

## Case Report

# IgA nephropathy in a patient presenting with scleritis

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### Abstract

Scleritis is a very uncommon manifestation in patients with IgA nephropathy. Here, we report the case of a patient presenting with diffuse anterior scleritis in which the laboratory disclosed microscopic haematuria and nephrotic range proteinuria. Renal function was normal. Serology for lupus, vasculitis and cryoglobulinaemia was negative. Rheumatoid factor was negative, and serum C3 and serum C4 were on the normal range. Serology for human immunodeficiency virus types 1 and 2, hepatitis B, hepatitis C, syphilis, and Lyme disease was also negative. A renal biopsy was performed and revealed IgA nephropathy. Oral steroids were then started, and 6 months later, the patient was asymptomatic. Scleritis did not recur, and ophthalmologic examination was normal; however, proteinuria was still in non-nephrotic range. Renal function still remains normal.

**Keywords:** IgA nephropathy; scleritis

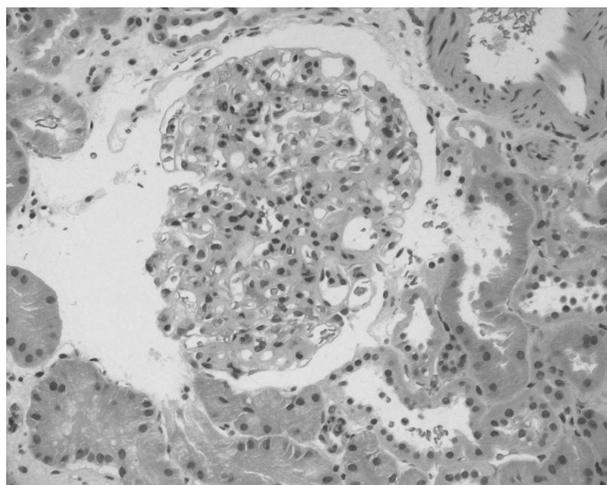
### Introduction

IgA nephropathy (IgAN) is considered to be the most common form of glomerular disease in the world [1]. Although IgAN is clinically restricted to the kidneys in most cases, there are associations with other conditions, particularly with a number of immune and inflammatory diseases, commonly rheumatic (i.e. ankylosing spondylitis, rheumatoid arthritis and Reiter syndrome), gastrointestinal (i.e. celiac disease), hepatic (i.e. alcoholic and non-alcoholic liver disease, and schistosomiasis), pulmonary (i.e. sarcoidosis), and cutaneous (i.e. dermatitis herpetiformis) [2]. Human immunodeficiency virus infection and hepatitis B (in endemic areas) have also been associated with IgAN [2]. Ocular involvement in patients with IgAN is infrequent, and the most common association occurs with uveitis [2]. Reports of the association between scleritis and IgAN are very scarce [3–5]. Here, we report the case of a patient presenting with scleritis, in which the laboratory disclosed micro-

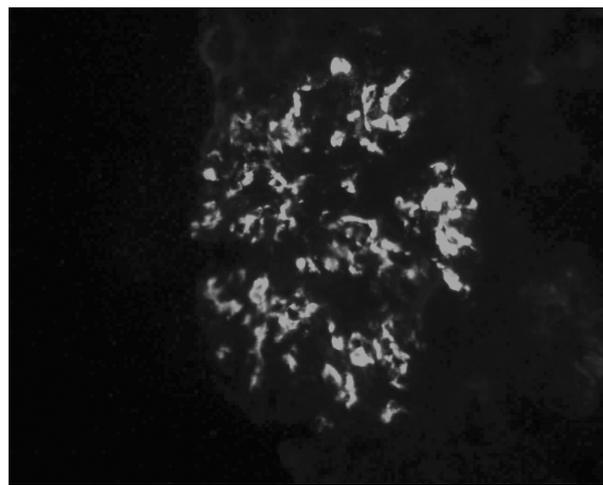
scopic haematuria and nephrotic range proteinuria, and renal biopsy revealed IgA nephropathy.

### Case report

A 34-year-old Caucasian female with prior history of hypertension was admitted at the Department of Ophthalmology of our hospital for bilateral red eye. She had no previous ocular trauma or surgery, and no other complaints. Ophthalmological examination showed bilateral diffuse anterior scleritis. Visual acuity was preserved. She was treated with subconjunctival injection of steroids with good result. On admission, blood pressure was of 150/100 mmHg, and heart rate was of 72 beats per minute. Ear, nose and mouth examination was normal, and cardiac, pulmonary, abdominal, neurological and lower limbs examination also revealed no changes. Neither joint tenderness or effusion nor rash, nor peripheral lymphadenopathies were detected. There was no genital ulceration. The laboratory disclosed normal renal function (creatinemia 1.18 mg/dL; uremia 40 mg/dL) and normal haemoglobin (12.9 g/dL). Urinalysis featured microscopic haematuria (150 erythrocytes/micro-litre) and proteinuria of 3.59 g/day. Serum albumin was 3.4 g/dL, and there was evidence of high total cholesterol level (total cholesterol, 254 mg/dL). Serum protein electrophoresis, serum protein immunoelectrophoresis, hepatic function tests, erythrocyte sedimentation rate and C-reactive protein were normal. Serology for lupus (antinuclear, anti-double strand deoxyribonucleic acid, anti-Smith, extractable nuclear and anti-ribonucleoprotein antibodies) and vasculitis (anti-neutrophil cytoplasmic antibodies) as well as the search for cryoglobulins was negative. Rheumatoid factor was negative, and serum C3 and serum C4 were on the normal range. Serology for human immunodeficiency virus types 1 and 2, hepatitis B, hepatitis C, syphilis, and Lyme disease was also negative. Chest X-ray was normal. Renal ultrasound revealed normal-sized and normoechoic kidneys, normal differentiation, and no hydronephrosis. A kidney biopsy was performed, and revealed diffuse mesangial hypercellularity (Figure 1) and diffuse granular



**Fig. 1.** Kidney biopsy showed diffuse mesangial hypercellularity (haematoxylin-eosin,  $\times 300$ ).



**Fig. 2.** Immunofluorescence microscopy showed diffuse granular mesangial deposits of IgA ( $\times 400$ ).

mesangial deposits of IgA (Figure 2). According to these, the diagnosis of IgAN was established. She started oral prednisolone (1 mg/kg/day), lisinopril (20 mg/day), losartan (50 mg/day) and simvastatin (20 mg/day). Six months later, she was asymptomatic, and no other episodes of scleritis occurred. Ophthalmological examination was normal. Proteinuria decreased but still persisted in non-nephrotic range (2.3 g/24 h). Renal function still remains normal.

## Discussion

Although IgAN is clinically restricted to the kidneys in most cases, there are associations with other conditions, particularly with a number of immune and inflammatory diseases, commonly rheumatic (i.e. ankylosing spondylitis, rheumatoid arthritis and Reiter syndrome), gastrointestinal (i.e. coeliac disease), hepatic (i.e. alcoholic and non-alcoholic liver disease, and schistosomiasis), pulmonary (i.e. sarcoidosis), and cutaneous (i.e. dermatitis herpetiformis) [2]. Human immunodeficiency virus infection and hepatitis B (in endemic areas) have also been associated with IgAN [2]. Ocular involvement in patients with IgAN is infrequent, and the most common association occurs with uveitis [2]. Our case describes the association of scleritis with IgAN. Reports of the association between scleritis and IgAN are very scarce [3–5]. Nomoto *et al.* [3] followed up 113 patients with various types of primary glomerular diseases for 1–33 months and verified that, of those patients studied, six patients exhibited scleritis. All of these six patients with scleritis were identified as having IgAN. Importantly, none of the patients other than those with IgAN had scleritis during the study period.

Scleritis is a scleral inflammatory disease that may also involve the cornea, adjacent episclera and underlying uveal tract. Scleritis is associated with a systemic disease in ~50% of cases [6]. The most common association is with rheumatoid arthritis. A number of other systemic disorders are also associated with scleritis, such as systemic lupus

erythematosus, relapsing polychondritis, systemic vasculitis, sarcoidosis and inflammatory bowel disease. In addition, infectious diseases (i.e. syphilis, Lyme's disease and herpes zoster) can also be the aetiology of scleritis [7]. Taking into account the association between scleritis and systemic diseases, a careful and complete examination of other organs should be carried out. In our patient, the absence of symptoms and signs of other organs involvement made the diagnosis of systemic disorder unlikely. On contrary, the presence of microscopic haematuria and nephrotic range proteinuria moved our attention towards a glomerular disease, and as such, a kidney biopsy was performed and revealed IgAN.

Scleritis can occur in association with IgAN. In patients with scleritis and asymptomatic urine abnormalities, IgAN should be considered and properly investigated. We hypothesize that abnormalities of the IgA immune system similar to the IgA nephropathy may be involved in the development of scleritis [3]. As described for episcleritis associated with IgAN, in which large numbers of dimeric IgA-secreting plasma cells were observed in the episcleral tissue [8], we speculate that ocular IgA could also display an important role in the pathogenesis of scleritis in patients with IgAN.

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*Conflicts of interest statement.* None declared.

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