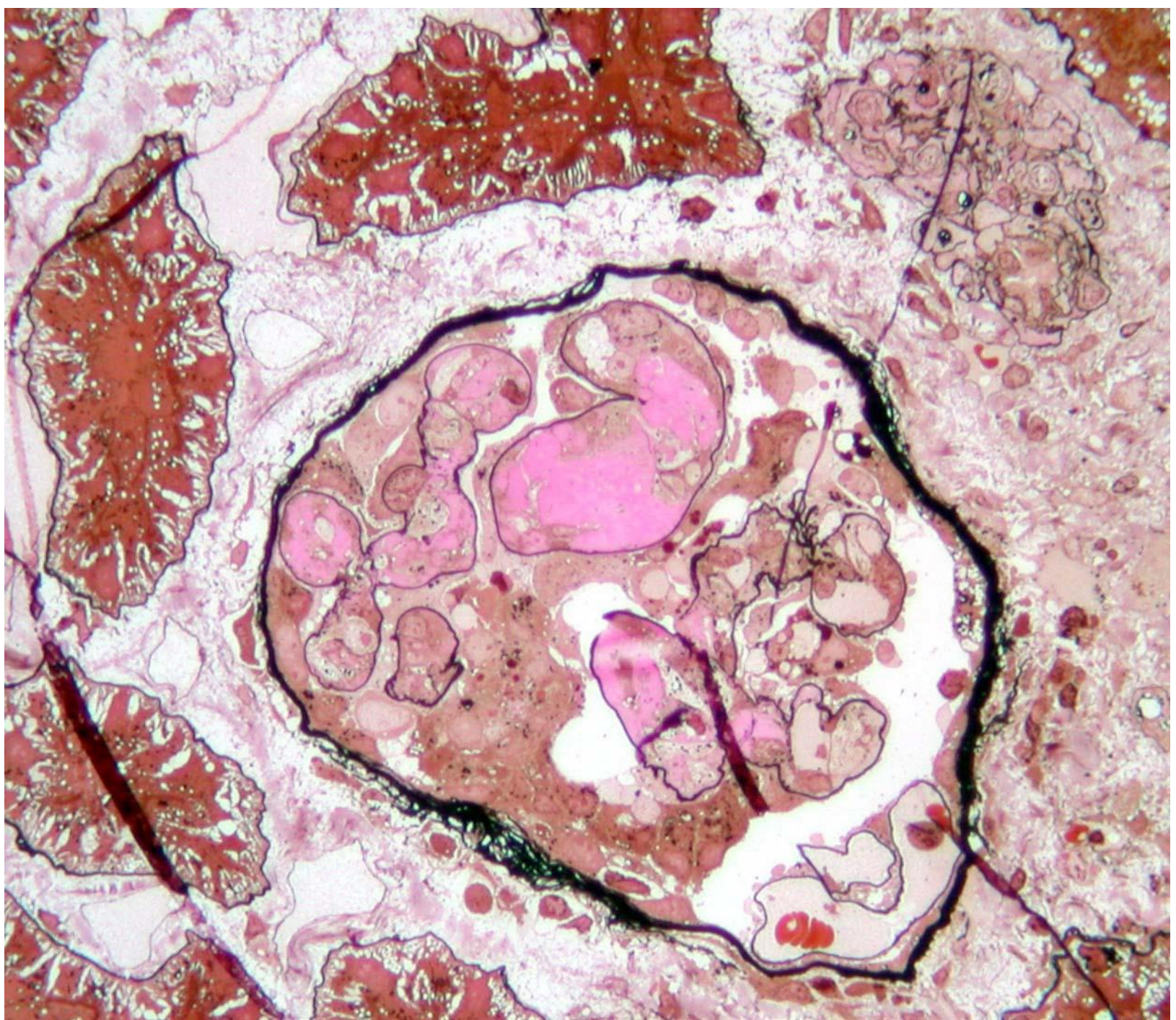




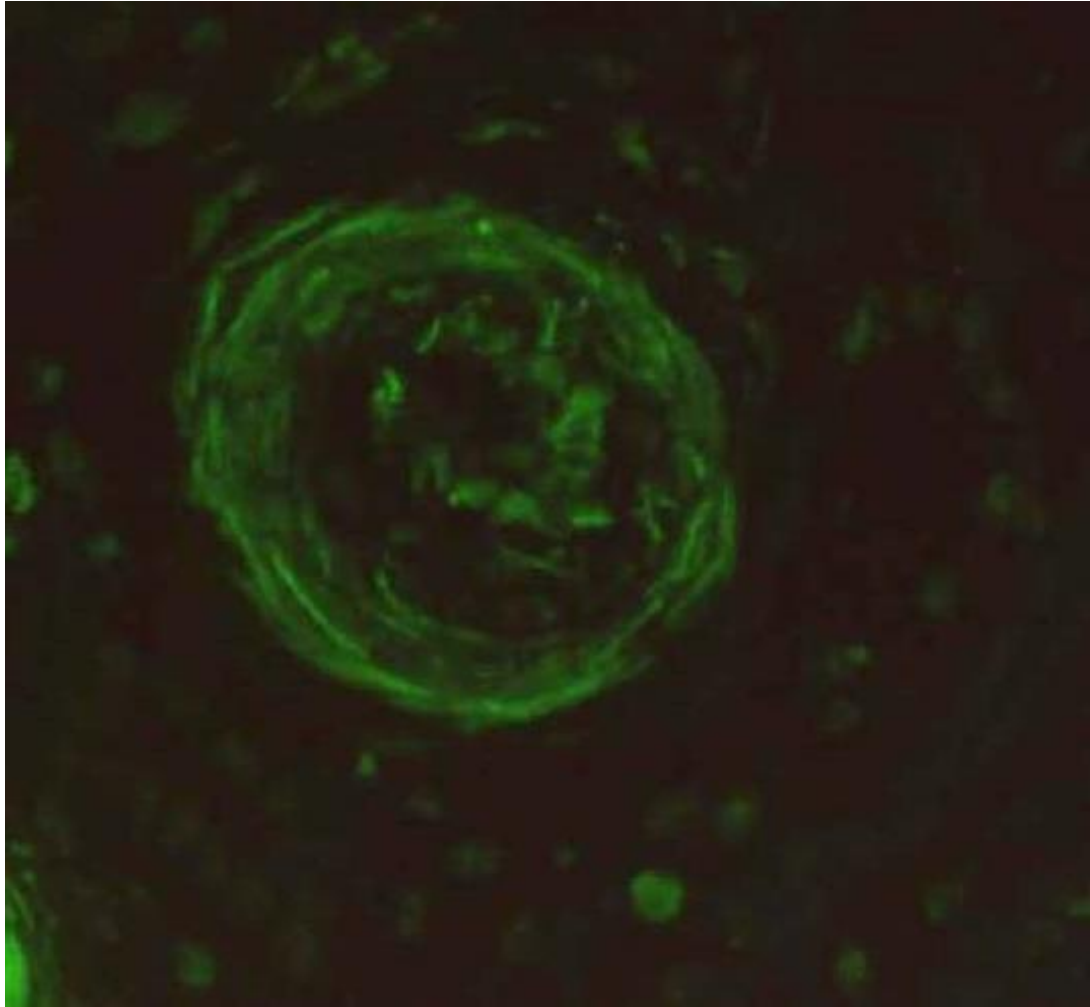
17954







C3
IgM
C1q
+/-
fibrinogène

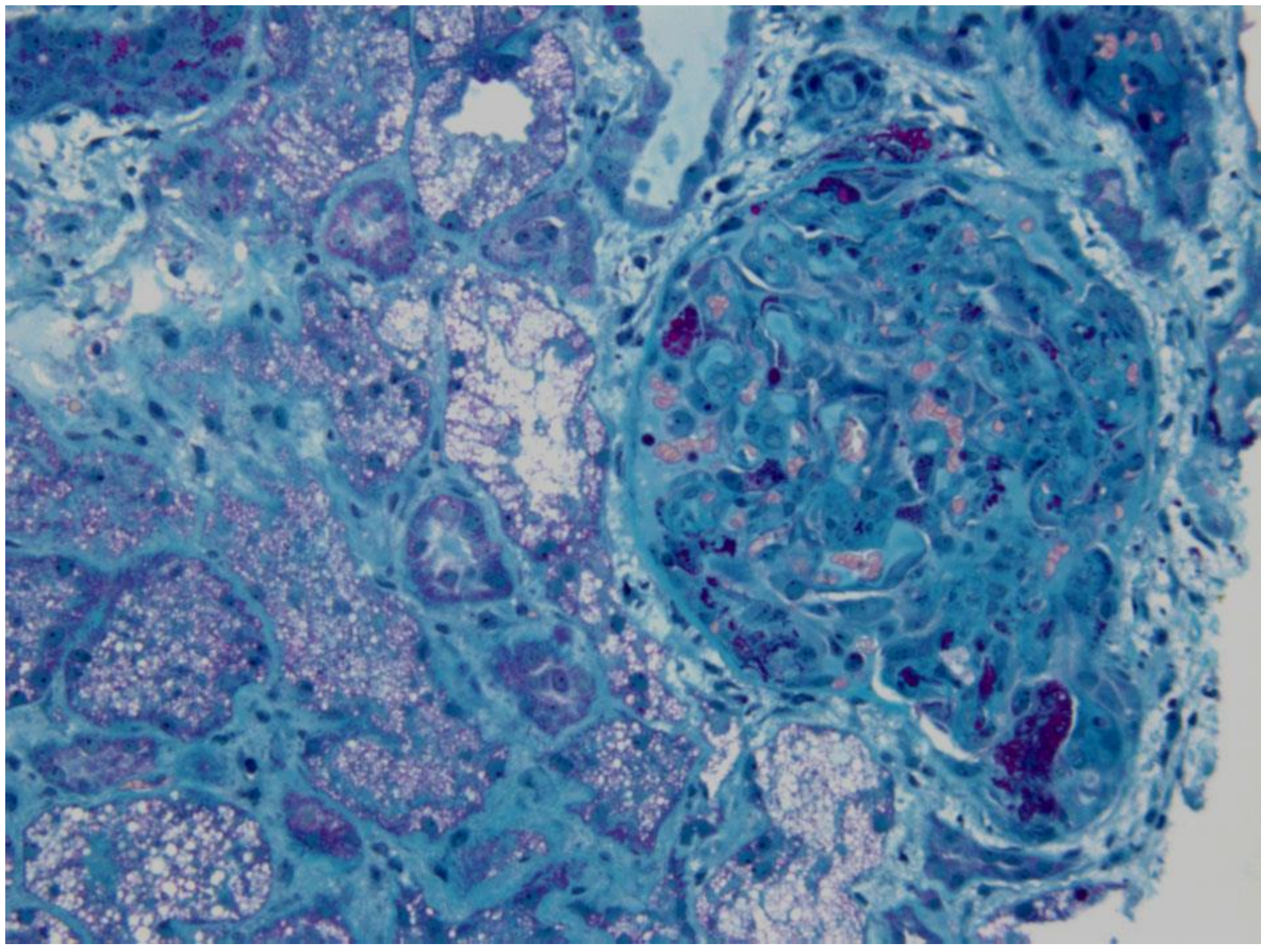


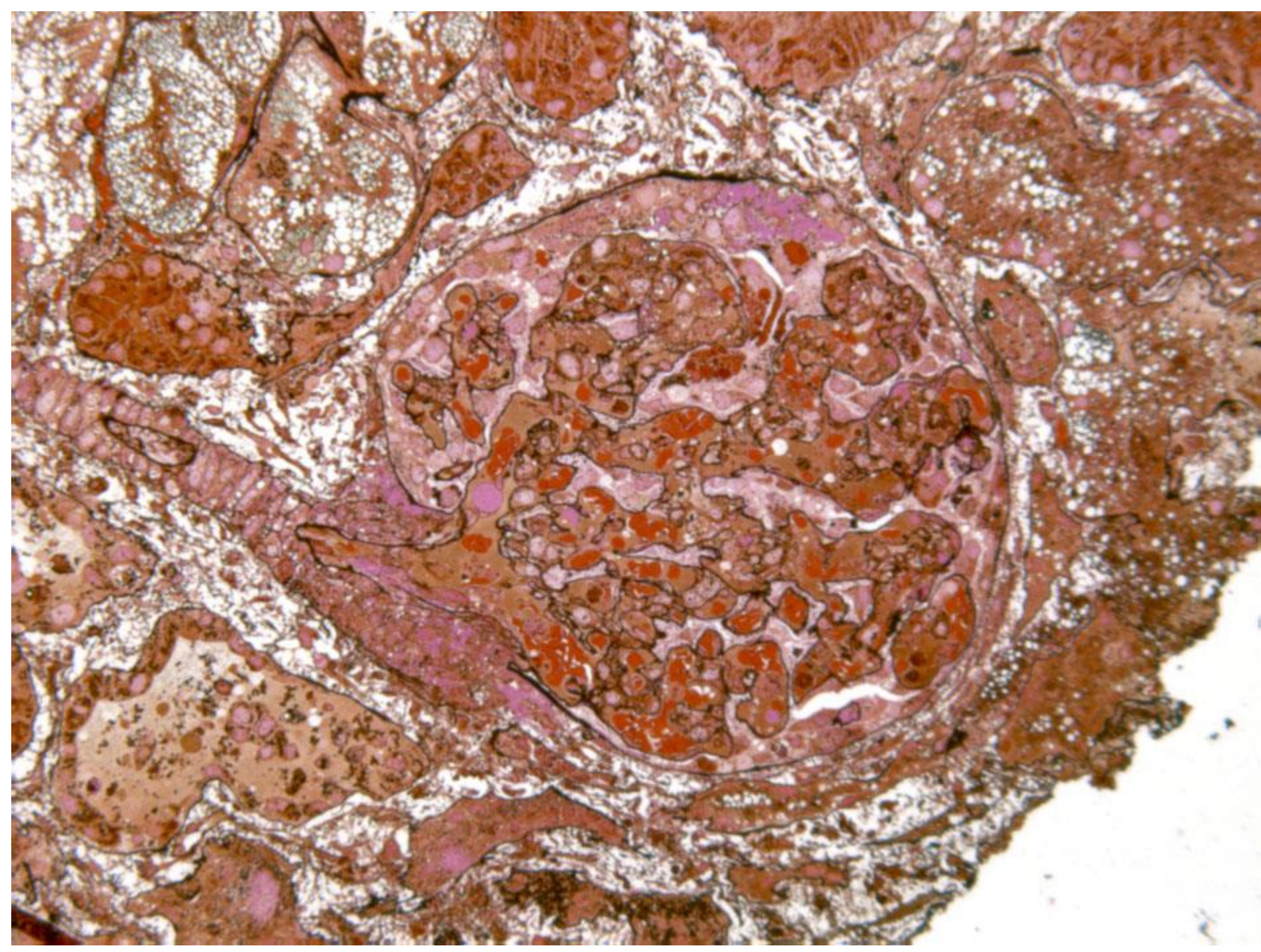
Exemple: MAT & kawasaki

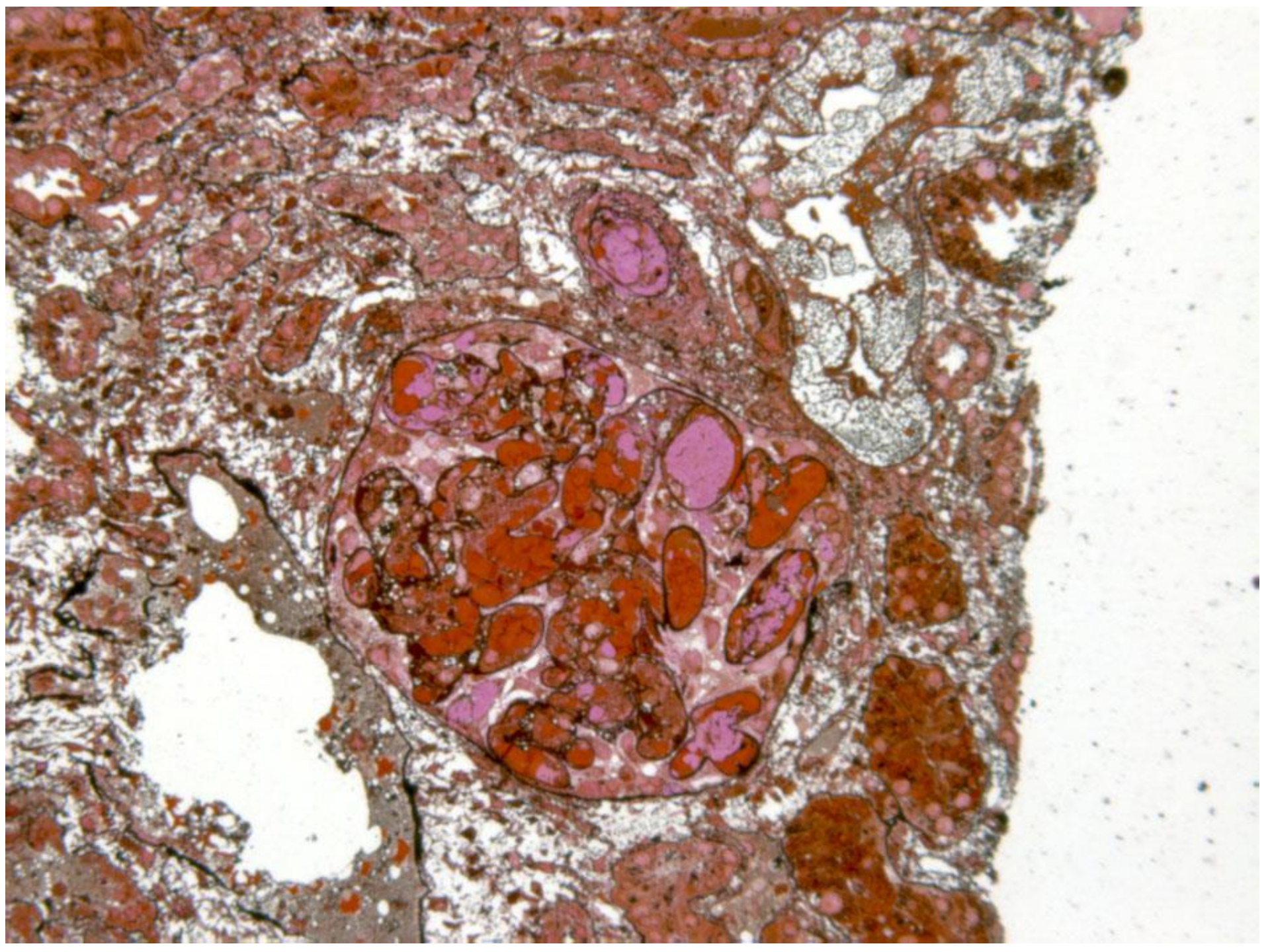
Il s'agit d'un enfant de **10 ans** hospitalisé pour prise en charge d'une maladie de **Kawasaki** atypique.

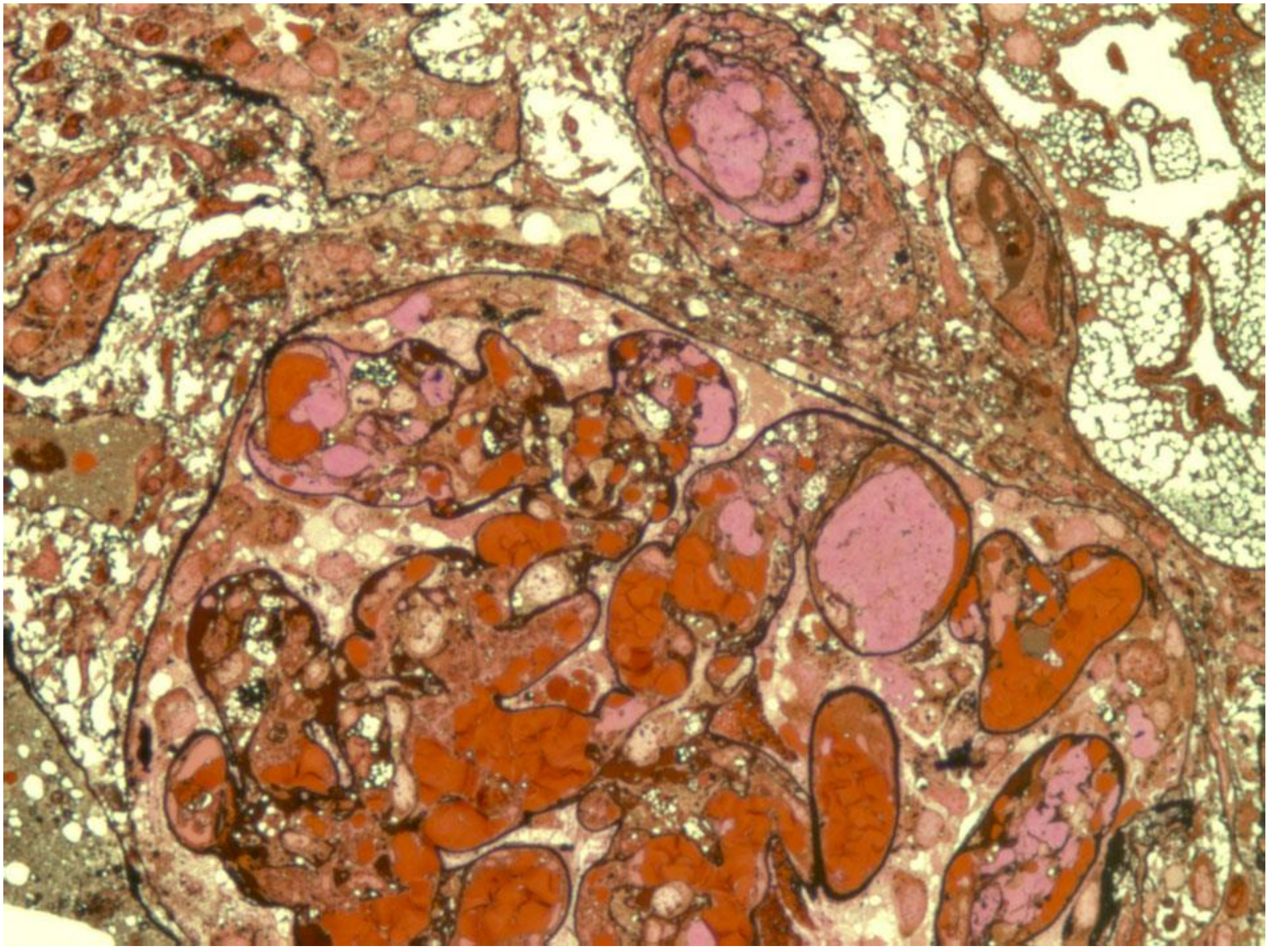
Fièvre évoluant depuis plus de 5 jours associée à une volumineuse adénopathie cervicale gauche, un érythème polymorphe, une conjonctivite sèche, un énanthème (chéilite) et une atteinte des deux coronaires (dilatations anévrismales). Le tableau clinique était compliqué par des œdèmes généralisés, une pleuro-pneumopathie hypoxiémiante, une HTA et une **oligoanurie**.

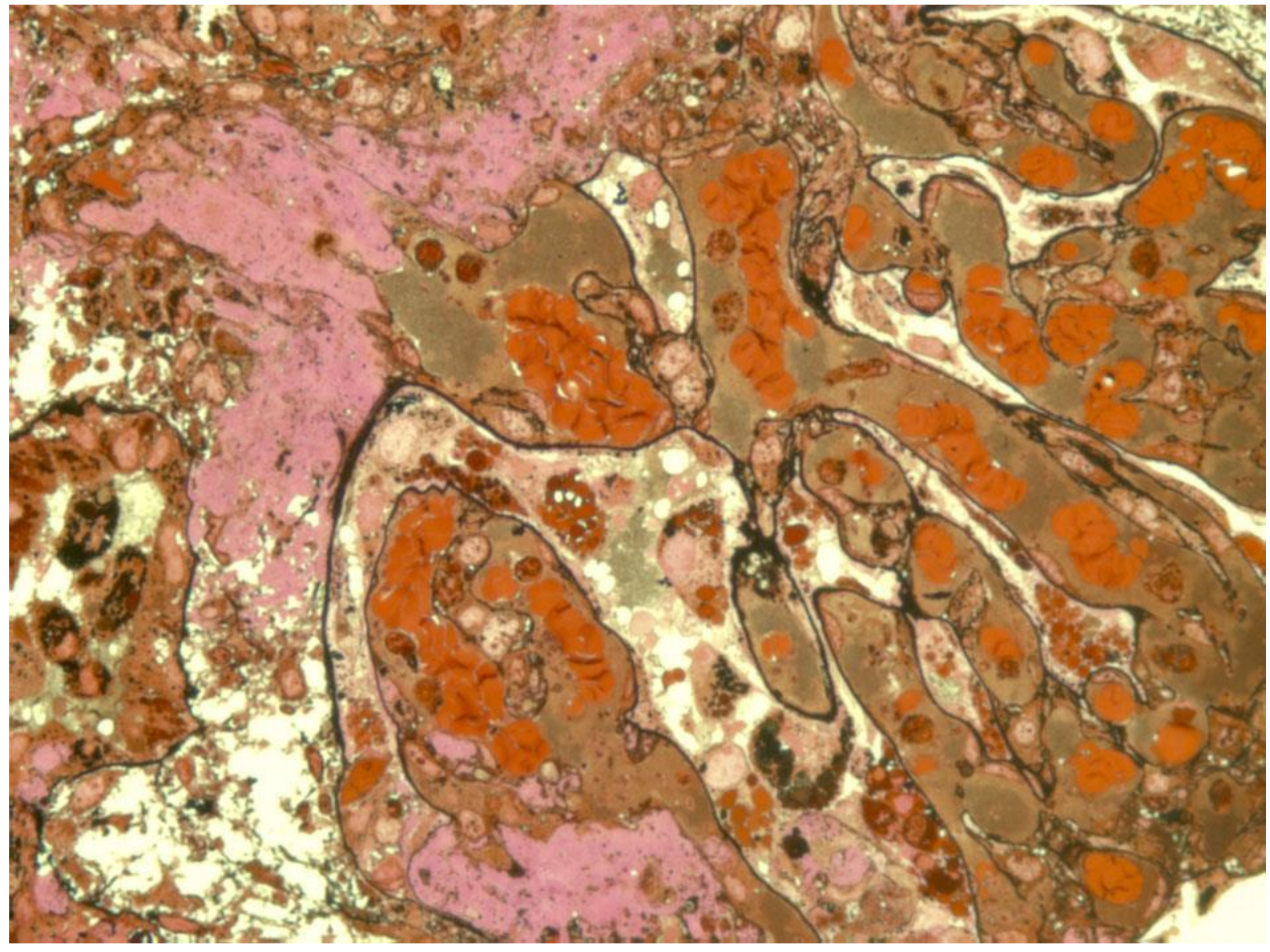
Le bilan biologique montrait un syndrome inflammatoire, une insuffisance rénale aiguë (créatinine à 200 $\mu\text{mol/l}$) et un syndrome néphrotique (albuminémie 20g/l, protéinurie 3.5g/l). Il n'y avait **aucun signe biologique d'hémolyse mécanique**. Le sédiment urinaire montrait une leucocyturie et une hématurie microscopique. L'échographie rénale retrouvait des reins hyperéchogènes.

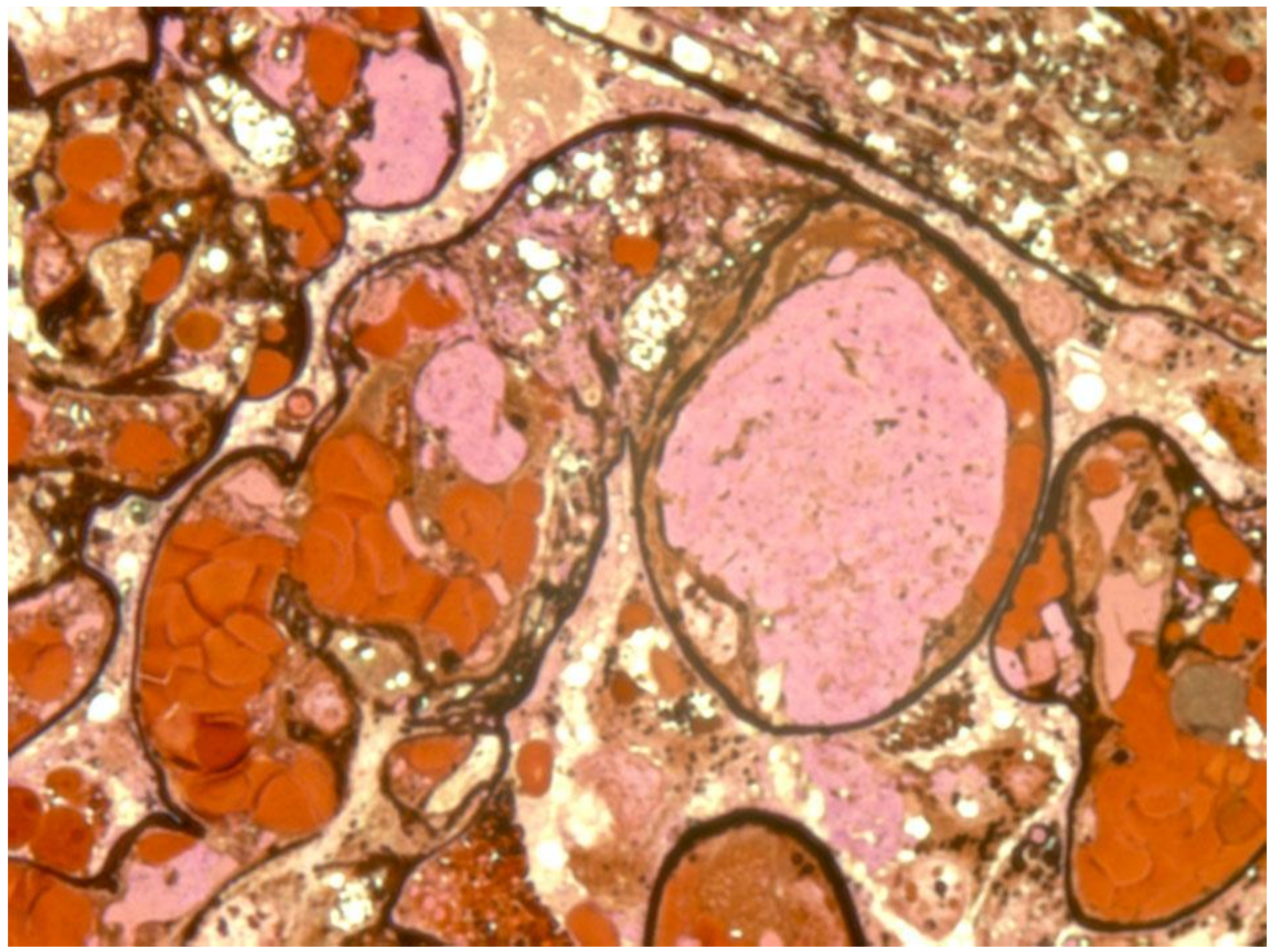


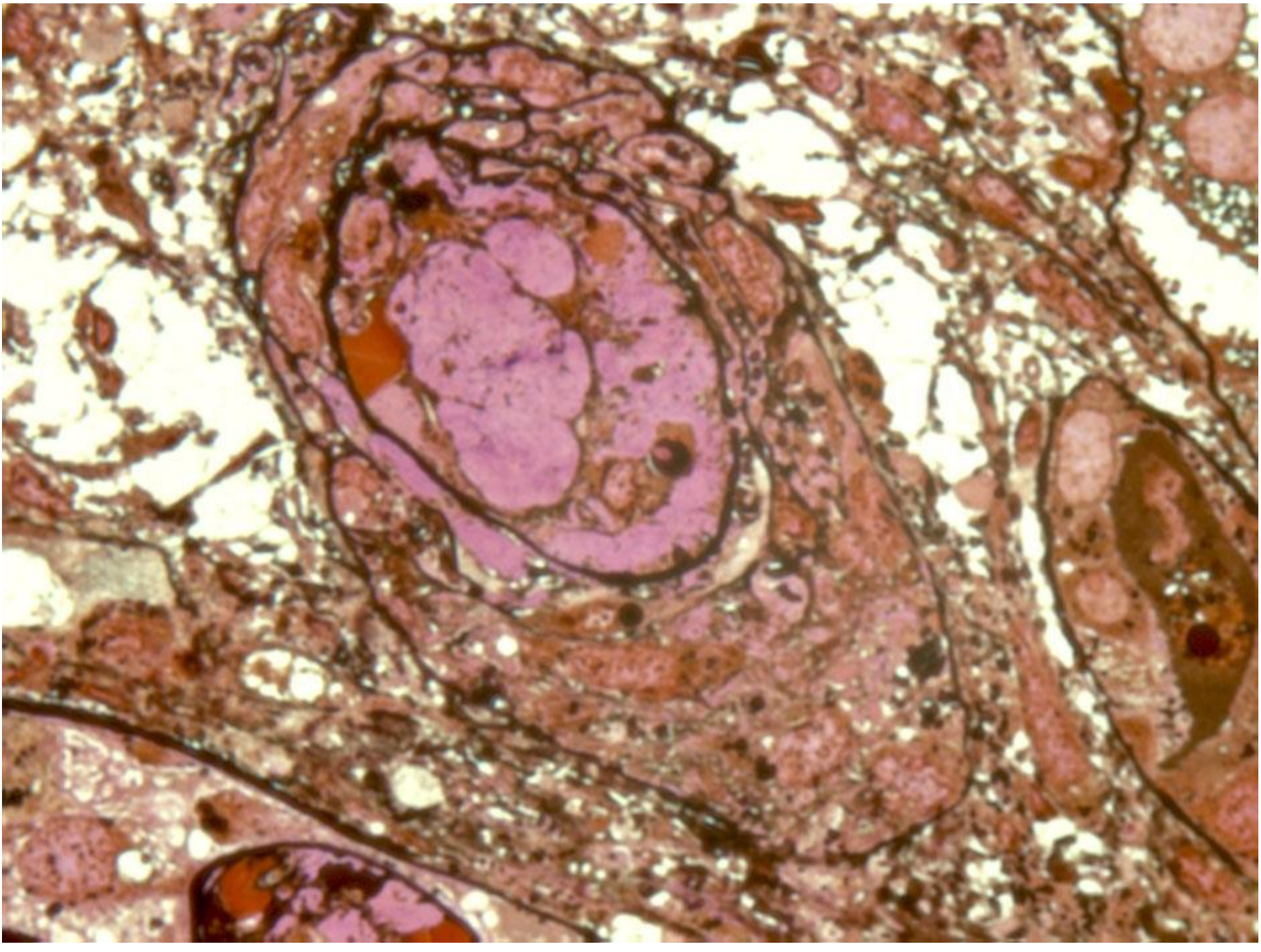








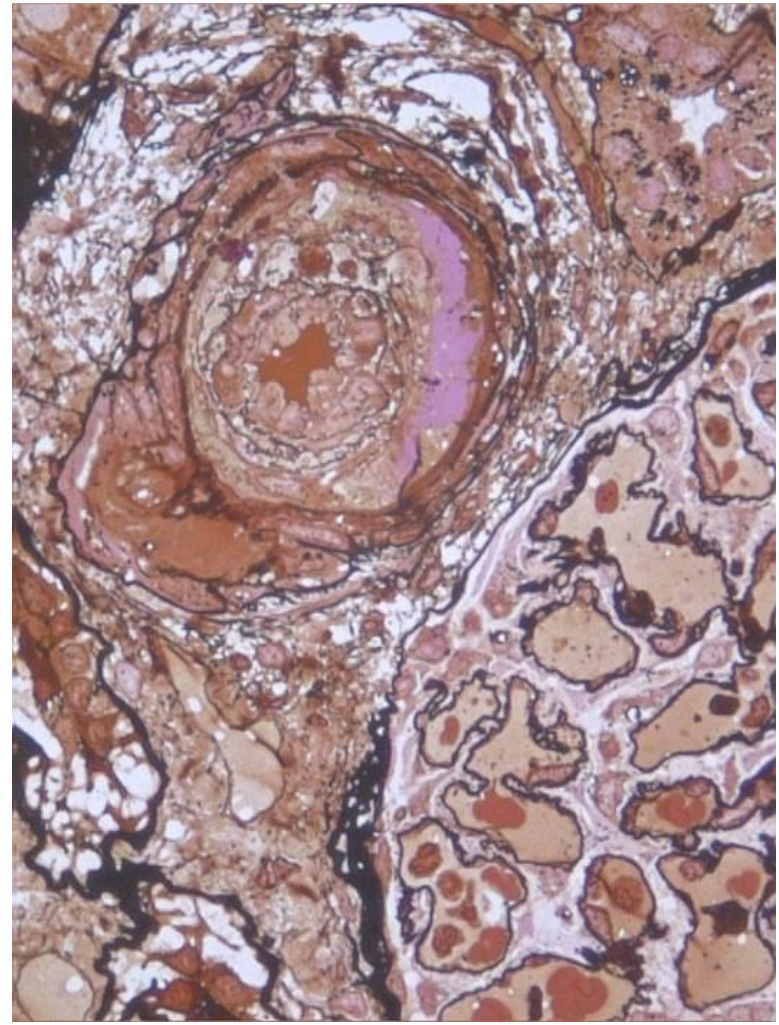
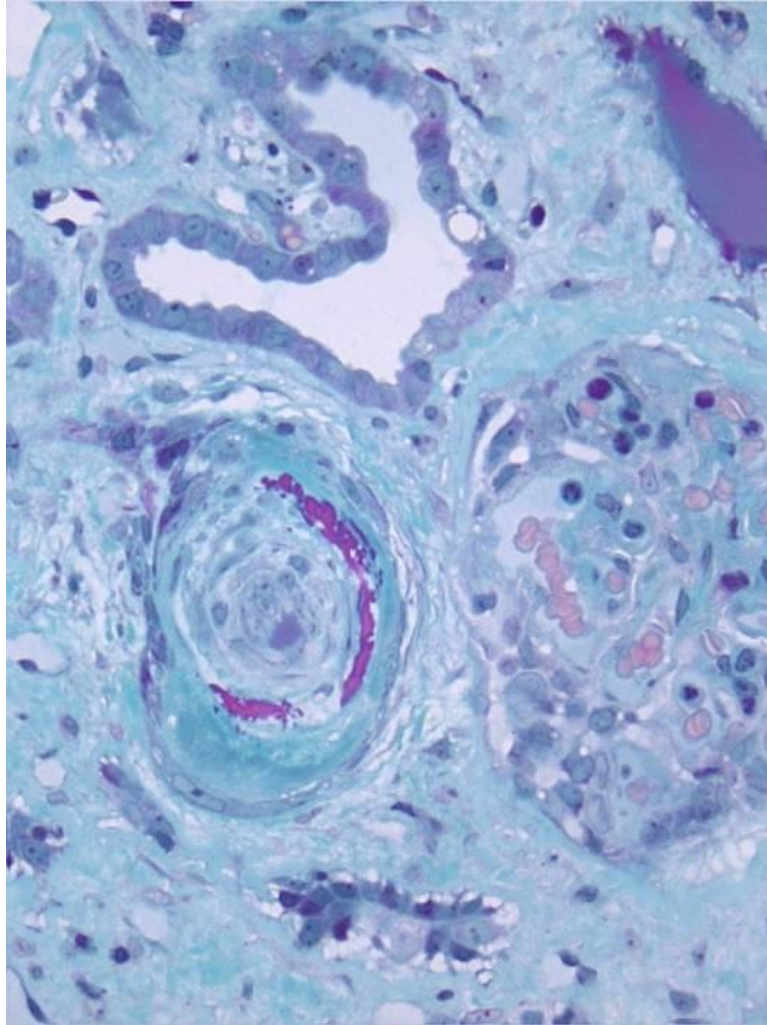


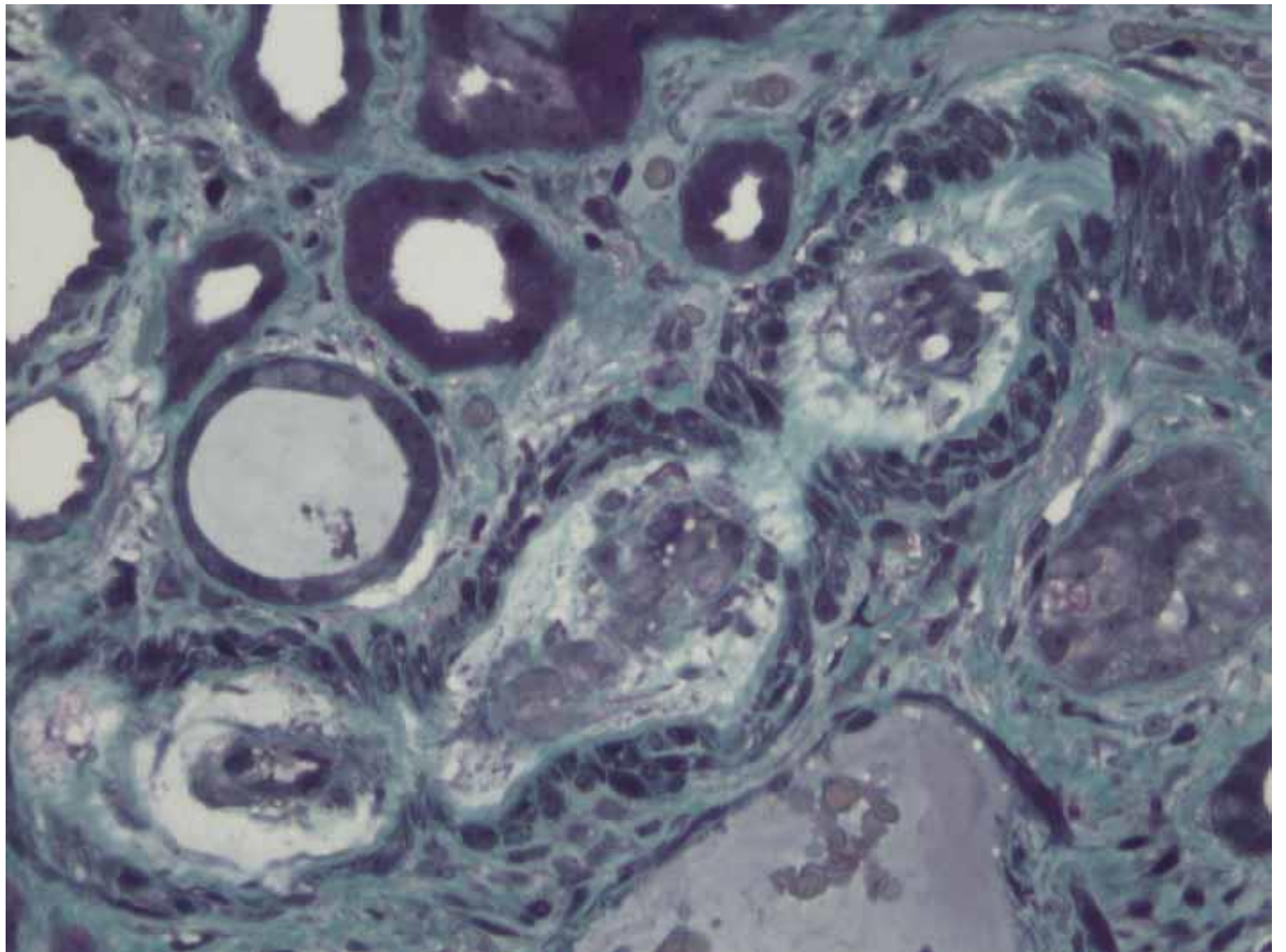


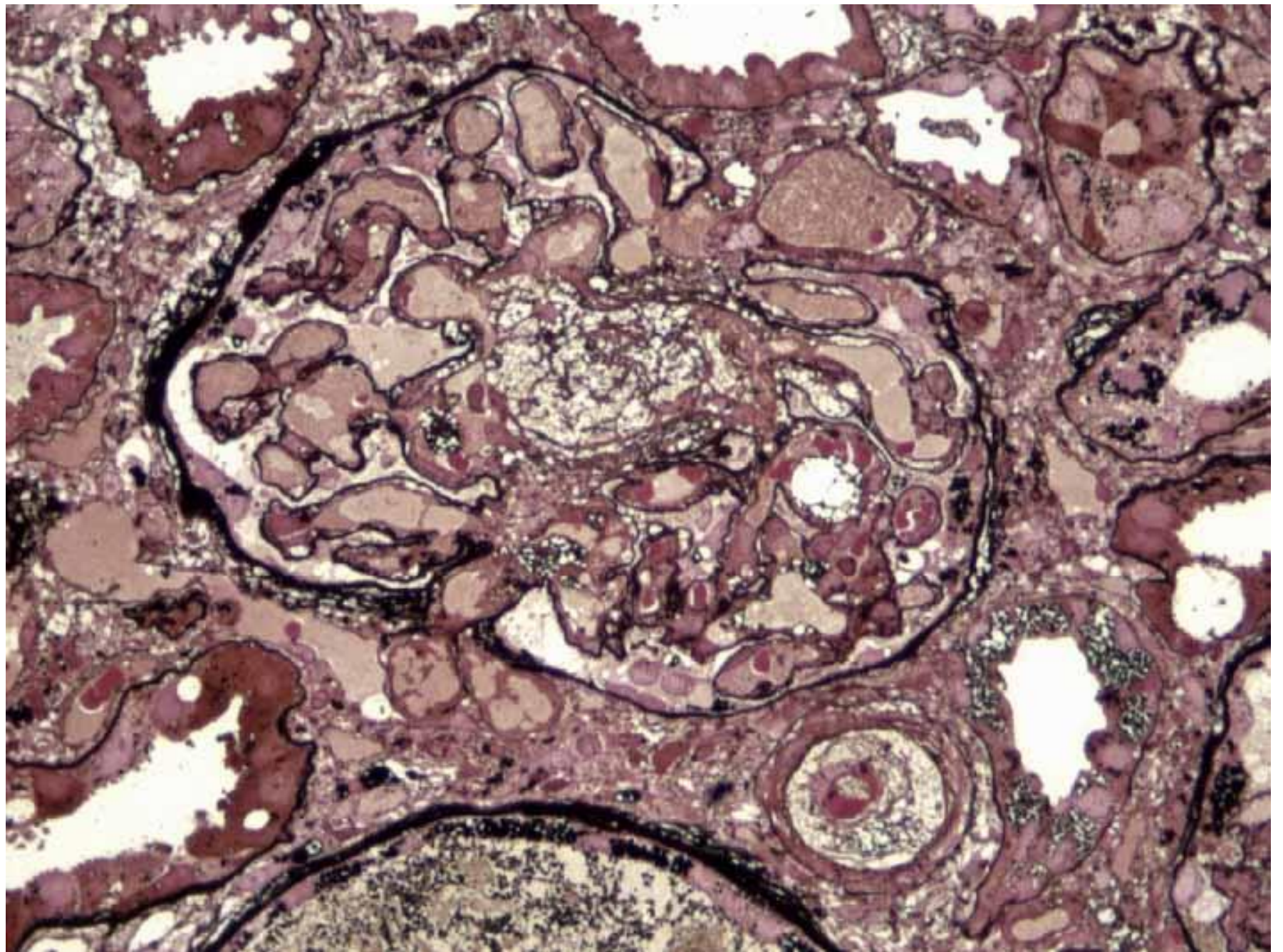
L'enfant a reçu un traitement par immunoglobulines IV, aspirine à dose anti-inflammatoire et corticoïde à 2mg/kg/j

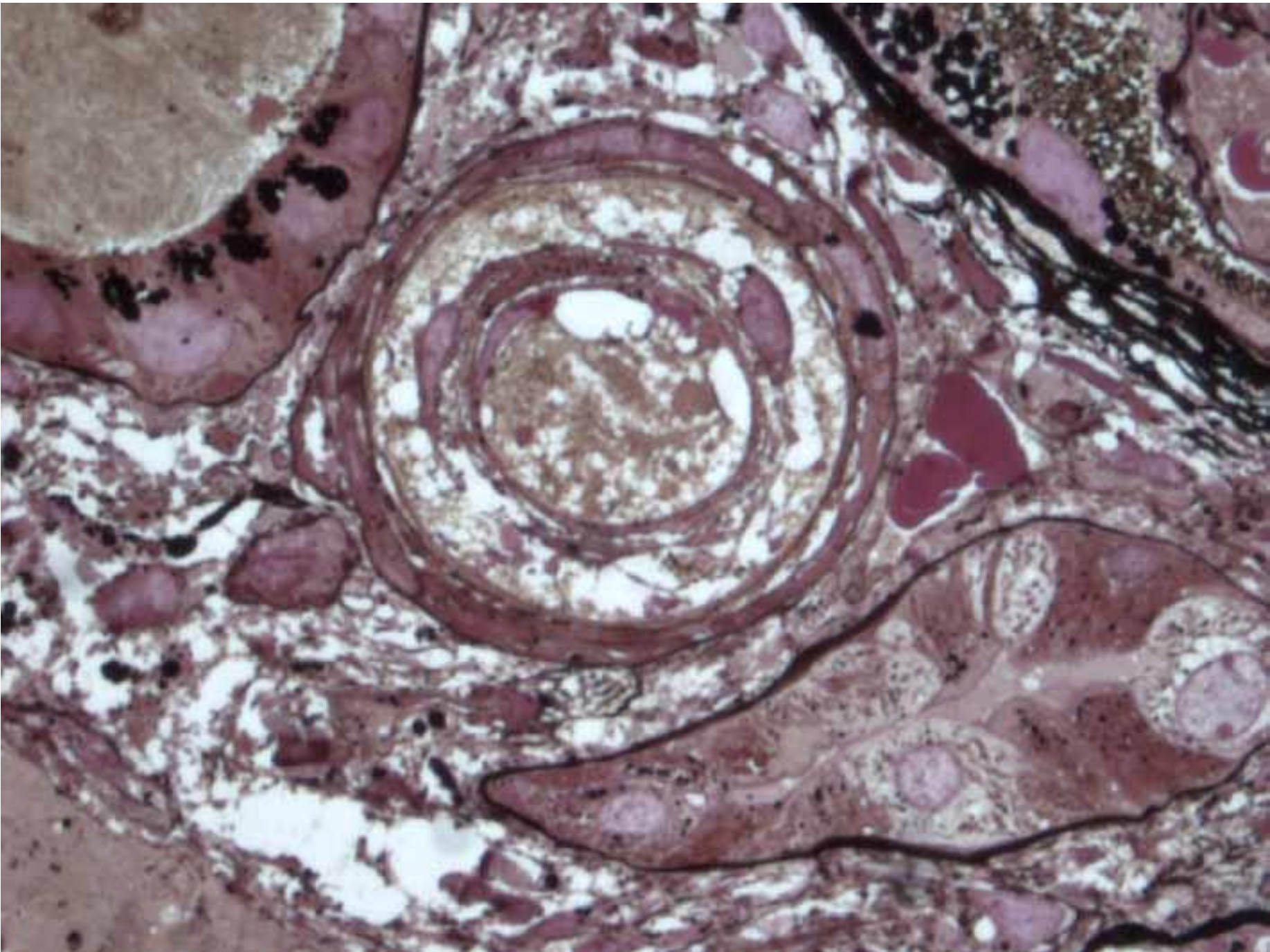
L'évolution à court et moyen terme a été satisfaisante avec normalisation de l'atteinte coronaire et de l'atteinte rénale. Une corticothérapie a été maintenue pendant une durée de 3 mois. Une ponction biopsie rénale de contrôle à 4 mois a retrouvé des lésions cicatricielles sur 15% des glomérules sans lésion de MAT.

Sclérodermie









Fréquence de la CRS

- 5 à 10% des sclérodermies.
- 80 à 90 % des cas dans les sclérodermies diffuses.
- 10% à 20% dans les sclérodermies localisées.
- 5% sans sclérodermie.

Penn H et al. QJM 2007.

Teixeira L et al. Ann Rheum Dis 2008

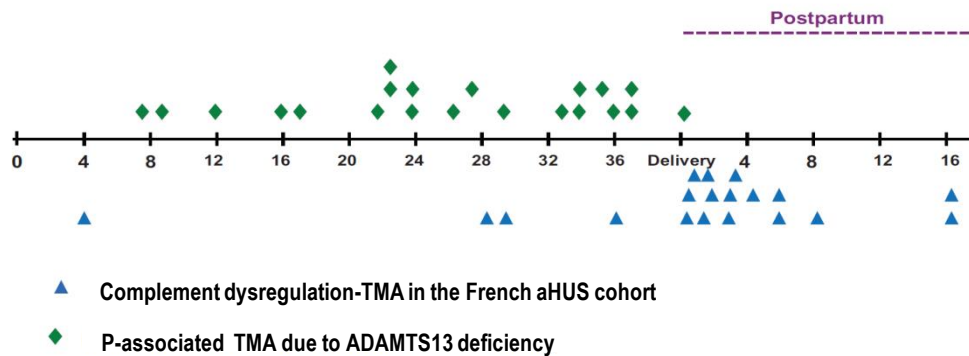
Tableau 2. Facteurs prédictifs de survenue d'une crise rénale sclérodermique

| |
|--|
| Atteinte cutanée diffuse |
| Progression rapide de l'atteinte cutanée |
| Durée d'évolution < 3 à 5 ans |
| Événement cardiaque récent |
| Péricardite |
| Insuffisance ventriculaire gauche |
| Anémie de survenue récente |
| Anticorps anti-ARN polymérase III |
| Traitement par prednisone > 15 mg/j dans les trois mois précédents |

Adapté d'après Steen 2003

MAT et grossesse

- T2-T3:
 - Lésions extra-rénales (PTT) > rein .
- Post-partum
 - De J2 à 1 mois.
 - Lésions rénales > extra-rénales.
 - Artériolo-glomérulaire +/- artérielle (HTA++++).



Merci de votre attention...

