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Autores:

Batista V.
Moreira T.
Meireles R.
Botelho C.

NEFROPATÍA POR IgA EN UN ADULTO JOVEN CON SÍNDROME NEFRÓTICO: REPORTE DE UN CASO

IgA nephropathy in a young adult with nephrotic syndrome: case report

Autores/Authorship:

Dr. Vasco Batista (Batista V.)

Internal Medicine Department, Centro Hospitalar do Tâmega e Sousa, Penafiel, Portugal.

Urb. Quinta de Carregoso, 234, 4580-294, Paredes.

00351915575322

vasco.abbatista@gmail.com

Dra. Teresa Moreira (Moreira T.)

Internal Medicine Department, Centro Hospitalar do Tâmega e Sousa, Penafiel, Portugal.

Dr. Ricardo Meireles (Meireles R.)

Internal Medicine Department, Centro Hospitalar do Tâmega e Sousa, Penafiel, Portugal.

Dr. Carlos Botelho (Botelho C.)

Nephrology Department, Centro Hospitalar do Tâmega e Sousa, Penafiel, Portugal.

RESUMEN

La nefropatía por IgA tiene una amplia gama de presentaciones clínicas, de las cuales el síndrome nefrótico es posiblemente el más infrequente. Individuos con nefropatía por IgA ocasionalmente desarrollan síndrome nefrótico, que es indistinguible de la enfermedad de cambios mínimos (ECM). En la mayoría de los casos, la incidencia de la nefrosis y la asociación de nefropatía por IgA con ECM fue mejor descrita con pacientes que respondían a la corticoterapia y sin progresión para enfermedad renal crónica. Relatamos un caso similar en un hombre joven que presenta síndrome nefrótico.

Palabras clave: nefropatía por IgA, enfermedad de cambios mínimos, síndrome nefrótico.

ABSTRACT

IgA nephropathy (IgAN) has a wide range clinical presentations, of which nephrotic syndrome is possibly the rarest. Patients with IgAN occasionally develop nephrotic syndrome, which is indistinguishable from minimal change disease.

There is a sudden onset of nephrosis, and the association of IgA nephropathy with minimal change disease was better described with patients having complete response to steroid therapy and no progression to CKD. We report a similar case in a young male presenting with nephrotic syndrome.

Keywords: IgA nephropathy, minimal change disease, nephrotic syndrome.

INTRODUCTION:

IgA nephropathy (IgAN) is a mesangial proliferative glomerulonephritis characterized by diffuse mesangial deposition of IgA. Is the most common cause of primary glomerulonephritis (GN) worldwide and a major cause of CKD and an important cause of end-stage renal disease (ESRD). It is likely tha IgAN is not a single entity but rather a common response to various injurious mechanism.[1]
The wide range of clinical presentation of IgA varies in frequency with age. No clinical pattern is pathognomonic of IgA.[1]
Clinical presentation is varied but asymptomatic hematuria with progressive kidney disease is the most frequent manifestation. It is rare for proteinuria to occur without microscopic hematuria.[1]
Whereas nephrotic-range proteinuria may occur, in particular in the presence of uncontrolled hypertension, full blown nephrotic syndrome is uncommon, occurring in only 5% of all patients with IgAN. Nephrotic syndrome may occur early in the course of the disease, with minimal glomeular change or with active mesangial proliferation GN. Alternatively, it may occur as a late manifestation of advanced chronic glomerular scarring.[1]
Although proteinuria is as anindependent predictor of unfavorable renal outcome, patients with nephrotic syndrome in IgA seem to have a different outcome.[2] Recent evidence have shown tha tmild IgAN with diffuse foot process effacement on electron microscopy (EM) corresponds to IgAN with superimposed minimal change disease (MCD) and that these patients have remarkably good prognosis with steroid.[3,4,5] therapy. We hereby report a case of IgAN presenting with nephrotic syndrome.

CASE REPORT

A 30- year-old, non-smoking Caucasian man, with no previous serious illnesses was referred to our clinic in May 2017 with a recent (one month) lower limb oedema.
Upon admission, clinical examination showed pronounced lower limb oedema and blood pressure of 150/80 mmHg. No history of fever or haematuria was disclosed; and no respiratory, gastrointestinal or dermatologic symptons were present.
Laboratory blood tests revealed normal haemoglobin levels; the leukogram and platelets were normal. The erythrocyte sedimentation rate was increased (ERS 35 m/1sthr). Renal and liver function levels, electrolytes and thyroid function levels were normal. The laboratory studies revealed hypoproteinaemia (3,9g/dl), hypoalbuminaemia (2,7g/dl); total cholesterol 267 mg/dl and LDL 179 mg/dl.
Urine analysis showed proteinuria and microscopic haematuria. Urine microscopy showed 15 erythrocytes per high-power filed. 24 hour urine protein excretion was 7,49 gr.

Blood and urine cultures were negative. Blood tests were also negative for autoimmunity diseases, monoclonal gammopathies, HCV, HBV, HIV or other opportunistic infections.

The renal ultrasound showed normal size kidneys.

In order to clarify the aetiology of the nephrotic syndrome, the patient underwent a percutaneous needle kidney biopsy.

Histological examination at light microscopy showed sclerosis in 1 of the 15 glomeruly and modest mesangial proliferation (Figure 1). Tubular epithelium had protein reabsorption droplets. Intesrtitium and vessels were normal.

Immunofluorescence showed granular mesangial IgA deposits (+++)- Figure 2.

Electron microscopy showed mesangial deposits and diffuse foot process effacement- Figure 3.

A IgaN was diagnosed with superimposed minimal change disease.

At the time of diagnosis treatment with prednisolone – 60 mg per day – was started, and a significant clinical improvement followed.

The patient evolved with regression of proteinuria – 0,5 gr/ 24 hours , disappearance of haematuria, and maintains a normal renal function at the 6 months follow up, under therapy with low dose of prednisolone and angiotensin II receptor antagonist.

DISCUSSION

IgAN is the most common cause of idiopathic glomerulonephritis in the world.[6] Although this disorder was initially thought to have a benign course, it is now recognized that slow progression to end-stage renal disease (ESRD) occurs in up to 50 percent of affected patients.[6]

The pathogenesis of IgAN is uncertain and its aetiology is unknown. Abnormal immune regulation results in formation of IgA containing immune complexes characterized by their high affinity for the mesangium.

The immuno-histological aspects of IgAN are characterized by mesangial proliferation and mesangial deposits of IgA.

The clinical presentation of IgAN is with frank haematuria, often recurrent, following an upper respiratory infection. However, the majority of patients are diagnosed following an evaluation for asymptomatic microscopic haematuria and/or mild proteinuria.[7]

The diagnoses requires a kidney biopsy, wich provides prognostic information.

IgAN presentation with nephrotic syndrome is rare and usually associated to endocapillary proliferation, segmental sclerosis, and crescent formation.

On the other hand patients with nephrotic syndrome associated with mild IgAN seem to have a better prognosis.[3]

These patients have a dual glomerulopathy with superimposed MCD.

Only electron microscopy can effectively document diffuse foot process effacement and make the diagnoses of superimposed MCD in these cases.

It seems to be a growing evidence that proteinuria in itself is not an independent worse prognostic factor.[5]

A recent cohort established a clear clinical-pathological difference between IgAN-MCD and IgAN without MCD.[4]

Almost all patients with IgAN-MCD in this cohort (n=247) had a complete response to steroids and none progressed to ESRD.[4]

Our case highlights the importance of renal biopsy in IgAN presenting with nephrotic syndrome. We recommend performing EM on all this cases, as the patients have improved prognosis with steroid therapy and extremely unlikely progress to ESRD.

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Conflict of interest statement: None declared

Figure 1- Mild mesangial proliferation

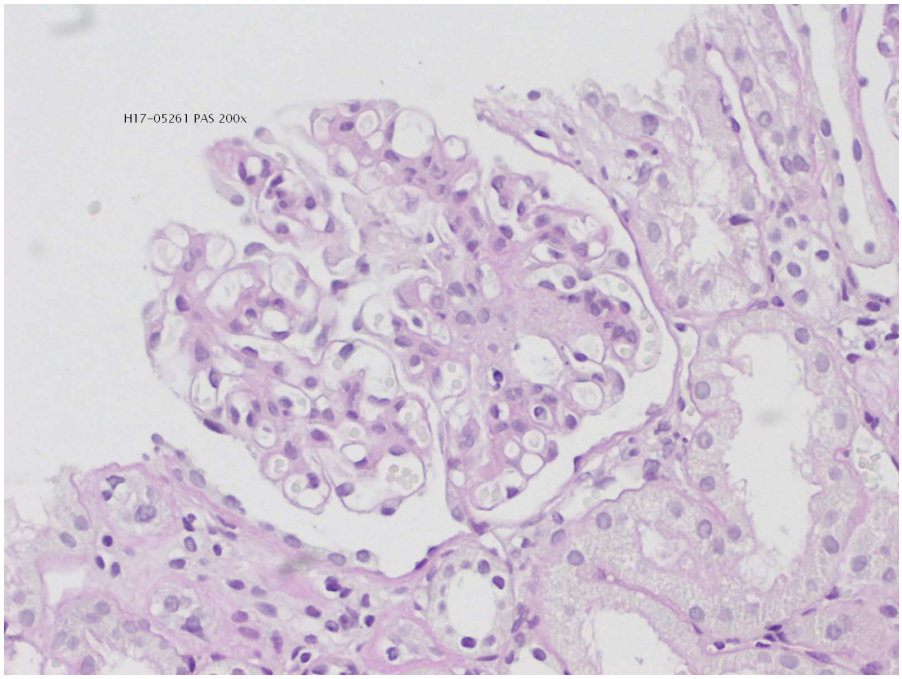


Figure 2- Segmental mesangial granular deposits of IgA (+++).

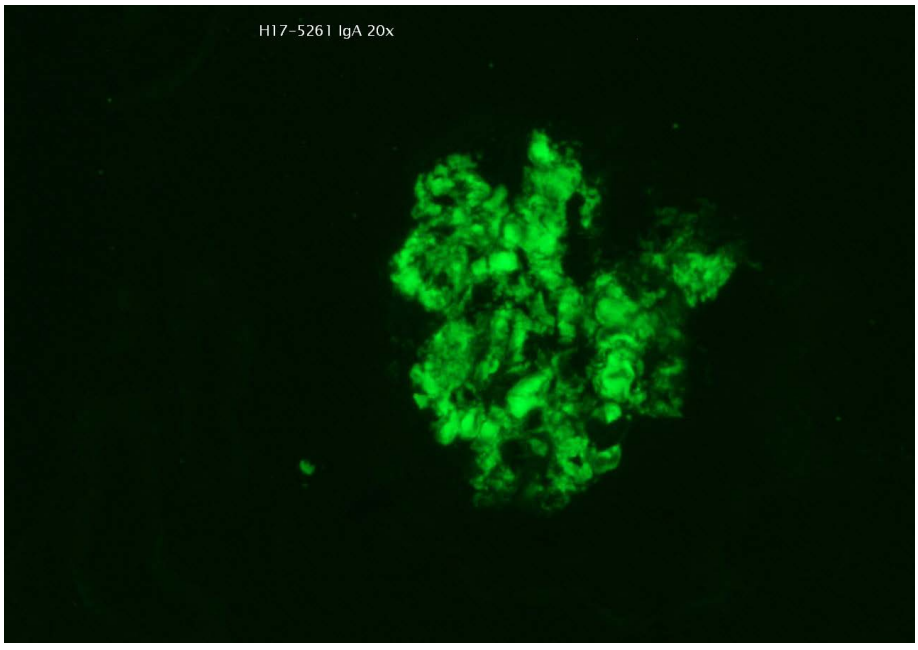
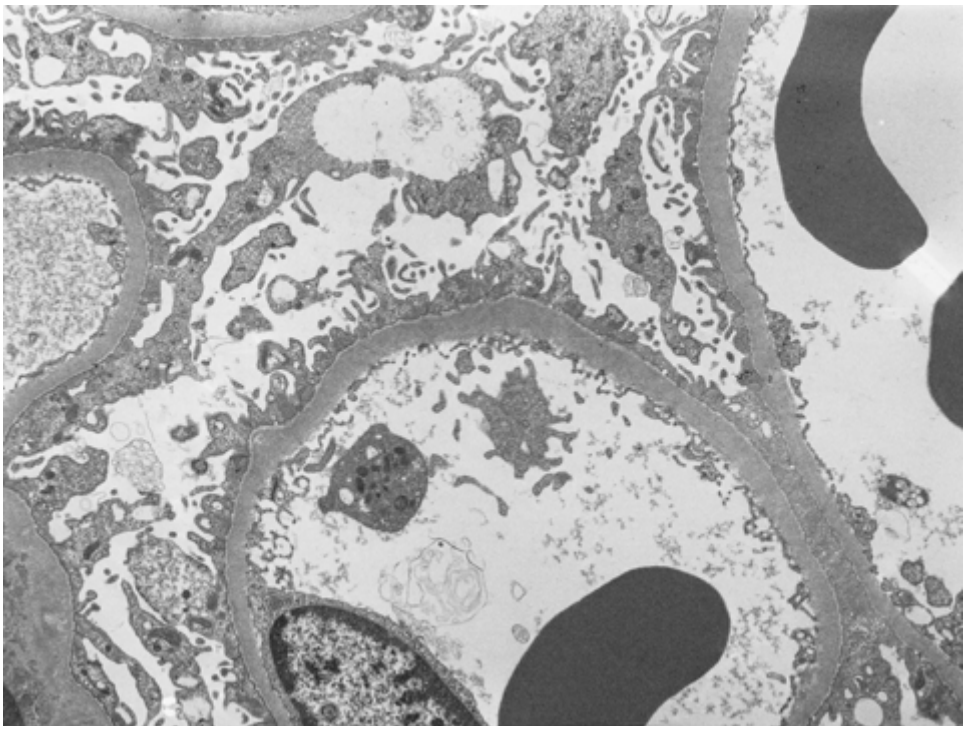


Figure 3- Diffuse foot process effacement over 50% of the capillary surface area involved (EM 4000x).



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