

Membranoproliferative glomerulonephritis with C3 and intramembranous dense deposits

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Membranoproliferative glomerulonephritis (MPGN) encompasses 7 to 10% of all biopsied glomerulonephritis. They are divided in: MPGN type I; MPGN type II and MPGN type III, being primary or secondary. MPGN type I are the most frequent, MPGN types II and III are very rare and difficult to diagnose without clinical and morphologic findings integration. MPGN type II or Dense Deposit Disease has a varied morphologic appearance with a few numbers of cases showing a membranoproliferative pattern by Light microscopy (LM). Electron microscopy (EM) is pivotal to confirm the diagnosis [1].

We present a case of 35 years old man, with nephrotic proteinuria and mild renal insufficiency since 2 years. The only relevant clinical data is facial lipodystrophy. Complement 3 (C3) was low and C3 nephritic factor negative. There were not other relevant abnormalities. Renal biopsy was fixed in buffered formaldehyde 10% and performed for LM. The frozen fragment, prepared for observation by fluorescence microscopy - immunofluorescence (IMF) -, was prepared to be stained with florescent anti-serums, against immunoglobulines (IgG, IgA and IgM) and complement factors (C3, C4, and C1q). EM was later done on tissue formaldehyde fixed reprocessed from paraffin-embedded for LM, because there was no tissue fragment fixed in glutaraldehyde.

LM showed variable endocapillary hypercellularity, with neutrophils infiltration. Capillary walls were thickened due to the deposition of elongate and ribbon-like deposits. Few double contours were visible (Figure 1a). IMF demonstrated the presence of C3 deposits in the capillary walls and mesangium (Figure 1b). EM confirmed the presence of an intramembranous dense deposit along basement membrane which was thickened (Figure 1c). LM and IMF findings favored the diagnosis of MPGN type II with C3 deposits and thickening of basement membrane. Nevertheless EM was essential to confirm intramembranous unequivocally dense deposits [2].

MPGN type II is a rare glomerulonephritis mediated by complement deregulation. The integration of clinical and morphologic findings is essential to get a correct diagnosis. In this setting EM is highly distinctive and required for a definitive diagnosis.

References

1. Zhou X. and Silva F., In Heptinstall's Pathology of the Kidney. Jennette J.C. *et al.*, (Eds.), Lippincott Wilkins: Philadelphia, PA., pp. 253-319, 2006.
2. Sethi S. and Fervenza F., N. Engl. J. Med., 366:1119-1131, 2012.

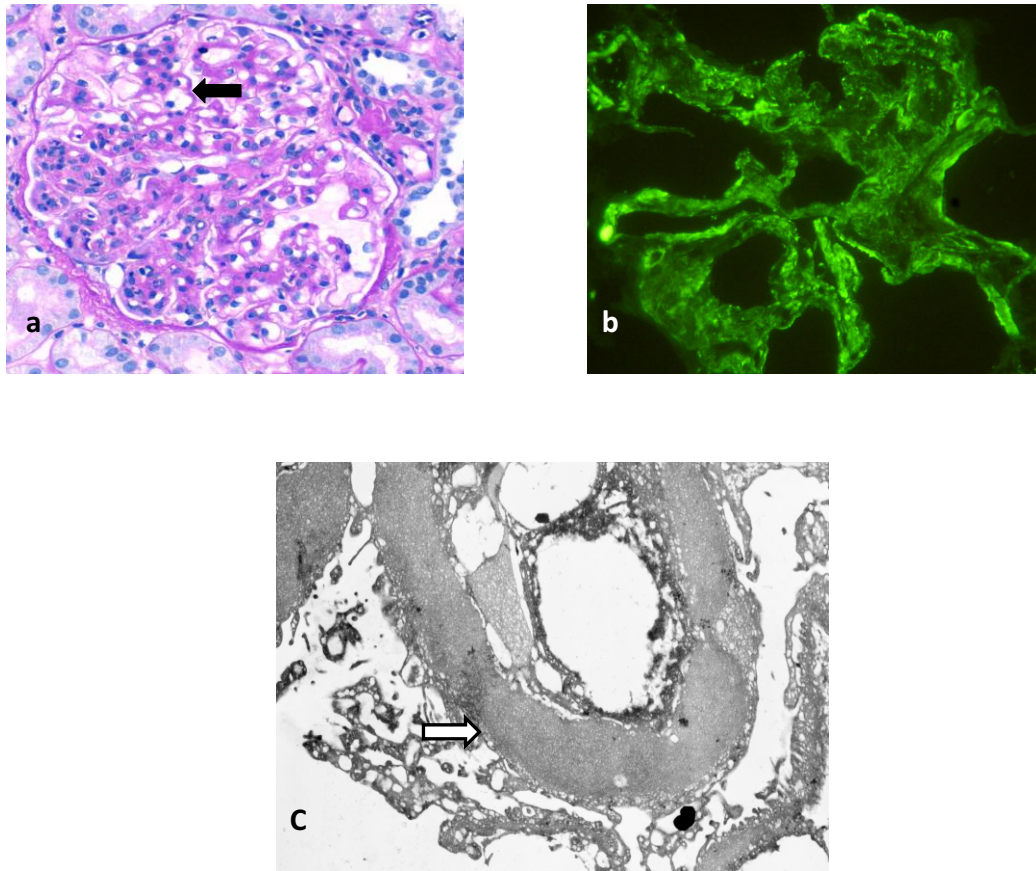


Figure 1 – a) Endocapillary hypercellularity, with neutrophils infiltration. Capillary walls were thickened due to the deposition of elongate and ribbon-like deposits \blackrightarrow (PAS X 400); b) IMF demonstrated C3 deposits in the capillary walls and mesangium; c) Presence of an intramembranous dense deposit along basement membrane which was thickened \Rightarrow (EM on tissue reprocessed from paraffin-embedded material; X 15000).