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LUPUS OR NOT LUPUS? That is the question: a case report about a full-house GNMP.

M. BENKIRANE, Z. ABOUDAR,N. HAMOUCHE, M. CHETTATI, W. FADILI, I. LAOUAD.

Nephrology, Morocco

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Abstract: Lupus nephritis diagnosis is defined by the EULAR and ACR lupus classification criteria and its diagnosis is usually easy when the SLE (Systemic Lupus Erythematosus) is known, but it may be challenging when the kidney manifestation is inaugural with no other organ envolvement especially no immunological criteria. We report the case of a 43 years old patient admitted for impure nephrotic syndrome with macroscopic hematuria and renal failure. The renal biopsy showed the aspect of a glomerulonephritis mesangio-proliferative (GNMP) with a full-house aspect in immunofluorescence. the presence of specific signs of Lupus nephritis helped retain the diagnosis of Lupus. And the positivity of the immunological assessment during the follow-up confirmed it.

INTRODUCTION

Full-house staining of glomeruli in renal histopathology is defined by the presence of IgA, IgM, IgG,C3 and C1Q deposits, it is highly suggestive of lupus nephritis especially for the C1q;other nonlupus entities can also be present with a similar pattern on immune fluorescence [1]. Different names are used for this entity with full house staining on immunofluorescence (IF) with negative serology for lupus;some authors used the term full-house lupus-like nephropathy [2]. Its differentiation from authentic lupus nephropathy stays problematic essentially due to possibility of seroconversion of SLE (Systemic Lupus Erythematosus) serum antibodies which allows a retrospective diagnosis of Lupus Nephritis [3].

CASE REPORT

Patient of 43-years-old,admitted for impure nephrotic syndrome with macroscopic hematuria and renal failure evolving for one month,alcoholic-smoker, withno history of nephropathy or known systemic disease. Diuresis initially maintained at 1l per day with no associated extra-renal signs.

Assessment on admission showed urea at 1.72 mg/l, creatinine 99 mg/l, protein 49 g/l, albumin 29g/l, calcemia 89 mg/l, hyperphosphatemia 109 mg/l, CRP negative, CPK normal and LDH slightly elevated to 327 IU/ml, ASLO negative.

A normal seric complement.

Proteinuria was 3.2 g/dl, with urine cytobacteriological study showing leukocyturia at 409,000/ml and hematuria at 430,860/ml, and a sterile culture.

The electrophoretic profile showed hypogammaglobulinemia and complement immunohisto chemistry revealed a monoclonal band corresponding to a kappa free light chain.

Weighted determination of light chains in the blood showed a normal Kappa/Lambda ratio.

And the myelogram showed 3% of plasmocytes with no abnormal cells.

Infectious assessment including heart sonography was with no abnormality.

Renal biopsy yielded 13 glomeruli, one of which was a sealing loaf. At high magnification, the glomeruli were lobulated, with accentuation of the mesangial meshwork and mesangio-capillary proliferation. Karyorrhexis and neutrophil influx were present. Silver staining reveals a double contour.

Immunofluorescence: a full-house appearance with both kappa and lambda deposits predominantly in the mesangial and capillary areas.
C1q deposits predominate in peri-tubular.

The etiological assessment of full-house mesangial proliferative glomerulonephritis showed a negative immunological work-up.

We decided to start a treatment based on intravenous methylprednisolone relayed by oral prednisone and monthly cyclophosphamide, as the karyorrhexis and tubuloreticular inclusions (TRI) were highly suggestive of a lupus.

The follow up showed after one month positive antinuclear at a titer of 1/320 and we could retrospectively retain the diagnosis of Lupus nephritis.

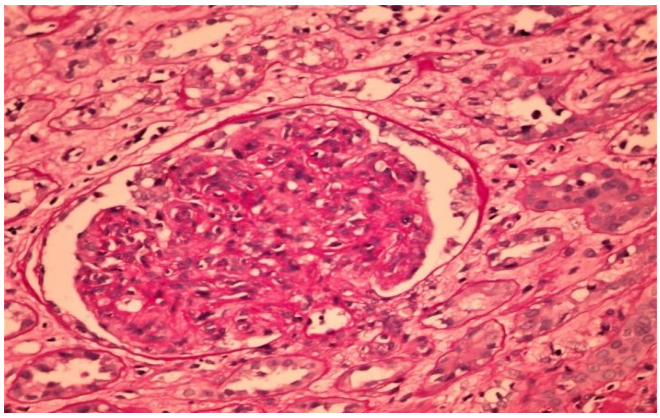

Figure 1: PAS coloration showing lobulation of the glomerulus.

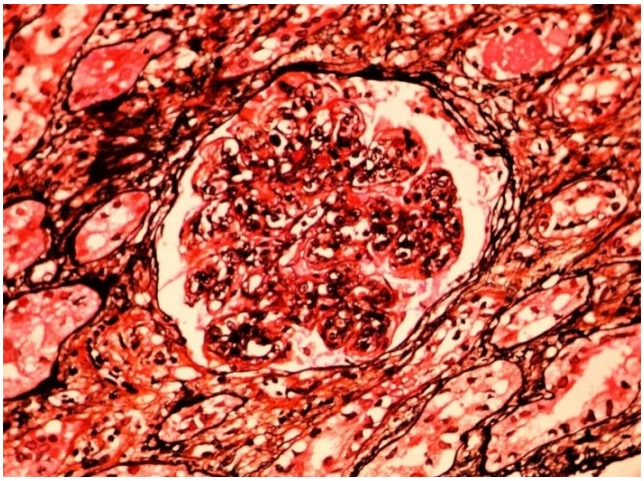

Figure 2: Silver staining showing a double contour aspect.

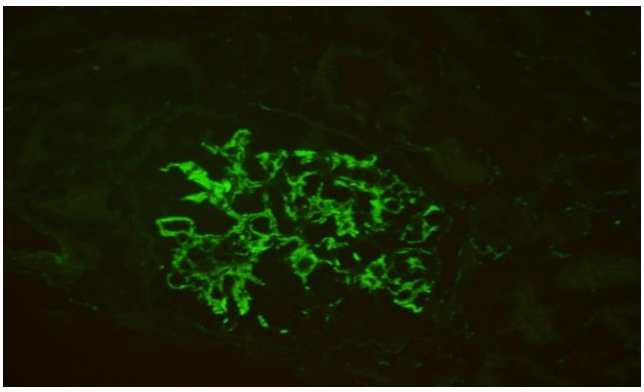

Figure 3: Full-House aspect in the immunofluorescence (IF) :Deposits of IgA, IgM, IgG, C3, C1q.

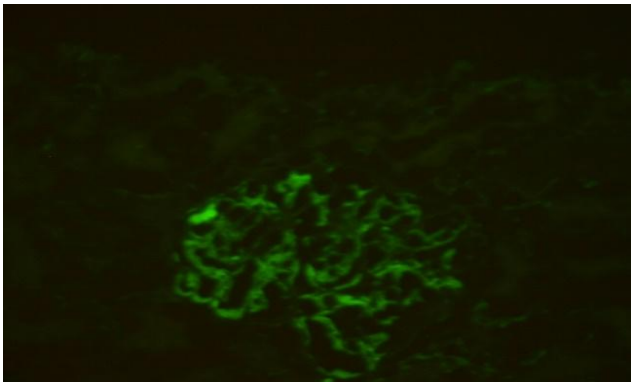

Figure 4: Full-House aspect in the immunofluorescence (IF) :Deposits of IgA, IgM, IgG, C3, C1q.

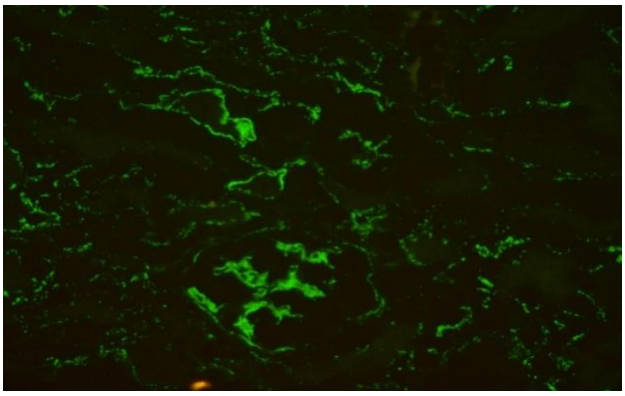

Figure 5: Full-House aspect in the immunofluorescence (IF) :Deposits of IgA, IgM, IgG, C3, C1q.

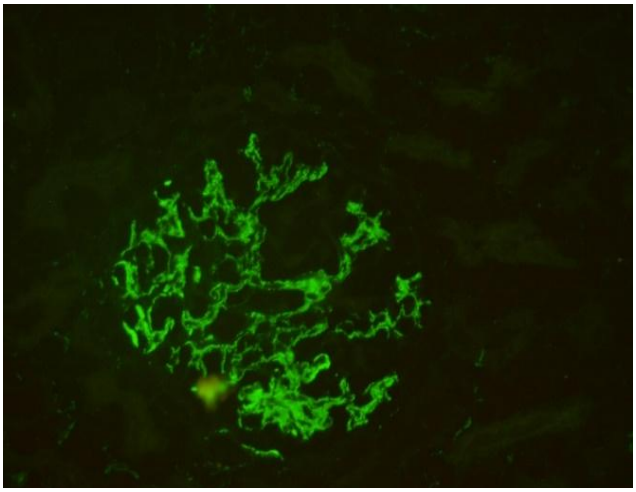

Figure 6: Full-House aspect in the immunofluorescence (IF) :Deposits of IgA, IgM, IgG, C3, C1q.

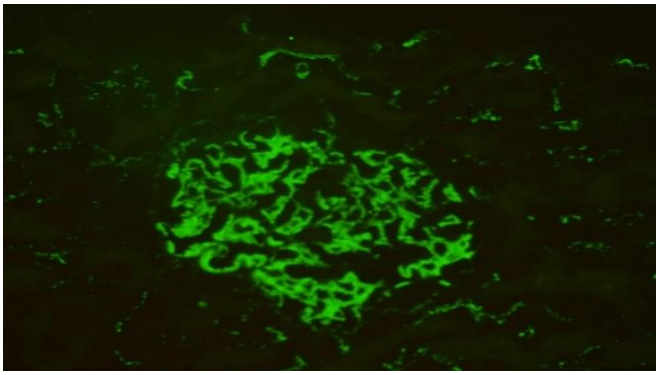

Figure 7: Full-House aspect in the immunofluorescence (IF): Deposits of IgA, IgM, IgG, C3, C1q.

DISCUSSION

This case report illustrates challenges encountered in the diagnosis of a full-house mesangial proliferative glomerulonephritis when histological study is highly in favor of a lupus nephritis without any other criteria of this pattern. A retrospective diagnosis is possible during the follow up if serum antibodies become positive: seroconversion as noticed in our case [4].

The 2019 EULAR/ACR classification criteria for SLE include positive ANA at least once as obligatory entry criterion; followed by additive weighted criteria grouped in 7 clinical (constitutional, hematologic, neuropsychiatric, mucocutaneous, serosal, musculoskeletal, renal) and 3

immunologic (antiphospholipid antibodies, complement proteins, SLE-specific antibodies) domains, and weighted from 2 to 10;patients accumulating ≥10 points are classified[7].

Lupus nephritis defined by the ACR lupus classification criteria is >0.5 g of proteinuria per day or a urinary protein/creatinine ratio (UPCR) of >0.5 or urinary protein greater than 3+ by dipstick analysis. As a second mandatory criterion, urinary cellular casts or an 'active' urinary sediment of more than five white blood cells per high-power field (in the absence of urinary tract infection) and doesn't precise diagnosis criteria if Lupus is not known and the renal manifestation is inaugural[8].

A study conducted by Rijnink et al. including 149 patients biopsied ,32 had FHN and none of them developed a SLE during 20 years follow up [5].

A case report published by Baskin E et al. of a ten years old girl with FHN with no other criteria of lupus was treated as a lupus nephritis ; this patient showed no seroconversion after one year, but the fact is that these patients may never become seropositive or develop SLE symptoms [6].

Another study conducted by Uzzo and al. concluded that SLE and nonlupus full house nephropathy are distinct clinical entities, with comparable outcomes; a small subset of patients develops SLE during follow-up; andnonlupus full house nephropathy is addressed by many different names in the literature [4].

CONCLUSION

Our case report is an illustration of how challenging the diagnosis of lupus nephritis can be in our clinical practice especially when the renal manifestation is inaugural and isolated. Seroconversion might confirm the diagnosis retrospectively, and still a rigorous study of the renal biopsy is the key of the diagnosis so as to justify an immunosuppressive therapy.

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