



Clinical Group

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### Case Report

## IgA Nephropathy Precedes the Onset of the Rheumatic Disease in a Female with Ankylosing Spondylitis

### Case Report

IgA nephropathy (known as Berger's disease) is the most common cause of glomerulonephritis. Its estimated prevalence between 25–50 cases per 100,000 individuals. Gross hematuria (40–50%) and microscopic hematuria (30–40%) are the most common findings which can be related to upper respiratory tract or gastrointestinal infection. Between 15–40% of affected individuals may progress to chronic renal failure [1–5]. The definitive diagnosis is made by renal biopsy findings which is characterized by mesangial IgA deposition. Although the condition is a limited non-systemic renal disease, some systemic diseases are sporadically associated with mesangial IgA deposition. Henoch-Schönlein purpura (HSP), systemic lupus erythematosus (SLE), hepatitis, dermatitis herpetiformis, celiac disease and ankylosing spondylitis (AS) have been closely linked to the IgA nephropathy [6,7].

The true prevalence of IgA nephropathy in AS is not yet known however, recent reports concluded a prevalence of hematuria (up to 20%) of patients, whereas histopathologically confirmed IgA nephropathy is present in about 2% [8]. Although AS tends to occur predominantly in men (3–4:1), the previous reports surprisingly described IgA nephropathy as exclusively occurring in the male population of AS, and following the course of the established rheumatic disease. Only one report in the literature had described IgA nephropathy in a female with AS [9]. In this report, we describe another female in whom the onset of AS occurred after the renal manifestations of the IgA nephropathy.

A- 31 year old Indian and a mother of one child was referred by the orthopedic surgeon for further evaluation of a 1 year long problem of back and heel pain that has worsened recently. The pain was worse in the early hours of the morning. At the age of 29 she was managed by the nephrologist for episodic microscopic

and gross hematuria, and subnephrotic proteinuria. The patient was neither hypertensive nor had a history of passing urinary stones, urinary tract infection, GI complaint, skin rash or a history suggestive of hepatitis or connective tissue disease (CTD). In addition to the hematological and biochemical work ups she underwent renal biopsy that furnished the diagnosis of IgA nephropathy. The immunofluorescent histology revealed eight glomeruli one of which was globally sclerotic, there was 3+ granular global mesangial positivity for IgA with negative IgG, trace IgM, negative C1q, C3 (3+), negative albumin, negative fibrinogen, trace kappa, and lambda(3+). She was commenced on ramipril since, with fairly reasonable response. The hematuria lessened significantly from 20–30 to 8–10/HPF and proteinuria from 956 to 785 mg /24 hours. At any point in time the renal function (serum urea and creatinine) remained within normal values. Likewise were the serum Ca, P, Na, K, CL, uric acid, albumin, glucose and the lipid profile. The MRI lumbar scan then showed focal right foraminal protrusion of L3–L4 and L4–L5 disc lesion, hemangioma in L2 vertebral body.

Our clinical evaluation revealed positive Faber's test along with positive Schober's test of 3 cm, chest expansion of 4 cm and tender heels. Other systems examination was unremarkable. Plain x-rays of the lumbosacral spine and sacroiliac joints (SIJ) (Figure 1a,b demonstrates grade II–III sacroiliitis) along with loss of lumbar lordosis and bilateral calcaneal spurs (Figure 2). The MRI of SIJ was also consistent with sacroiliitis and sclerosis (Figure 3). The family history was negative for same condition. The latest biochemistry and hematology work ups were within normal values including CRP of 4.22 mg/dL (N <5) and ESR of 8 mm/HR. Serum ANF, Hepatitis B surface antigen, RF, cyclic citrullinated peptide (CCP) antibody were negative. Both of anti-ds-DNA antibody and serum complement components were normal. The serum IgA concentration was raised at 460 mg/dl (N 61–356 mg/dl). HLA B27 was negative. Reports on the concomitance between the IgA nephropathy and spondyloarthropathy (SpA) began to appear in the literature since the mid-seventies [10,11–13], indicating that individuals with SpA are more prone to develop IgA nephropathy than the general population. Its association with other seronegative

spondyloarthropathies such as Behcet's disease, psoriatic arthritis, Reiter's syndrome and the postenteric arthritides have been previously reported [5,6,10,12,13].

IgA nephropathy and AS are believed to share a common etiopathogenic mechanism. This mechanism involves the decreased expression of the receptor responsible for the clearance of IgA and its immune complexes on the surface of

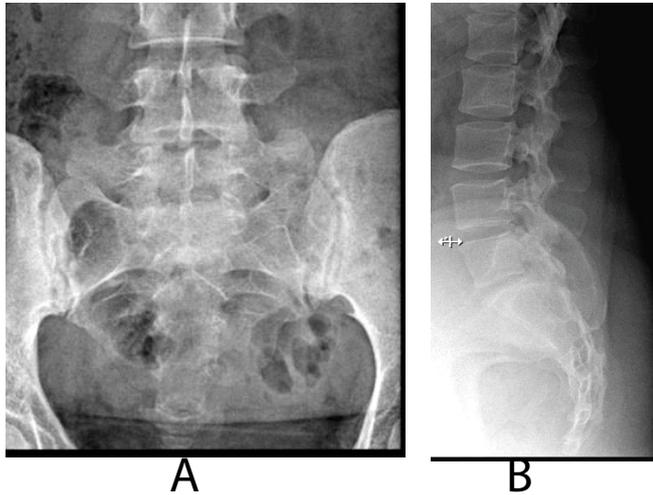


Figure 1a,b: Showing Grade II & III sacroiliitis & straightening of the lumbosacral spine.



Figure 2a,b: Demonstrating bilateral calcaneal spur.

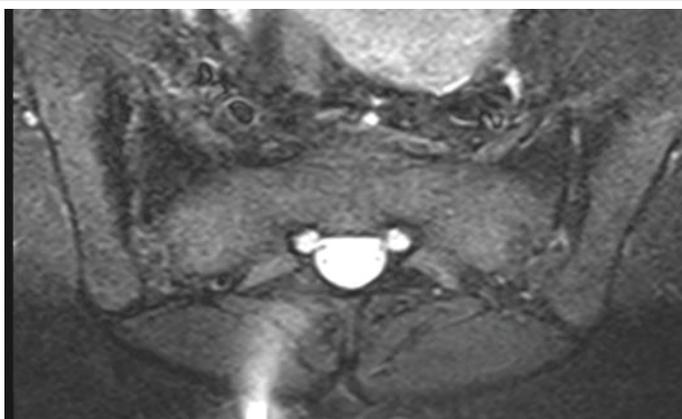


Figure 3: Axial STIR image showing bilateral and symmetric sacroiliitis Grade II-III, with subchondral sclerosis and shallow erosions. No present inflammatory activity.

monocytes and neutrophils [12,14]. Interestingly, all the cases previously reported were of males and perhaps in accordance with the fact that the prevalence of AS in men is higher than that in women. Most were B27 positive [3,5,9,11,12,15]. The first case a female with AS/IgA nephropathy reported earlier was in fact a B27 positive patient [9].

Our patient with renal biopsy proven IgA nephropathy had clinical and radiological sacroiliitis and calcaneal spurs. She met the diagnosis of definite AS according to New York criteria [16]. This would make the patient as the second reported women with such concurrence in the English literature, yet of interest her renal features had manifested earlier than the usual rheumatic disease and differed from the previous one of being B27 negative.

This could suggest that there is a subset of AS patients in whom the concurrence of IgA nephropathy is not HLA- B27 associated or a male-gender specific. Rheumatologists and nephrologists' alike need to be aware of the presence of such association in a subset of females with HLA B27 –negative AS.

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