Fibrillary glomerulonephritis: a rare entity with unique ultrastructural characteristics

Glomerulonefrite fibrilar: uma entidade rara com características ultraestruturais únicas

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¹University Hospital of Nephrology, Skopje, R of North Macedonia. Fibrillary glomerulonephritis (FGn) is characterized by deposition of randomly arranged polyclonal immune deposits in glomerular matrix^{1,2}. A 56-year-old hypertensive patient presented to our hospital with proteinuria (3.04 g/24 hours) and an elevated serum level of creatinine (391 µmol/L). Electron microscopic evaluation of kidney biopsy specimens set the diagnosis of fibrillary glomerulonephritis (Figure 1). There was no evidence of monoclonal components in the blood and urine. Antinuclear and anti-double-stranded DNA antibodies, complement components C3 and C4 and markers of viral hepatitis were also negative.

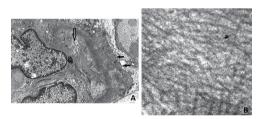


Figure 1. Electron microscopy evaluation demonstrates deposition of organized fibrils with consecutive expansion of mesangial matrix and permeation of lamina densa of the glomerular basal membrane (GBM) (Image A: hollow arrow). Visceral epithelial cells (podocytes) show complete effacement of foot process (Image A: black arrows). At higher magnification (x 60.000), the fibrils are randomly arranged and non-branching, measuring 15 to 18 nm in diameter (Image B).

The benefit of immunosuppressants is limited,³ and half of patients progress to kidney failure within 2 years¹⁻³.

AUTHORS' CONTRIBUTION

VK contributed with the collection, analysis and interpretation of data and writing of the article. GS contributed with the performing of the kidney biopsy, collection, analysis and interpretation of the data. PDV contributed with the performing of the kidney biopsy, collection, analysis and interpretation of the data, and approval of the final version.

CONFLICT OF INTEREST

None to declare.

REFERENCES

- 1. Rosenstock JL, Markowitz GS, Valeri AM, Sacchi G, Appel GB, D'Agati VD. Fibrillary and immunotactoid glomerulonephritis: distinct entities with different clinical and pathologic features. Kidney Int. 2003 Apr;63(4):1450-61.
- Azevedo A, Cotovio P, Góis M, Nolasco F. Rare diagnosis in a patient with diabetes with nephrotic proteinuria. BMJ Case Rep. 2019 Jan;12(1):bcr-2017-223835.
- 3. Pronovost PH, Brady HR, Gunning ME, Espinoza O, Rennke HG. Clinical features, predictors of disease progression and results of renal transplantation in fibrillary/immunotactoid glomerulopathy. Nephrol Dial Transplant. 1996 May;11(5):837-42.

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