

**Background:** Thrombotic thrombocytopenic purpura (TTP) and Microscopic polyangiitis (MPA) rarely occur in conjunction. Autoimmune TTP is caused by autoantibodies to the ADAMTS13 protein that cleaves von Willebrand factor. MPA is a small-vessel vasculitis that typically affects the small blood vessels of the lung and kidney [1]. We present a case of concurrent TTP and MPA which we believe could highlight a pathogenetic correlation and treatment overlap between the two diseases.

**Objectives:** To report a rare case of concurrent TTP and MPA and to educate physicians on optimal treatment strategies.

**Methods:** Case report and literature review.

**Results:** A 67-year-old patient with history of stroke and sickle cell trait presented to the emergency department due to altered mental status and fever. Serology on admission was notable for marked acute kidney injury and anemia requiring transfusions. Urinalysis showed hematuria and proteinuria. Autoimmune serology was remarkable for p-ANCA positivity at 1:80 with myeloperoxidase antibody > 30 pmol/L; a diagnosis of MPA was made. Patient's serology was also remarkable for thrombocytopenia with a nadir under 60 and evidence of thrombotic microangiopathy on peripheral blood smear and hemolysis labs. ADAMTS13 autoantibodies were reduced with less than 3% functionality and an inhibitor of 1/0 BU, confirming a concurrent diagnosis of TTP. Patient was initiated on high-dose steroids and therapeutic plasma exchange (TPE) with resultant normalization of platelets and improvement in renal function. TPE was discontinued, and the patient was transitioned to rituximab with continuation of high-dose steroids. The patient was ultimately discharged home with a steroid taper and maintenance rituximab dosing.

**Conclusion:** Per literature review, three other cases of concurrent TTP and MPA have been reported [2-4]. Further investigation into pathogenetic and treatment overlap of MPA and TTP may be warranted. Although rare, when occurring together the presentation is often severe, and urgent initiation of treatment is critical. TPE is the mainstay of treatment in TTP but is usually only recommended in MPA patients with anti-GBM antibodies, those at high risk of progression to end stage renal disease, or those unresponsive to first-line therapies [1,5,6]. Rituximab, which acts to decrease B cell activity and overall antibody production, has been shown to reduce disease activity in both conditions [1,5]. This case report suggests that, in the rare event that TTP and MPA are concurrently diagnosed, the combined use of rituximab and TPE in conjunction with steroids may induce remission more efficiently than either treatment alone.

#### REFERENCES:

- [1] Chung SA, Langford CA, Maz M, et al. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Antineutrophil Cytoplasmic Antibody-Associated Vasculitis. *Arthritis Rheumatol*. 2021. doi:10.1002/art.41773
- [2] Hirsch DJ, Jindal KK, Trillo AA. Antineutrophil cytoplasmic antibody-positive crescentic glomerulonephritis and thrombotic microangiopathy. *Am J Kidney Dis*. 1995. doi:10.1016/0272-6386(95)90663-0
- [3] Nagai K, Kotani T, Takeuchi T, et al. Successful treatment of thrombotic thrombocytopenic purpura with repeated plasma exchange in a patient with microscopic polyangiitis. *Mod Rheumatol*. 2008. doi:10.1007/s10165-008-0107-3
- [4] Syed Mujtaba Ali Naqvi, Rachel Whittaker, Arti Saraswat, Nisarfarthi Kazimuddin, Rishi Agarwal; A Rare Case of Concurrent New-Onset Microscopic Polyangiitis and Thrombotic Thrombocytopenic Purpura in a 77-Year-Old Woman. *Blood* 2021. doi: https://doi.org/10.1182/blood-2021-148966

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AB1851

#### TERMINATION OF PREGNANCY IN LUPUS NEPHRITIS: A BIOETHICAL ANALYSIS WITH THE INTEGRATIVE METHOD

**Keywords:** Best practices, Pregnancy and reproduction, Systemic lupus erythematosus

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**Background:** Termination of pregnancy in patients with rheumatic diseases is controversial due to current lack of evidence. Around one fourth of pregnancies in patients with rheumatic diseases are terminated due to medical reasons. Only 0.026% of articles in Rheumatology relate to bioethics despite the high frequency of ethical dilemmas in the treatment of pregnant patients with autoimmune diseases. [1]

**Objectives:** To analyze from a bioethical perspective the case of a pregnant patient with lupus nephritis unresponsive to treatment, for whom termination of pregnancy is considered as a therapeutic measure.

**Methods:** The integrative model was applied combining different normative ethical theories. The central moral dilemma and main stakeholders were identified. The case was analyzed from utilitarian, deontological and virtue ethics perspectives, followed by integration to establish a global conclusion.

**Results:** *Case presentation.* A 30-year-old woman was diagnosed with systemic lupus erythematosus in 2019 in Mexico. Induction therapy for lupus nephritis was started; however, it was interrupted due to COVID-19 pandemic. When she returned for follow-up, despite treatment was reinstated, she persisted with proteinuria without achieving complete remission criteria. Multi-target therapy was started; however, patient stopped treatment for economic reasons and became pregnant shortly after. She presented for prenatal control follow-up at 20 gestational weeks with edema in the lower extremities and hypertension, for which she was hospitalized in intensive care. Renal replacement therapy was initiated without further response to treatment. A multidisciplinary team suggested pregnancy termination as a therapeutic option, which the patient refused. *Bioethics analysis.* Figure 1 shows the background factors and initial steps for defining and analyzing the bioethical dilemma. Three different normative ethical theories were used. *Utilitarianism.* The consequences of the action are the central component, and the decisions made must be profitable and effective for the majority. Terminating the pregnancy would seek to protect the mother's life, since saving one life represents a greater benefit than losing two. For the patient's family, the costs of continuing with the pregnancy would be unsustainable. For the institution, the termination of the pregnancy would lead to a more efficient use of human and material resources. Therefore, interrupting the pregnancy is justifiable. *Deontology.* Duty-based ethics center the action itself regardless of consequences, as well as valuing autonomy. Seeking all therapeutic alternatives, including termination of pregnancy, is a good action itself. However, considering the patient's autonomy, her desire to continue pregnancy must be taken into account. Therefore, both terminating and continuing the pregnancy are justifiable. *Virtue ethics.* Aretology centers the importance of decisions in the person who performs the action. Core values involve enabling flourishing and personal growth. For both the family and the institution, flourishing would imply preserving the life of the patient. Therefore, the termination of pregnancy is justifiable.

**Conclusion:** Using the integrative method, we conclude that termination of the pregnancy is justifiable from the three ethical theories and is an appropriate resolution to the ethical dilemma. However, considering the patient's desire to continue pregnancy, health professionals should offer psycho-emotional support. The lack of recommendations about medically indicated termination of pregnancy in rheumatology leads to complex moral decisions, making the bioethical analysis of paradigmatic cases essential to ensure the best possible action and as a precedent for future similar situations.

#### REFERENCE:

- [1] Caplan, L., Hoffecker, L. and Prochazka, A.V. (2008) "Ethics in the rheumatology literature: A systematic review," *Arthritis & Rheumatism*, 59(6), pp. 816–821. Available at: <https://doi.org/10.1002/art.23703>.

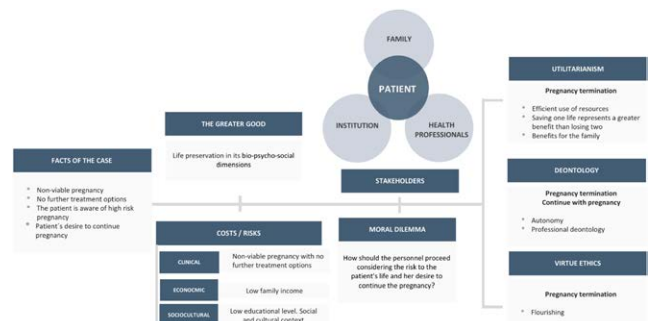


Figure 1. Integrative method and analysis based on normative ethical theories

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#### DERMATO-NEURO SYNDROME PRECIPITATED BY COVID-19 INFECTION IN THE SETTING OF SCLEROMYXEDEMA

**Keywords:** Skin, COVID

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**Background:** A black female in her 40s presented with a nonpruritic rash for 10 months consisting of bumps on the face, hands, forearms, and thighs. She had no prior treatment. Past medical history was significant for pulmonary embolism (PE) 6 years prior. She had no personal or family history of autoimmune disease. Physical exam revealed numerous smooth 2-3 mm skin-colored papules over the bilateral forearm dorsa, hands, anterior thighs, and face. Serum protein electrophoresis revealed monoclonal IgG lambda gammopathy. Skin biopsy of her left elbow showed dermal fibroplasia with mucin deposition. IgG was less than 1.5 grams/deciliter; bloodwork was otherwise stable. The diagnosis of scleromyxedema was rendered.

**Objectives:** The objective of this clinical case was to evaluate a neurologic sequela of COVID-19 infection in a patient with scleromyxedema.

**Methods:** One month following diagnosis of scleromyxedema, our patient was diagnosed with COVID-19 five days before admission to the emergency department with altered mental status and aphasia. Rheumatology was consulted due to malignant hypertension and acute kidney injury with question of scleroderma-like renal crisis in the setting of recently diagnosed COVID-19 infection, although she had no other features of systemic sclerosis. The infectious disease team was consulted due to COVID-19-induced inflammatory reaction.

**Results:** The patient's creatinine kinase and brain natriuretic peptide were elevated. Creatinine and potassium trended upwards. She developed seizures and became hemodynamically unstable with rapidly declining clinical status. She was transferred to the intensive care unit, where she developed respiratory arrest, shock, hyperkalemia, and acidemia. She received escalating doses of pressors but experienced frequent arrhythmic disturbances and developed asystole. Resuscitation efforts were unsuccessful; she expired within 24 hours of consultation.

**Conclusion:** Dermato-neuro syndrome (DNS) is a potential complication of scleromyxedema associated with confusion, dysarthria, seizures, and coma. The patient's clinical presentation is consistent with DNS in the setting of scleromyxedema likely precipitated by COVID-19. Intravenous immunoglobulins are first-line treatment for scleromyxedema; however, it is associated with risk of venous thromboembolism. The patient was considered for treatment as an outpatient but deferred due to history of PE. She was reevaluated for treatment upon presentation to the hospital, but given the severity and rapidity of her condition, it was already too late. This is the second reported case of COVID-19 induced DNS in a patient with scleromyxedema. Given the severity, we recommend early initiation of treatment in patients with scleromyxedema and aggressive treatment for those contracting COVID-19.

#### REFERENCES:

- [1] Haber R, Bachour J, El Gemayel M. Scleromyxedema treatment: a systematic review and update. *Int J Dermatol.* 2020;59:1191-1201.
- [2] Flannery MT, Humphrey D. Deep Venous Thrombosis with Pulmonary Embolism Related to IVIg Treatment: A Case Report and Literature Review. *Case Rep Med.* 2015;971321.
- [3] Lee YH, Sahu J, O'Brien MS, D'Agati VD, Jimenez SA. Scleroderma Renal Crisis-Like Acute Renal Failure Associated With Mucopolysaccharide Accumulation in Renal Vessels in a Patient With Scleromyxedema. *J Clin Rheumatol.* 2011;17:318-322.
- [4] Hoffman-Vold AM, Distler O, Bruni C, et al. Systemic sclerosis in the time of COVID-19. *Lancet Rheumatol.* 2022;4:e566-575.
- [5] Fritz M, Tinker D, Wessel AW, et al. SARS-CoV-2: A potential trigger of dermato-neuro syndrome in a patient with scleromyxedema. *JAAD Case Rep.* 2021;18:99-102.

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AB1853

#### SPONTANEOUS ABORTION AS DIFFERENTIAL DIAGNOSIS OF INTERMITTENT GLOMERULAR PROTEINURIA IN CLINICALLY AND SEROLOGICALLY INACTIVE SLE

**Keywords:** Systemic lupus erythematosus, Pregnancy and reproduction, Kidneys

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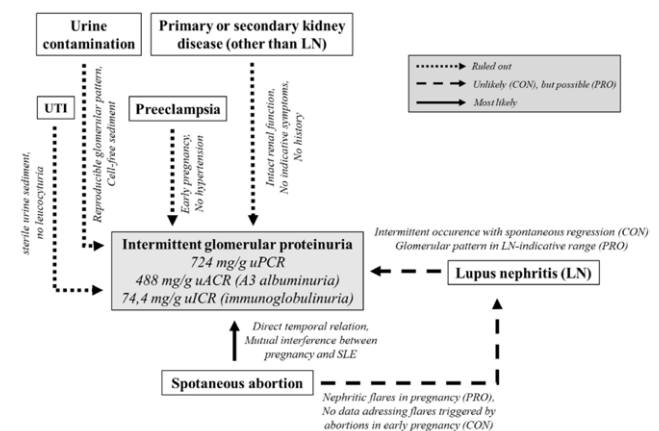
**Background:** In SLE, new-onset proteinuria and albuminuria raise concern for renal involvement, even when isolated and subnephrotic. In women of childbearing age, pregnancy-related complications are other causes of renal protein loss. Distinguishing renal disease from pregnancy-related complications of SLE is difficult because SLE is associated with adverse maternal and fetal events, while pregnancy is also a cause of disease relapses. Common to all causes of pathologic proteinuria in SLE is that they are related to disease activity and/or the need for intervention, as proteinuria does not normalize on its own.

**Objectives:** We report a case of a patient with inactive SLE who intermittently developed significant isolated proteinuria and albuminuria of glomerular origin that was directly related to spontaneous abortion in early (possibly twin-) pregnancy.

**Methods:** A 39-year-old female with SLE on hydroxychloroquine presented at our clinic for a routine check-up. Physical examination and laboratory tests were performed. To that date, SLEDAI-2K during most visits had a score of 0, indicating inactive disease.

**Results:** The patient reported feeling well, and the physical examination was unremarkable. Stable low-grade thrombocytopenia was the only relevant abnormality in recent years. On the current examination, the urine showed significant proteinuria and A3 albuminuria in the protein/albumin to creatinine ratio (uPCR 724 mg/g, uACR 488 mg/g). IgG/creatinine was markedly elevated (74.4 mg/g), and  $\alpha$ 1-microglobulin was normal, indicating nonselective glomerular proteinuria without tubular impairment. There were no other abnormalities suggestive of SLE activity. After she recalled having vaginal spotting a week earlier, a pregnancy test was performed and was positive. Subsequently, the  $\beta$ HCG level increased insufficiently, and ultrasound detection of a live embryo was not successful, but the presence of two amniotic cavities was suspected. An early incomplete miscarriage was diagnosed, and the gestational age was calculated to be 6+5 weeks. Shortly thereafter, a planned suction curettage was performed. One week later, she had a final vaginal bleed. At this time, the urine showed a decrease in proteinuria of over 50% (uPCR 317 mg/g, uACR 210 mg/g, IgG 26.9 mg/g). Three weeks later, urine protein was completely normalized, which proved stable 12 weeks after initial diagnosis. Throughout the follow-up period, she did not have any SLE relapse.

**Conclusion:** After exclusion of differential diagnoses (chart), causality of the miscarriage with the urinary findings seems evident. To date, there have been no reports of the concomitant occurrence of early pregnancy miscarriage, possibly LN-indicative glomerular proteinuria, and its spontaneous regression in a clinically and serologically inactive SLE patient. Recently, observations were confirmed that a large proportion of patients with a uPCR <1g/g had LN histology, whereas a relevant proportion had inactive sediment or normal serology like our patient. As the kidney is the organ most affected in SLE pregnancy, it is important to be aware of intermittent proteinuria, as the consequences may be very different from persistent proteinuria as would be expected in nephritis.



**Diagnostic algorithm of intermittent glomerular proteinuria. Clustering in probability of etiology. Arrow labeling indicates diagnostic steps leading to "rule-out" or "rule-in" of diagnosis.**

**Figure 1.**

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AB1854

#### CORRELATION OF THE OUTCOMES MEASURED BY THE PATIENTS AND BY THE MEDICAL TEAM ACCORDING TO THE LEVEL OF ADHERENCE AND AN EDUCATIONAL TOOL FOR PATIENTS

**Keywords:** Outcome measures, Patient reported outcomes

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