MACRODACTYLY
case report & literature review

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המרכז הרפואי
האוניברסיטאי סורוקה

Macrodactyly or Local gigantism, has also been described under many names such as *megalodactyly*, *dactylomegaly*, *macrodactylia fibrolipomatosis* , *macrodystrophia lipomatosa* .

It is a rare congenital malformation of unknown cause, characterised by progressive enlargement of all mesenchymal elements of a digit.

definition, *n.*

Forms: α. ME–15 **diffinicioun**, etc. **diffinition** , etc., 15 *Sc.* **diffinitioun**; **definition**.

The action of defining, or stating exactly what a thing is, or what a word means.

A precise statement of the essential nature of a thing; a statement or form of words by which anything is defined.

Barsky's defined macrodactyly as overall enlargement of phalangs, tendons, nerves, vessels, subcutaneous fat, fingernails & skin.

Static macrodactyly is present at birth and the digit appears to have increased in size and grows proportionately with the rest of the body.

Progressive macrodactyly is characterized by disproportionate growth – the digit affected increases in size faster than can be attributed to normal growth patterns of unaffected digits.

Macrodactyly of the hand is much more common then in the foot.

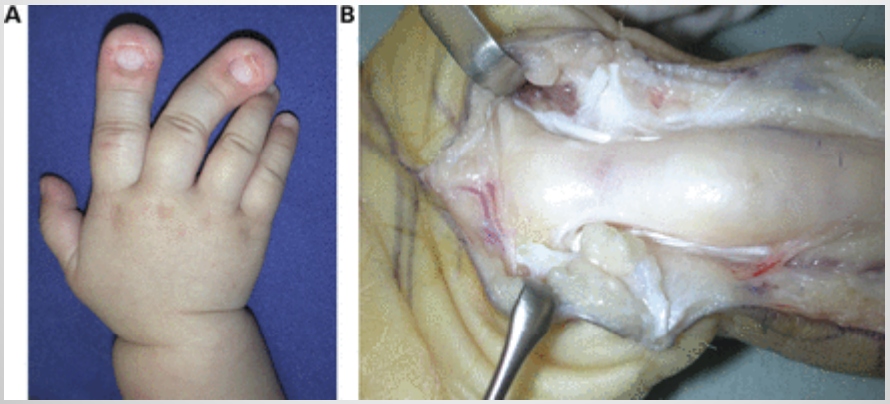
Due to the rarity of the condition, the prevalence is unknown but predominantly associated with males more than females.

Since the first description by Annadle in 1866,

167 cases were published by 1998, two thirds appeared in the hand.

Mostly single digit is affected, and less common two adjacent digits.

The index finger is the most affected one, followed by the long, thumb, ring, and little fingers.



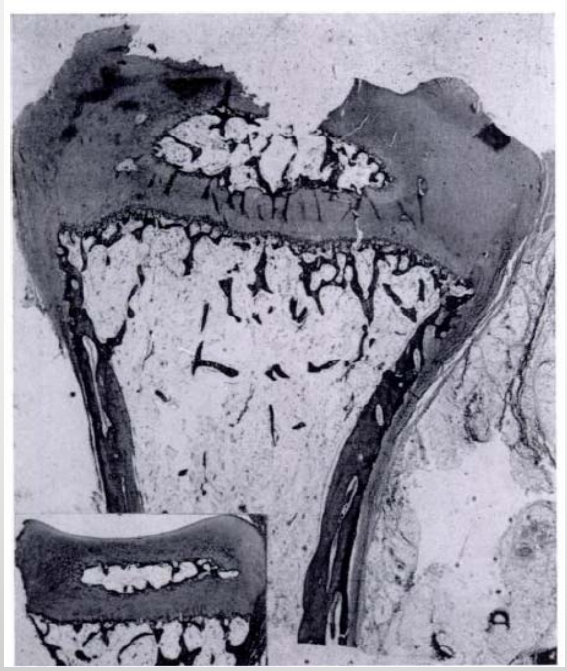
Barsky (1940'S) *postulated that an* interaction of two or more intrinsic and extrinsic agents (genetic and/or environmental) are responsible for the deformity.

He further conceptualised that some disturbance of growth limiting factor occurs in the affected local area, which continues to increase in size.

Brooks and **Lehman** (1924), **Moore** (1942), & **Inglis** (1950) , all *believed that neurofibromatosis (via neurogenic disorder mechanism)* is the main cause of rapid localized overgrowth.

However, **Thorne *et al*** *(1968) in their study of thirty* cases did not find any evidence of neurofibromatosis.

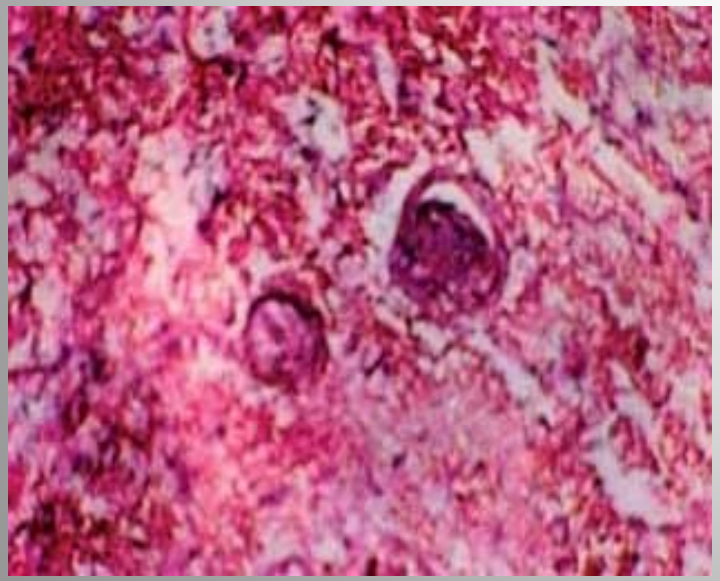
Authors like Ben-Bassat *et al (1966)* *El-Shami* and Dennyson *et al* added the possibility of abnormal mechanism in the affected involved bone.

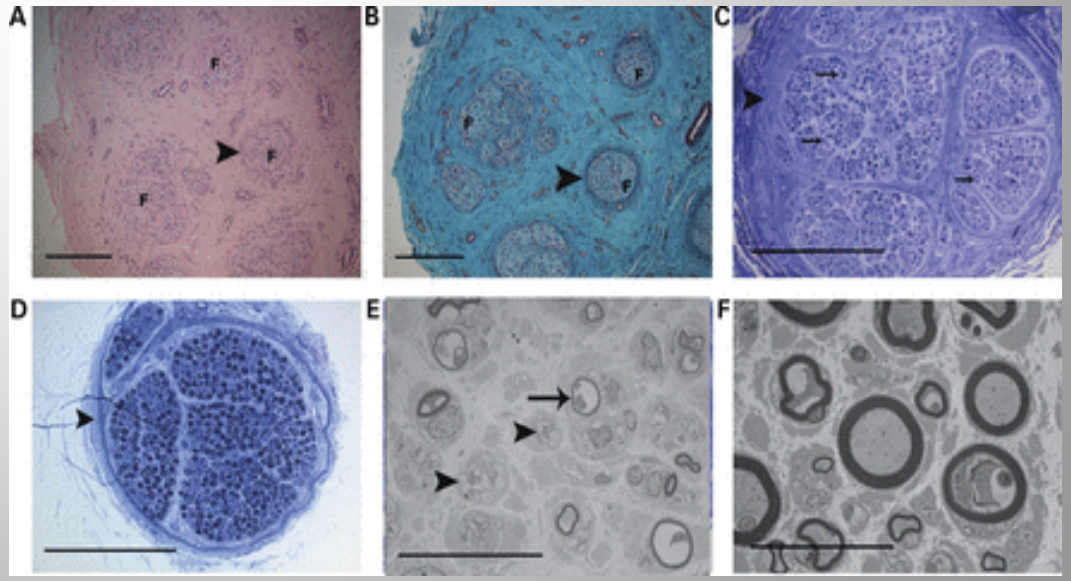


Histopathologic findings suggest that:

In **pedal Macrodactyly** excessive proliferation and accumulation of fibro-fatty tissue is the basic lesion.

In **Macrodactyly of the hand** involvement of nerves might be the fundamental lesion -> hypertrophied and tortuous digital nerves.





diff dx:

AV Maleformation

Hemangioma

Lymphangioma

Plexiform neurofibromatosis

Fibrous dyplasia

lipoma

Ollier’s disease

Klippel-Trenaunay, Proteus, Maffuci syndromes, tuberous sclerosis

Surgical options vary, and include but are not limited to:

Debulking of the soft tissue and fat.

Shortening procedures (Barsky and Tsuge).

Ray resection.

Epiphyseal Arrest.

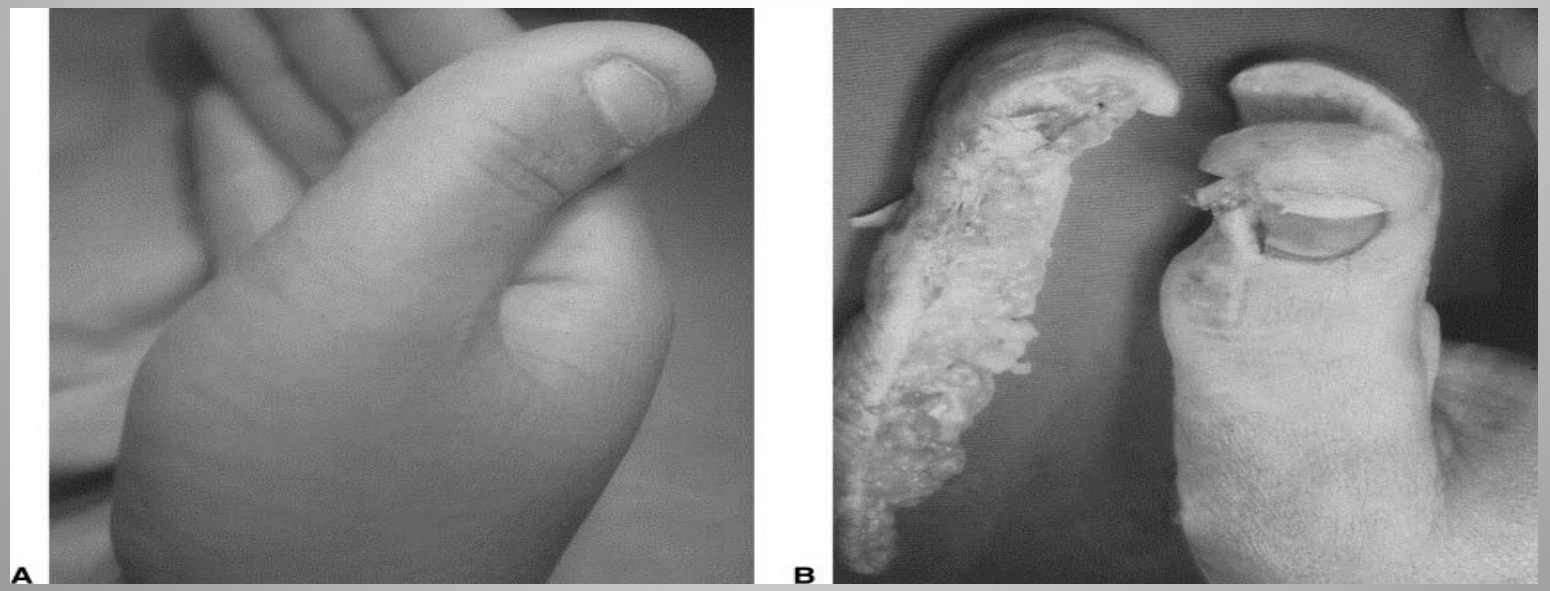
Other surgical options include amputation and wedge osteotomies.

A.H.G. - CASE REPORT

49 years old Bedouin male was born with a primary congenital macrodactyly of his Rt. hand thumb.

First time to be very Prominent by the age of 1 year.

Soft tissue debulking was performed at his early childhood but re growth of the surrounding tissues has occurred.



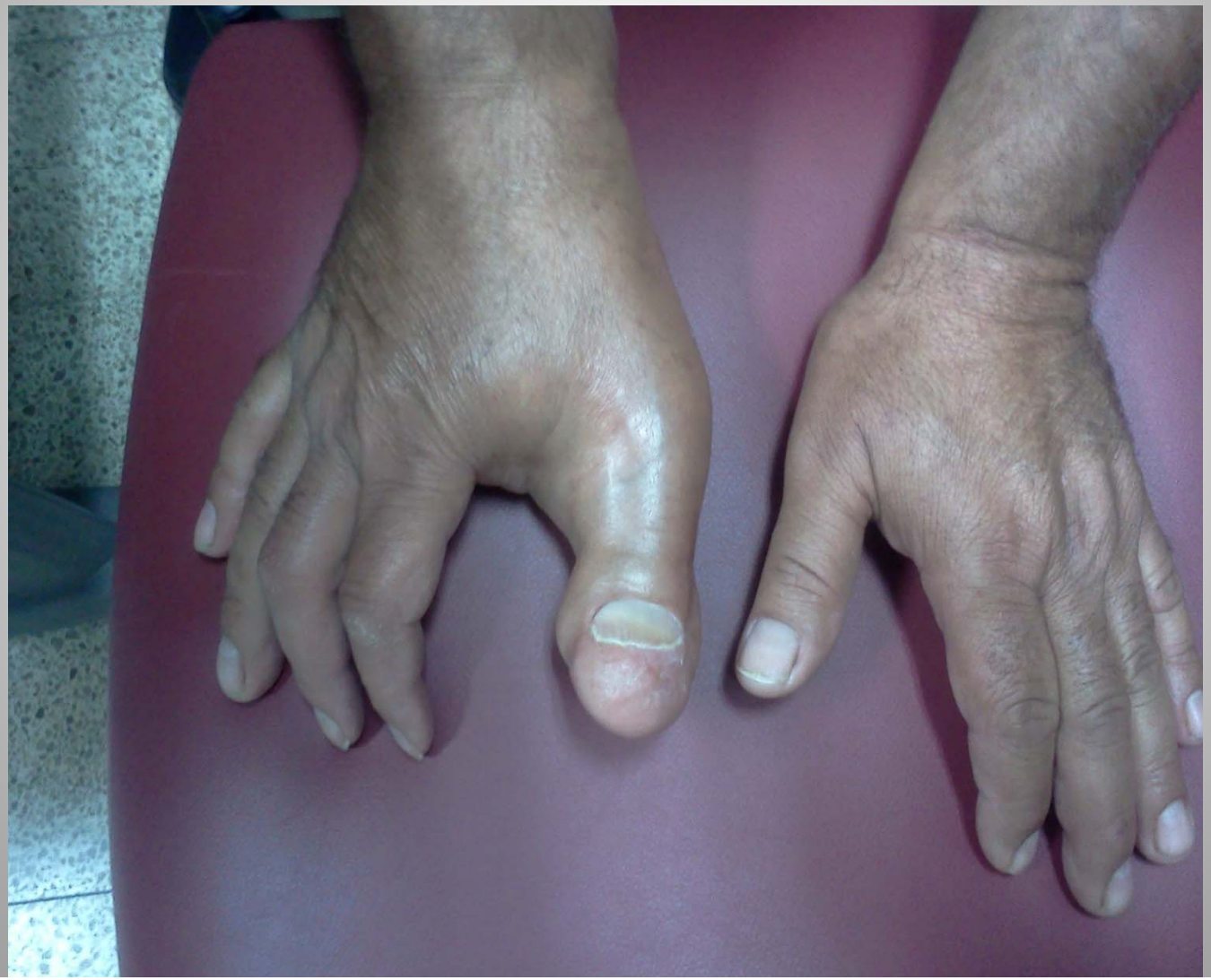
At the age of 12 years he was operated again.

The surgical procedure included:-

- Excision of fibrofatty tissue.
- Transfer of EPB to opponense to provide opposition.
- Arthrodesis of the DIP & PIP in order to cause epiphyseal - arrest.

Since then he was followed up in some medical centers without further treatment.

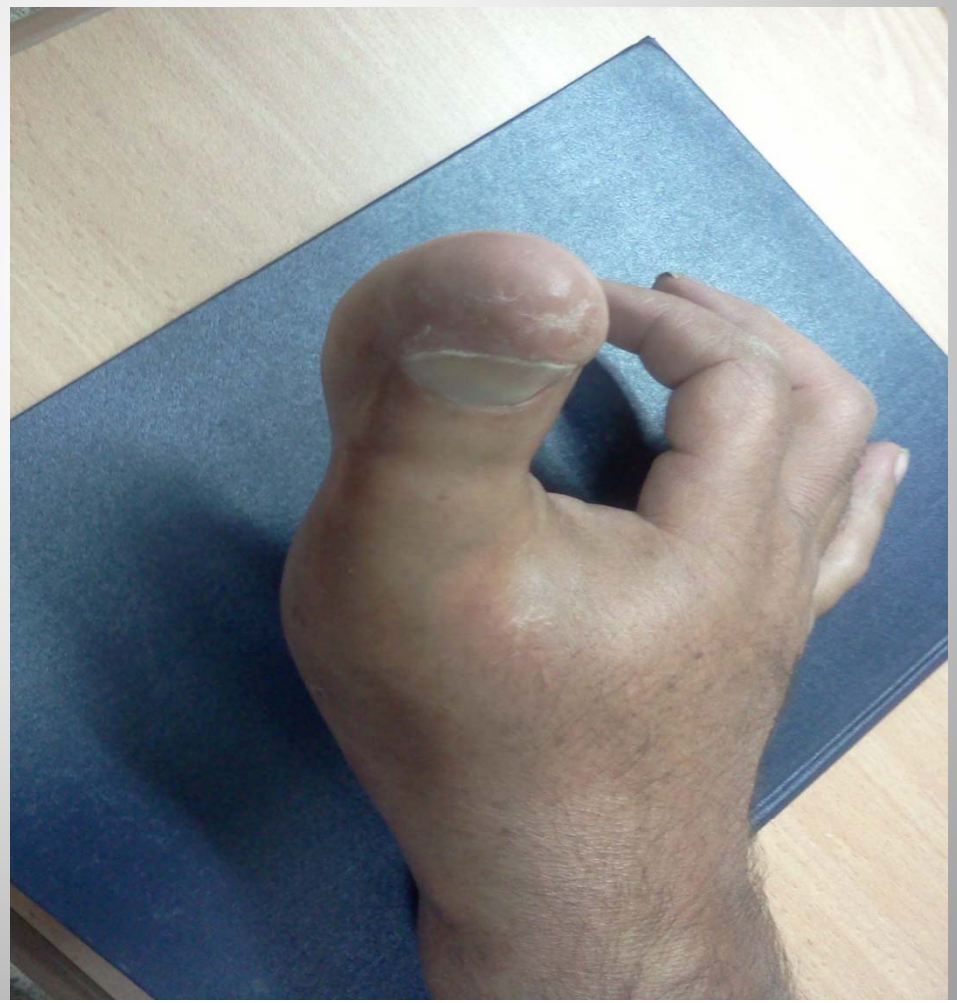
Lately he appeared at our clinic complaining about constant pain in the thumb area and deteriorating sensibility of the 1st to 3rd fingers.



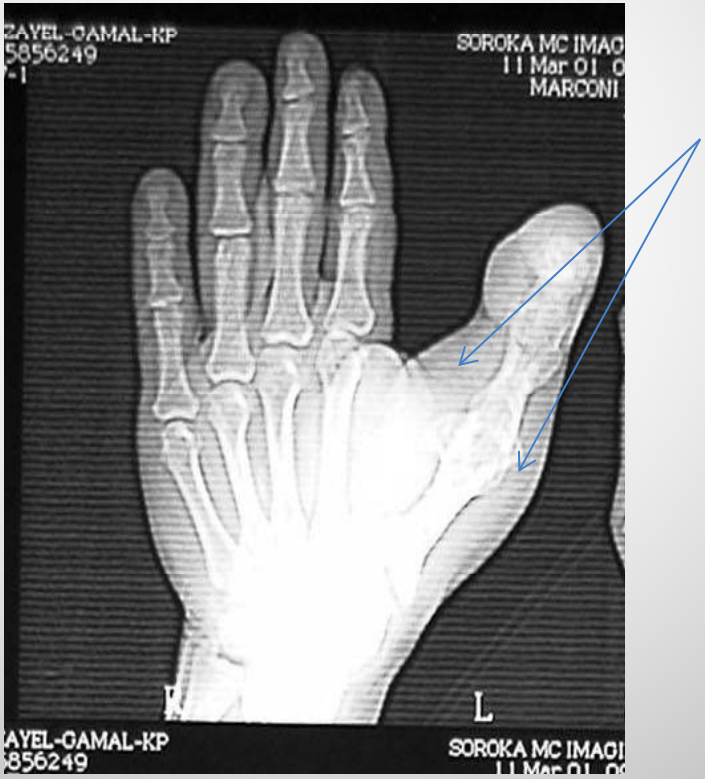
On ph-ex:

Huge thumb with no active movements.

Loose of sensation at the distribution of the median nerve.



On X-RAY & CT : s/p arthrodesis of the Interphalangeal joints and soft tissue significant thickening.



On EMG – no Motor or sensory response of Rt. Median nerve!

- FCR & Rt. APB – loose of Muscle unit.

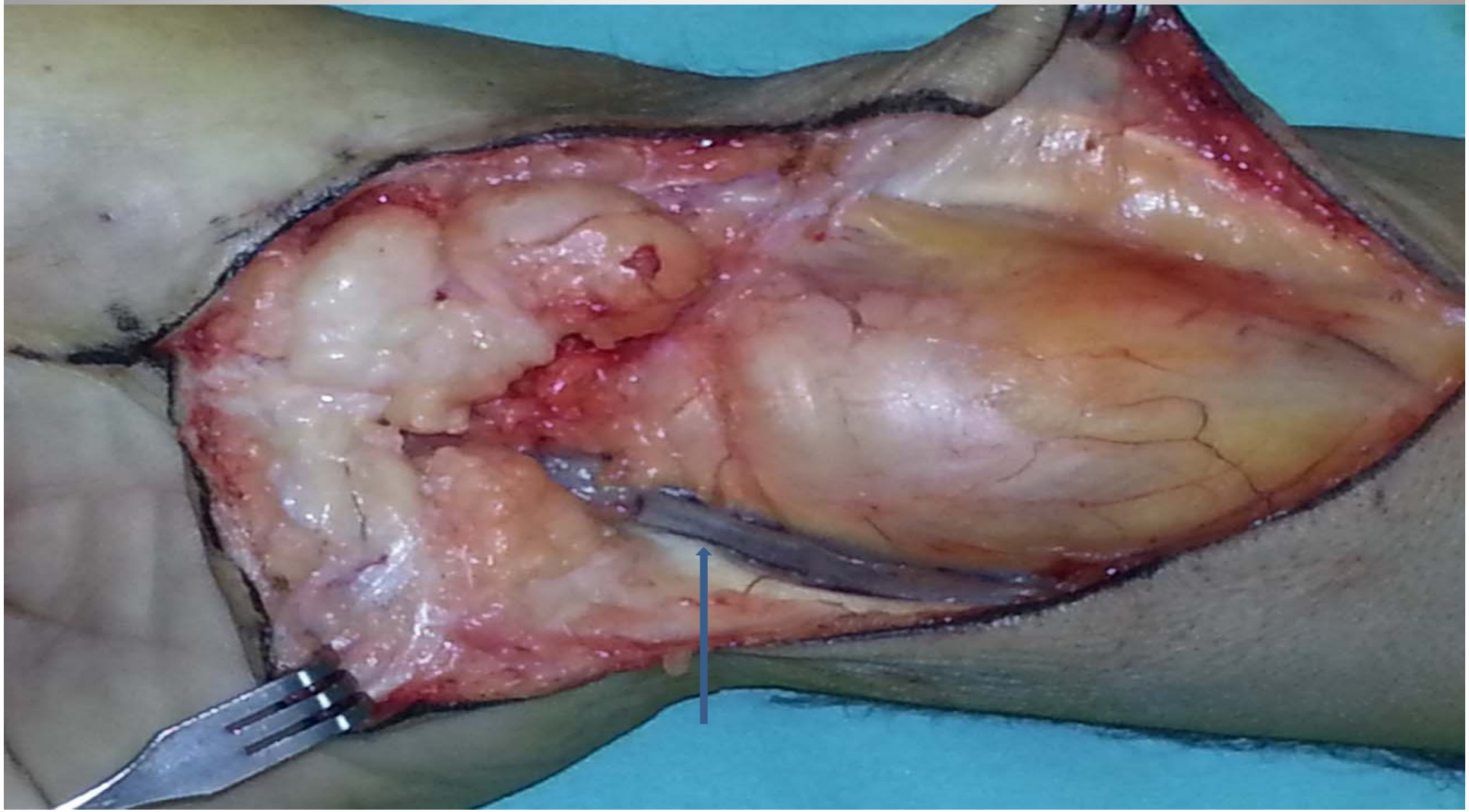
- ulnar & radial nerve – functional.

It is important to emphasize that the patient didn’t want to have any major surgery.

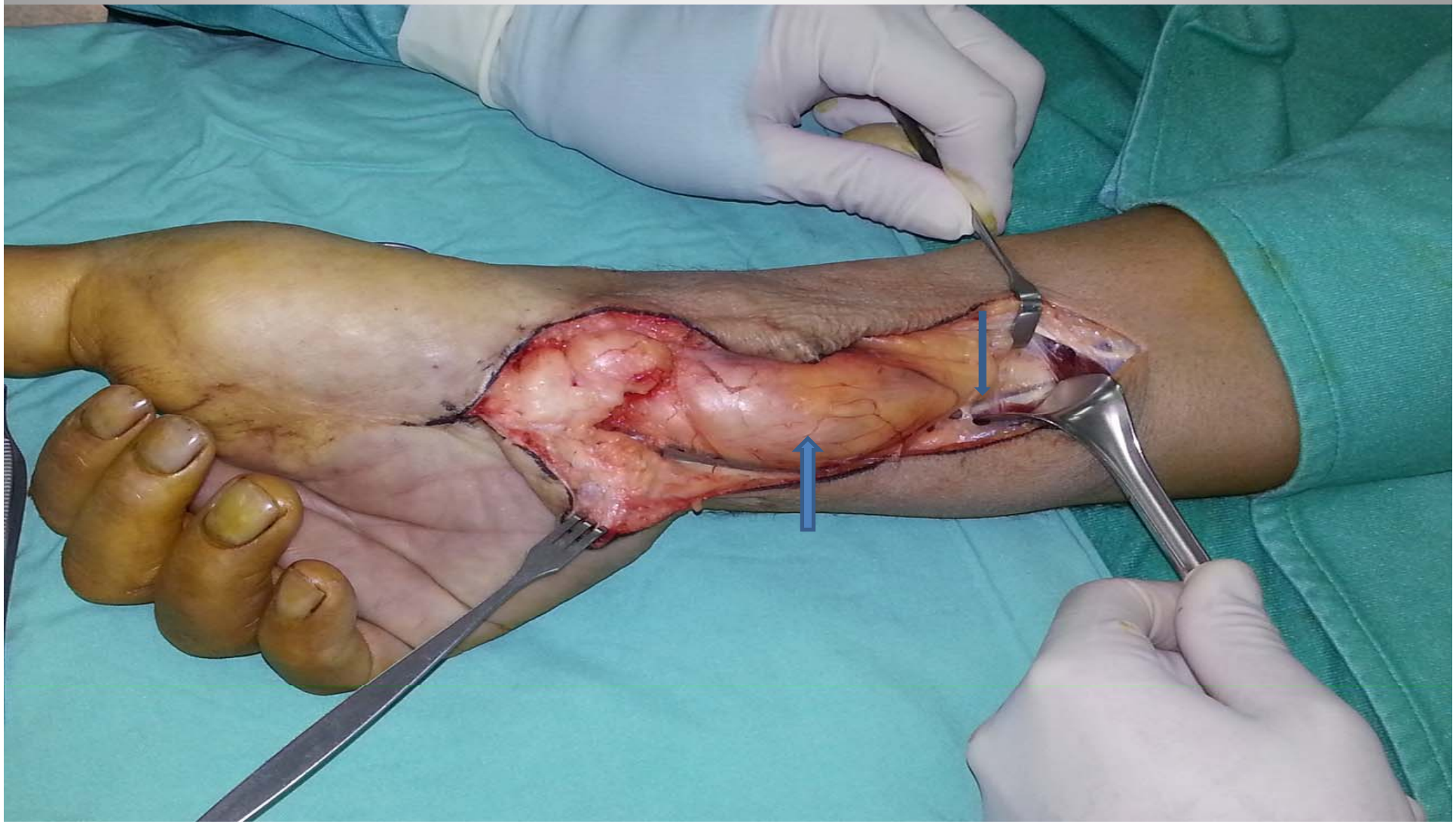
He only wished relieving of the pain & gaining some senstion back to his fingers.

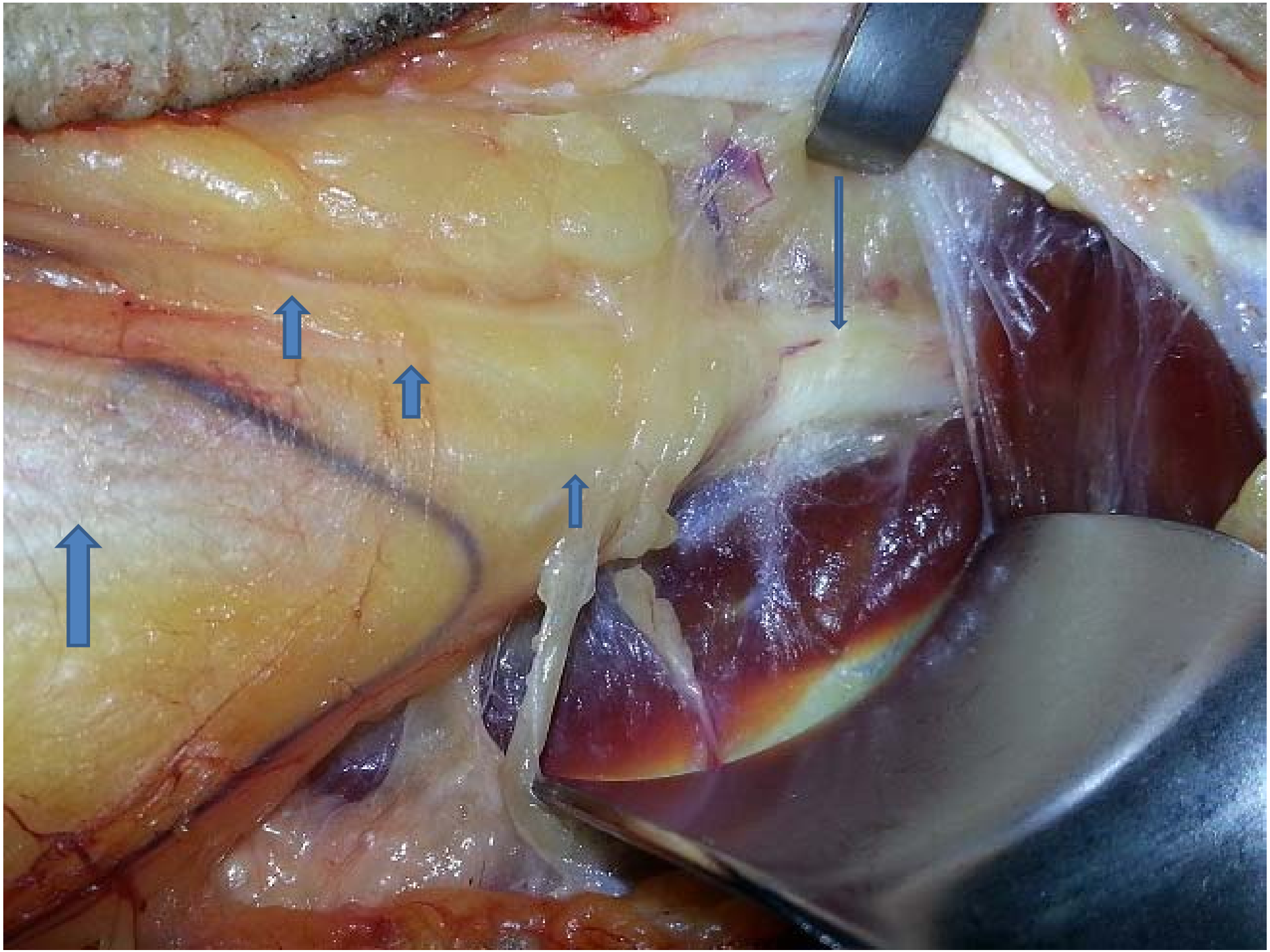
Having that in mind we planed him for explorative operation.

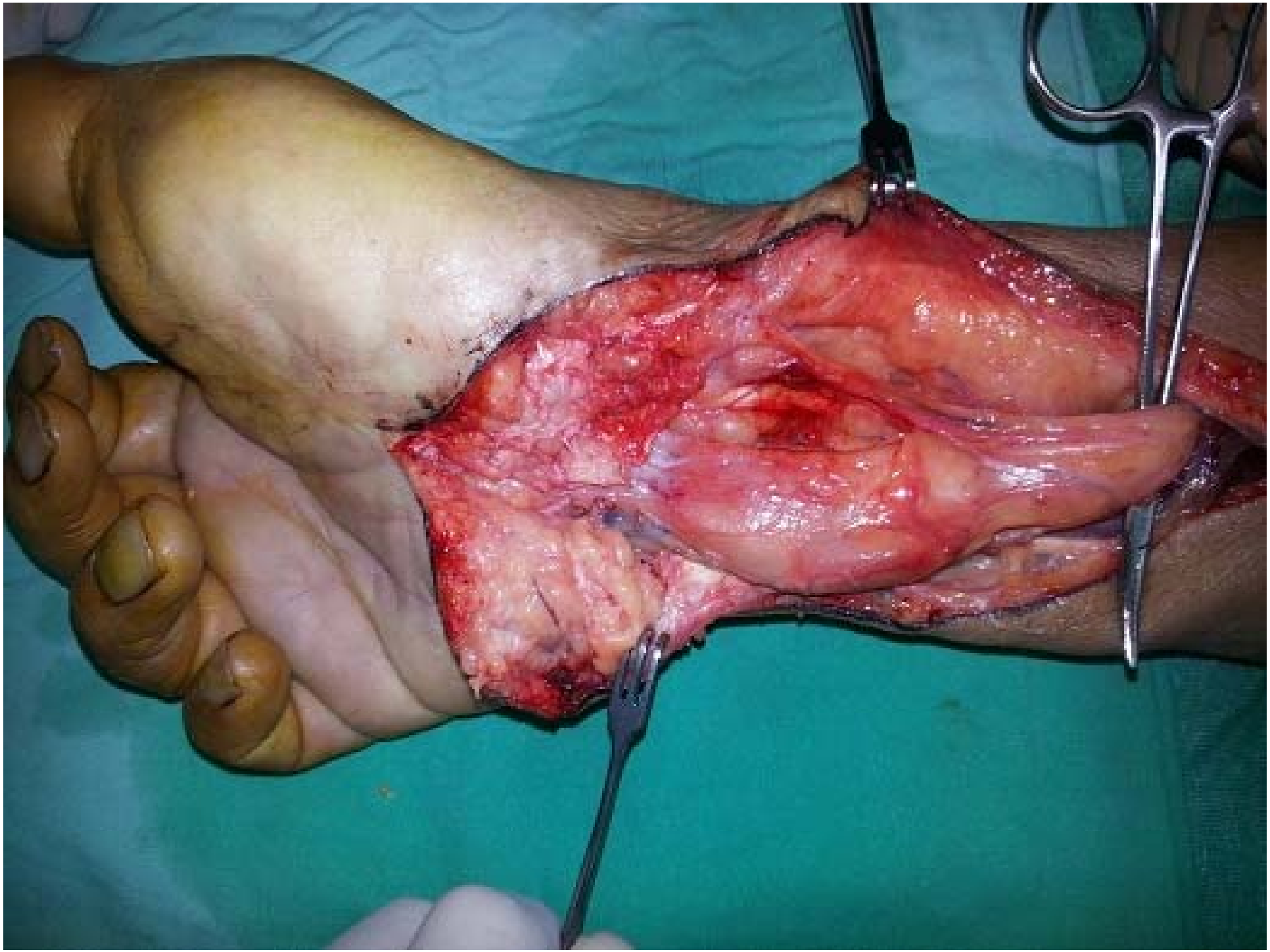
Carpal tunnel release was performed with exploration of the Guyon canal.

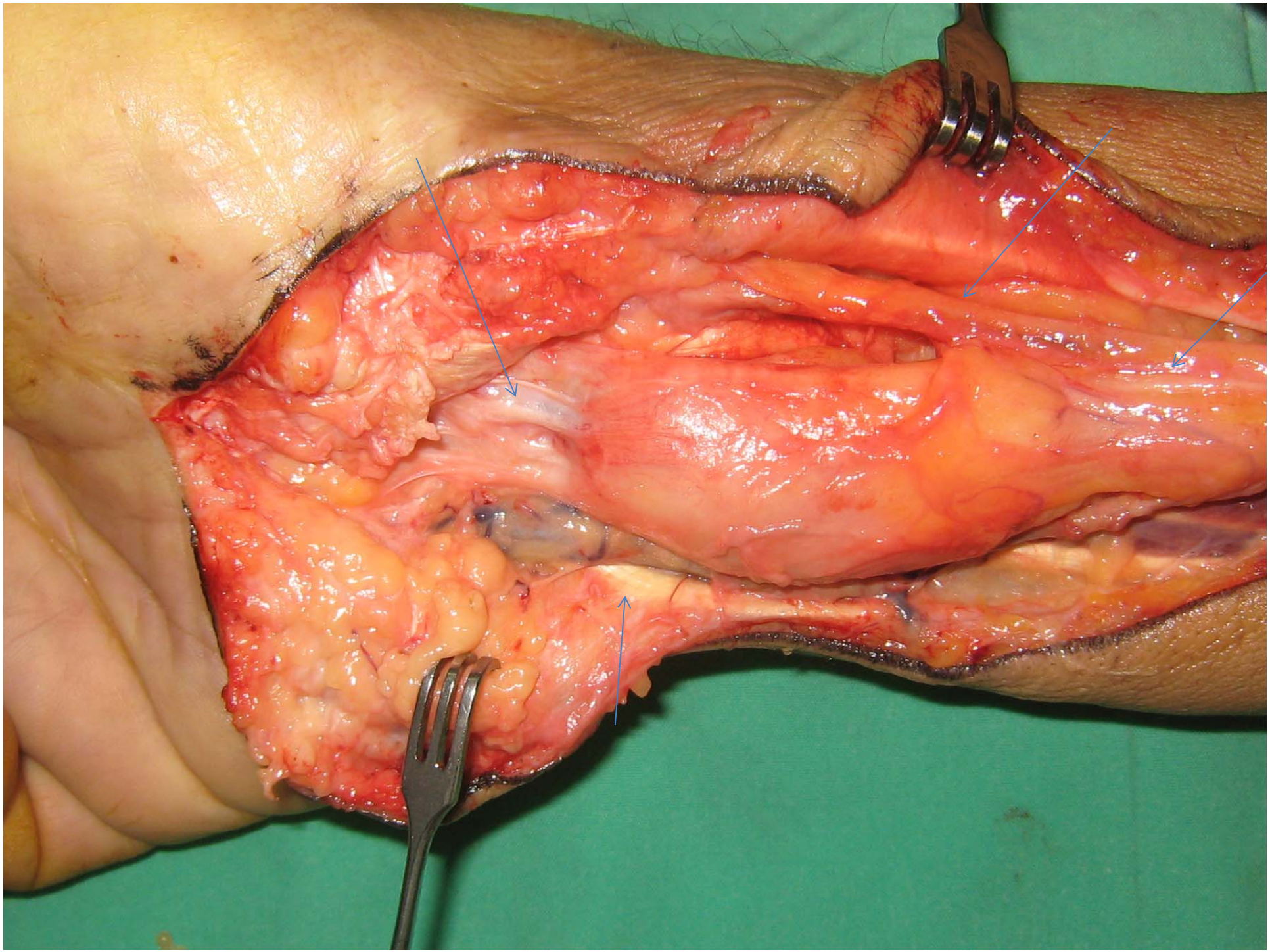


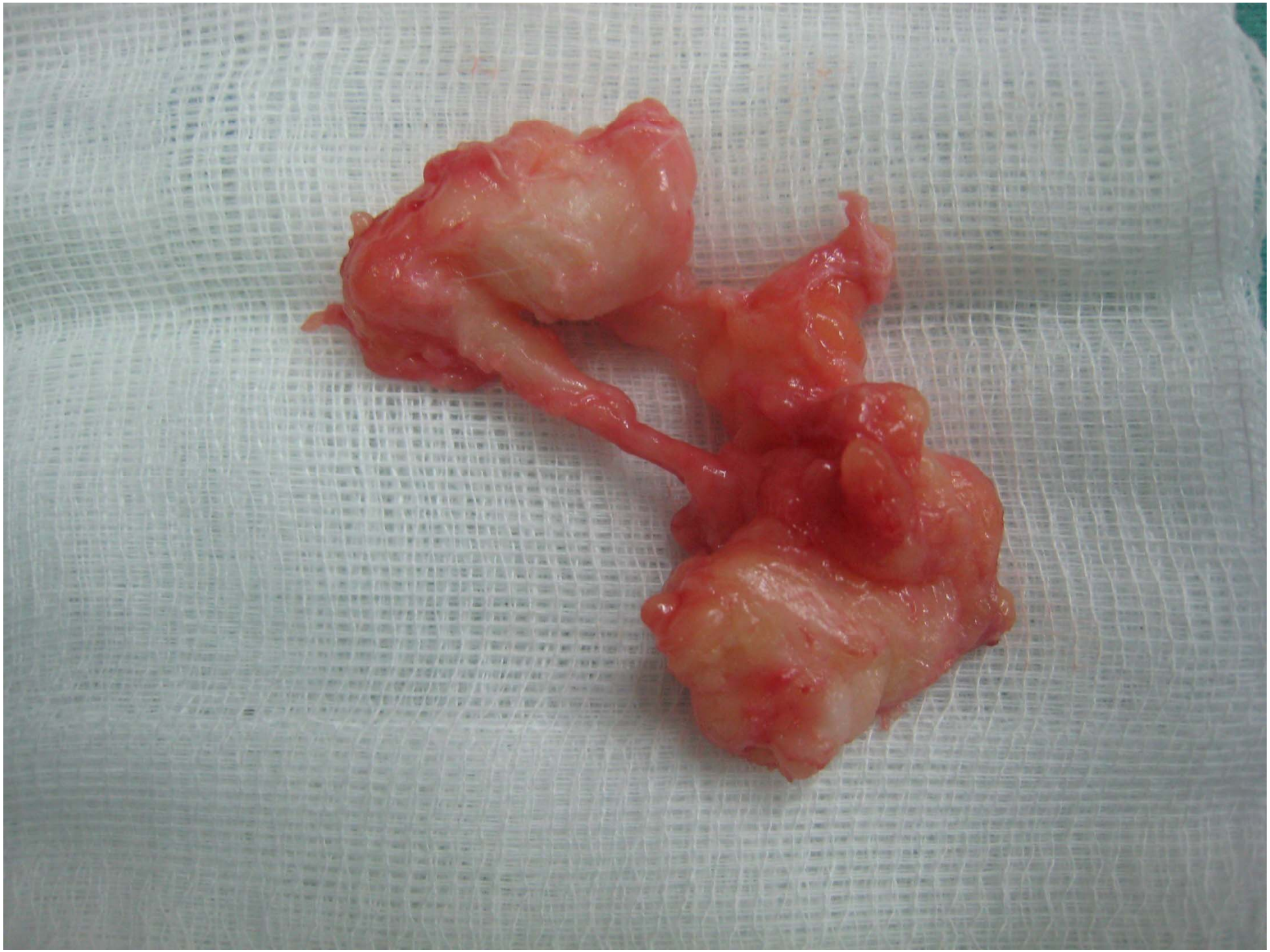
Median nerve with normal size at the forearm transforms to a giant nerve.











The presented case correlates with what **Feriz** (1925) coined as ***macrodystrophia lipomatosa progressiva.***

This rare form comprises 10% of all cases of congenital Macrodactyly.

It characterized by excessive proliferation of fibroadipose tissue in the distribution of the median nerve in the upper extremity or the planter nerve in the lower extremity.

Immediately post op. the patient reports a **significant pain relieve and slight improvement of sensation.**

THANK YOU!!!