DIAGNOSTIC CHALLENGE IN A YOUNG PATIENT WITH KIDNEY AMYLOIDOSIS

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Male patient at the age of *40 years*, was hospitalized due to nephrotic syndrome and anasarca. He has been *paraplegic* for the past 20 years.
Family history was negative for kidney diseases.
Initially he presented with deep vein thrombosis of the leg and *severe nephrotic syndrome* (proteinuria of 20 g/dU) was detected. A large *decubitus ulcer* of sacral region, which has been deteriorating for a past year, was also verified at that time.

Prior to kidney biopsy extensive diagnostic tests were performed, and *immunofixation* results showed positivity for *monoclonal IgG lambda*. Also, *7% of monoclonal plasma cells* in the bone marrow were found (expressing the CD38 + CD138 + CD19- s phenotype by monoclonal expression of lambda immunoglobulin light chains).

A **kidney biopsy** was performed and a diagnosis of *renal amyloidosis* was made (Figure 1).
Immunohistochemical positivity for AA was found, while staining for the kappa and lambda chains showed a polytypical reaction. Given the fact that laser microdissection and mass spectrometry is considered the most reliable method for amyloidosis typing, formalin paraffin embedded block with kidney specimen was sent to National Amyloidosis Centre in London for **mass spectrometry** to be performed. Amyloid deposits were present in glomeruli, tubules and vessels.
The proteomic analysis of the amyloid supported the result of the immunohistochemical staining, diagnosis of AA kidney amyloidosis remained.

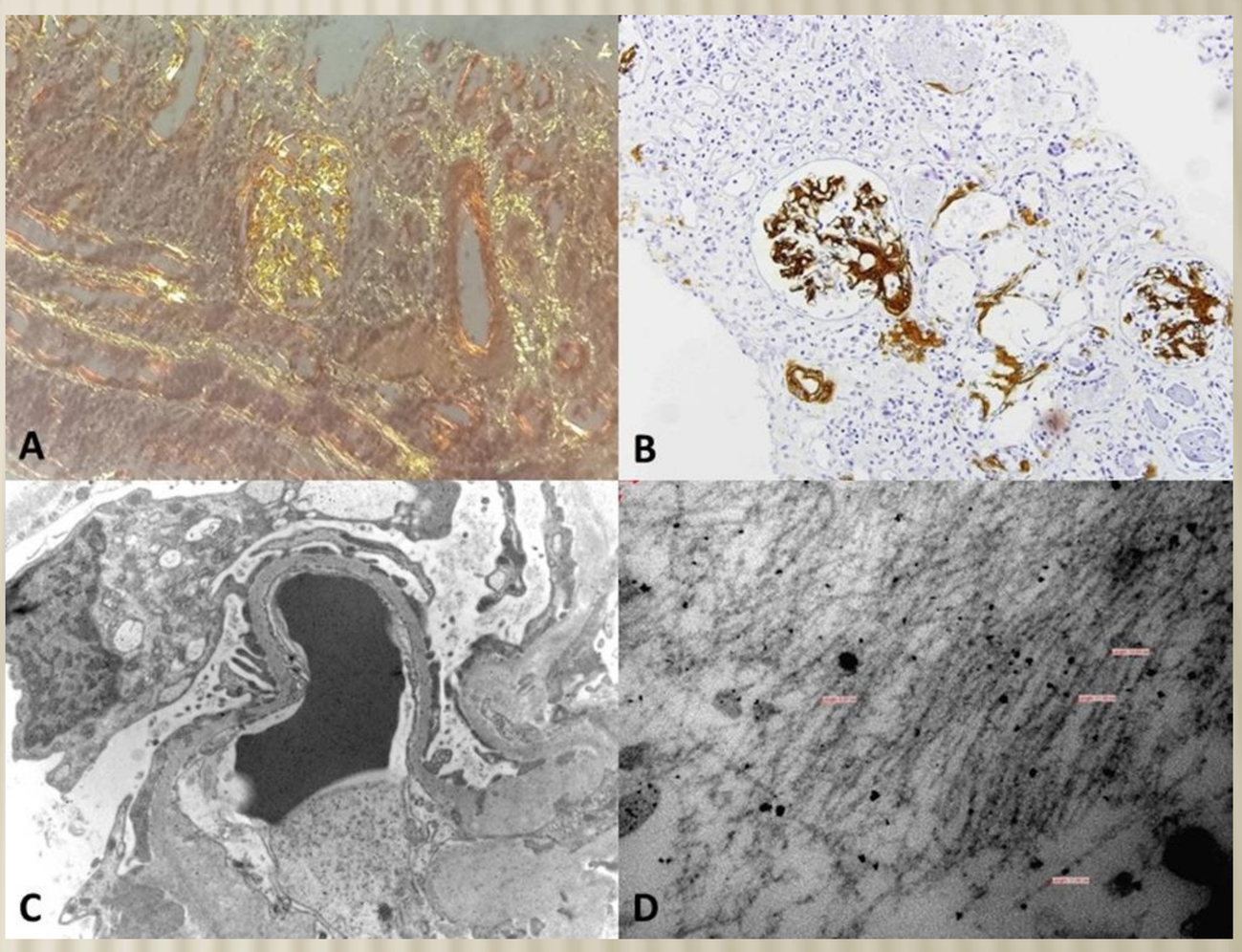

***Figure 1** A) Birefringence under polarized light, Congo Red stain, magnification x200. B) Positive AA, immunohistochemistry, magnification x 200. C) Amyloid in mesangium and basement membrane. TEM, original magnification x 10000. D) Amyloid fibrils measuring 10 nm in average. Fibrillar material in mesangium and basement membrane. TEM, original magnification x 100000*

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The concluding opinion was that there were no criteria for diagnosis of monoclonal gammopathy of renal significance. The main cause of *AA kidney amyloidosis* was probably a *chronic inflammation* derived from massive decubitus ulcer. Only option to prevent further kidney deterioration was to treat the underlying inflammatory disease.