Kimura's Disease Presenting as Proliferative Glomerulonephritis with Complete Heart Block

Sir,

We report this case of Kimura's disease in a 35-year-old Indian male presenting with pleural effusion and nephrotic–nephritic Syndrome. Unique features in the case are that it is first reported case presenting simultaneously with diffuse proliferative glomerulonephritis and complete heart block.

This case presented with progressive swelling of feet and breathlessness on exertion of 2 weeks duration not associated with oliguria, frothuria, hematuria, joint pain, and rash. On examination, his weight was 62 kg, and his blood pressure was normal. He had anasarca and right pleural effusion. The rest of the physical examination was unremarkable. There was a history of painless cervical lymphadenopathy 1½ years back, biopsy of which showed reactive lymphoid tissue with thickened capsule along with many eosinophils consistent with Kimura's disease. He was initiated on steroids and showed marked improvement within 3 months. The patient developed complete heart block requiring permanent pacemaker insertion 1 month before presenting to us. His hemoglobin was 10.5 g/dl, white blood cell was 9800/mm^3 with 36% eosinophils, and absolute eosinophil count of 3528/mm^3; serum IgE levels were high (1210.0 IU/ml), urea was 76 mg/dl, and creatinine was 2.9 mg/dl. Urine examination showed albumin 3+ and full-field red blood cells. The 24 h urine protein was 3.7 g. Mantoux test was negative; chest X-ray showed mild-to-moderate right-sided pleural effusion which was exudative with eosinophil predominance. High-resolution computed tomography chest showed moderate pleural effusion. Serum C3 was very low and C4 was normal. Angiotensin-converting enzyme and immunological tests (antinuclear antibodies, anti-dsDNA

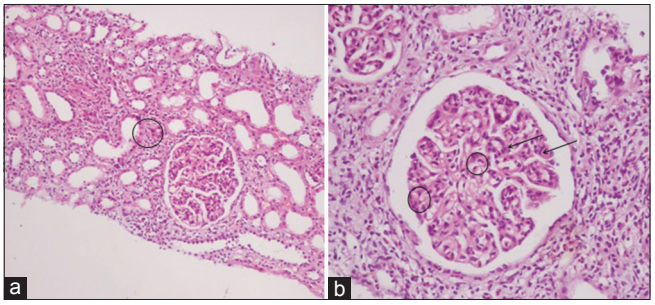

Figure 1: (a) Low power showing endocapillary proliferation (black arrow), densely mixed interstitial infiltrate (H and E, ×100), (b) high power view showing endocapillary proliferation (black arrow), infiltration by neutrophils and eosinophils, interstitium shows eosinophils (circles) (H and E, ×200)

antibodies, antiextractable nuclear antigens, antineutrophil cytoplasmic antibodies, and rheumatoid factor) were normal. Coomb's test was negative. Total cholesterol was 260 mg/dl, low-density lipoprotein cholesterol was 102 mg/dl, high-density lipoprotein cholesterol was 38 mg/dl, and triglyceride 285 mg/dl. Stool examination was normal. Parasitic serology, serologic tests for human immunodeficiency virus (HIV), hepatitis B virus, hepatitis C virus, syphilis, and QuantiFeron were negative.

Renal biopsy showed diffuse proliferative glomerulonephritis characterized by global endocapillary proliferation without any crescents. Interstitium showed dense inflammatory eosinophils infiltrate [Figure 1a and b]. Immunofluorescence study showed deposition of IgG, IgM, C3, and C1q along glomerular capillary walls. His old lymph node biopsy was reviewed, which showed features of Kimura disease [Figure 2a and b]. Prednisolone was initiated at the dose of 1 mg/kg. Pleural effusion, heart block, and eosinophilia improved and creatinine also decreased to 1.2 mg/dl within 10 days of steroid initiation. Steroid was continued with the same dose for 3 months. His proteinuria decreased gradually and then disappeared. Thereafter, steroids were tapered, and he was followed up in outpatient department for 2 years without any relapse.

Kimura's disease is a benign, chronic inflammatory soft tissue disorder of unknown origin. Most cases of Kimura's disease have originated in China, Japan, or Southeast Asia, and the disease is uncommon in Caucasians and rare in Africans. There is a marked male predominance. The peak age of onset is during the third decade of life. Kimura's disease was first described in 1937 in the Chinese literature by Kimm and Szeto and initially recognized as "eosinophilic hyperplastic lymphogranuloma." The definitive histological description was published by Kimura *et al*. in 1948, and thus, the disease has borne the author's name. Since that time, there has been a gradual increase in the number of reports of the disease.

The pathophysiology of Kimura's disease remains unclear. Some of the hypothetic triggers may be an infection or toxin, which initiates autoimmune phenomenon. Other studies have shown elevated granulocyte-macrophage colony-stimulating factor, tumor necrosis factor-alpha, soluble interleukin (IL)-2 receptor, IL-4, IL-5, IL-10, and IL-13.[1]

An estimated 12%–16% of patient with Kimura's disease exhibit proteinuria, of which 59%–78% have nephrotic syndrome. Yuen *et al*. published eight cases of Kimura's disease of age 9–35 years with nephropathy from 1998 to 2004.[2] Available literature shows that the Renal histology patterns reported are IgA nephropathy, membranous glomerulonephritis, minimal change glomerulonephritis, IgM nephropathy, and mesangial proliferative glomerulonephritis. A recent case report was published from India of mesangial-proliferative glomerulonephritis

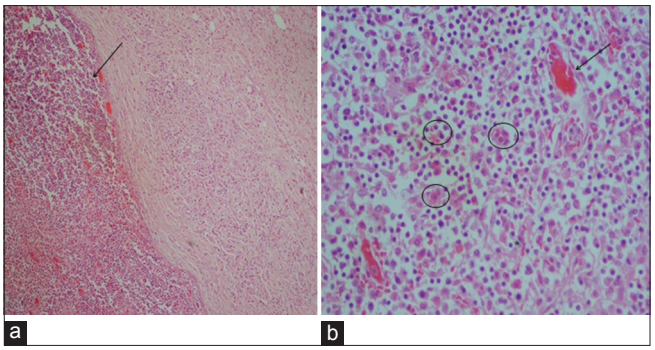

Figure 2: (a) Histopathological image shows lymphoid tissue with follicular hyperplasia (arrows); (H and E, ×400), (b) image shows eosinophilic infiltration (circles) vessels with flattened endothelial lining (arrows; H and E, ×400)

in a patient with Kimura's disease presenting as nephrotic syndrome.[3] To the best of our knowledge, diffuse proliferative glomerulonephritis has not been reported in patients with Kimura's disease till date. We observed prompt response to steroids in our patient. Goel *et al*. also observed gradual improvement in symptoms and complete resolution of lymphadenopathy and pleural effusion poststeroid initiation in a 60-year-old male.[4]

We could find only one case report of Kimura's disease with complete heart block. Horigome *et al*. described a 13-year-old boy with hypereosinophilia associated with Kimura's disease, which showed repeated life-threatening syncopal attacks during daily activities or at rest. Coronary arteriography demonstrated small aneurysms with irregular vessel walls of both coronary arteries and the absence of organic stenotic lesions. Recent articles also showed acute necrotizing eosinophilic myocarditis, the most severe form of eosinophilic myocarditis, is generally fatal, and rarely complicated by complete atrioventricular block and responds well to the steroid treatment.[5]

Management strategies range from conservative observation for asymptomatic patients to surgical excision, steroid therapy, or radiotherapy for symptomatic patients Xiao-Rong Ma *et al*. published a case report showing leflunomide as a therapeutic modality for patients who is unresponsive to corticosteroids or with frequently relapsing disease.

One must think for Kimura's disease when an Asian patient presents with nephropathy, skin lesions, eosinophilia, and heart block. Prognosis of renal improvement is good with steroid treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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