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Case Report

Remission of secondary membranous nephropathy in a patient with Kimura disease after surgical resection

Sunhwa Lee [1], Yong Jin Yi [1], Hyung Ah Jo [1], Hyuk Huh [1], Kyung-Hwan Kim [2], Dong Ki Kim [1], Hajeong Lee [1,*]

[1] Department of Internal Medicine, Seoul National University Hospital, Seoul, Korea
[2] Department of Thoracic Surgery, Seoul National University Hospital, Seoul, Korea

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Abstract

Kimura disease (KD) is an eosinophilic, granulomatous, benign, chronic inflammatory disease with an unknown etiology. A 33-year-old woman visited our hospital because of a palpable, left subclavian mass, a left scapulo-anterior pseudoaneurysm, and nephrotic syndrome. Her subclavian lymph node biopsy examination result was consistent with KD, and results of a renal biopsy indicated secondary membranous nephropathy. After renal histological examination confirmed nephropathy, treatment with prednisolone and cyclosporine was initiated, which was maintained for over 1 year. However, this therapy only provided a transient improvement in proteinuria. One year after commencing the treatment, both proteinuria and azotemia aggravated as the left axillary mass doubled in size. Finally, the mass was surgically excised, following which the azotemia rapidly normalized and proteinuria resolved within 1 month. This case shows that tumor resection in a patient with KD with secondary nephropathy may resolve secondary renal manifestations. Furthermore, reversible renal dysfunction may be caused by unknown secreted molecules.

Introduction

Kimura disease (KD) presents as a subcutaneous mass, most frequently occurring in the head and neck regions. The disease is characterized by a benign, angiolymphomatous proliferation with eosinophilic infiltration. The histological characteristics of the disease are hyperplastic lymphoid tissue that contains well-developed lymphoid follicles, marked infiltration of eosinophils, vascular proliferation, and fibrosis. It is also associated with the development of renal disease, especially nephrotic syndrome [1]. Previously, surgical resection was the treatment of choice. However, treatments with steroid and immunosuppressive agents were also reported to produce beneficial effects [2], although discontinuation of steroid often results in recurrence of the masses [3]. In patients with renal manifestations, however, surgical resection is usually not the first-choice treatment. We herein report the case of secondary membranous nephropathy (MN) associated with KD which recovered after surgical resection of the mass.

Case report

A 33-year-old woman presented with a soft-tissue mass in her left axillary area. A percutaneous biopsy was performed, and histological findings showed atypical lymphoid hyperplasia with many eosinophilic infiltrations, which are consistent with KD (Fig. 1). Neither proteinuria nor hypereosinophilia was noted. One-month treatment with oral prednisolone could not

* Corresponding author. Division of Nephrology, Department of Internal Medicine, Seoul National University Hospital, 101 Daehak-ro, Jongno-gu, Seoul 110–744, Korea.
E-mail address: dewyhj@hanmail.net (H Lee).

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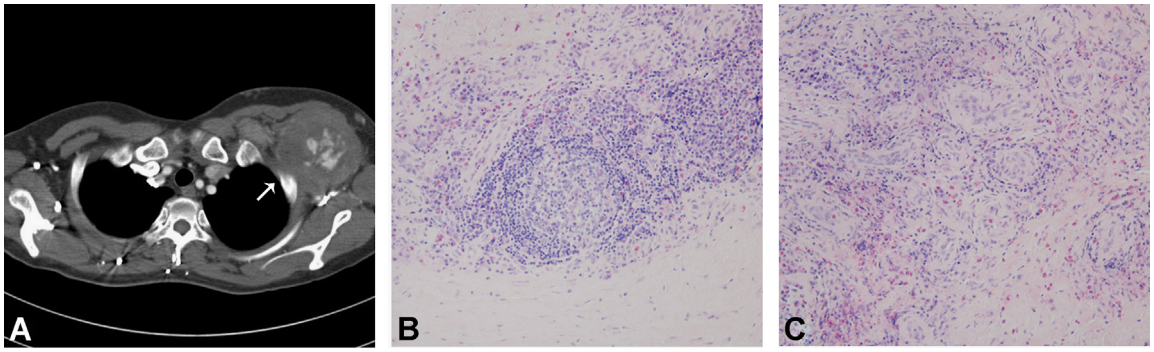

Figure 1. Left subclavian mass consistent with Kimura disease. (A) A left scapulo-anterior pseudoaneurysm in the left axillary area (arrow). Light microscopic images of hematoxylin and eosin–stained sections of the subclavian lymph node biopsy specimen show (B) a well-developed lymphoid follicle with eosinophilic infiltration and (C) vascular proliferation (200 ×).

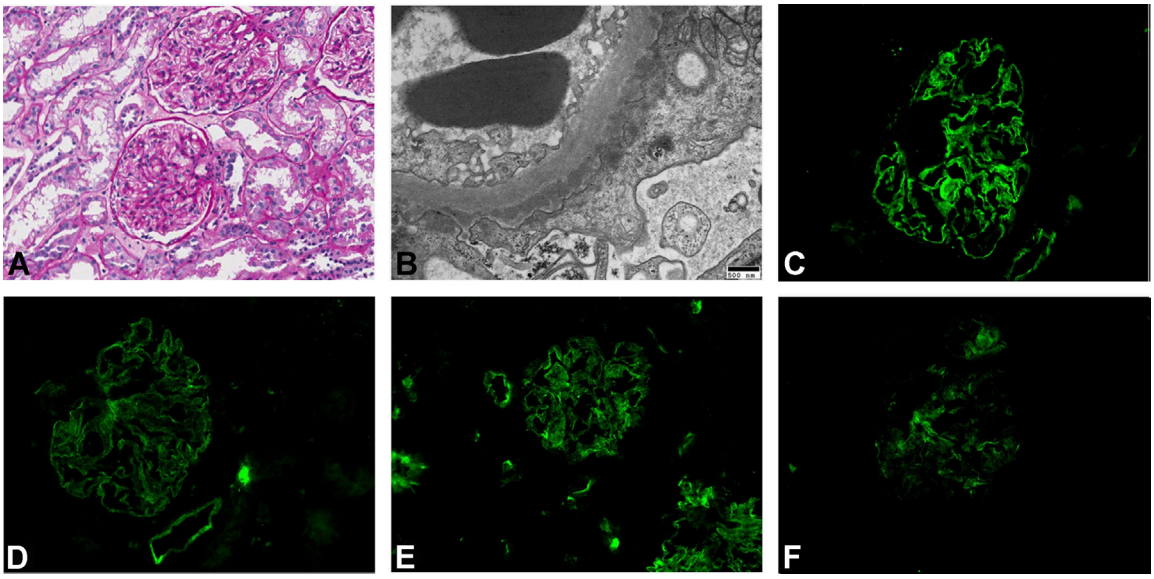

Figure 2. Renal biopsy specimens diagnosed with membranous nephropathy. (A) Light microscopic image of a biopsy specimen shows hypercellular mesangial cells (Hematoxylin and eosin-stained, 200 ×). (B) An electron microscopic image of a similar specimen reveals electron-dense deposits in some subepithelial areas and diffuse foot process effacement (20,000 ×). Using immunofluorescence microscopy, granular deposits of (C) IgG, 3+, (D) IgM, 2+, (E) C3, 3+, and (F) C1q, 1+ were detected in the mesangium (400 ×). Ig, immunoglobulin.

diminish size of the mass. One year after the diagnosis of KD, a 6-cm, left scapulo-anterior pseudoaneurysm was noted (Fig. 1), which required a stent-graft insertion. Following the graft insertion, the size of pseudoaneurysm decreased. Hypereosinophilia (12,450/mm^3) was noted during this period. Treatment with prednisolone was continued for 1 year, and eosinophilia gradually improved to < 500/mm^3 within this period.

One and half year after the diagnosis of KD, the patient presented with a month-long history of foamy urine and a weight gain of 5 kg; pitting edema of both lower extremities was also observed.

Upon admission, the patient's blood pressure was 98/61 mmHg, white blood cell (WBC) count was 7,200/mm^3 (with 33.8% eosinophils), and hemoglobin and platelet levels were 14.5 g/dL and 287,000/mm^3, respectively. Serum blood urea nitrogen and creatinine levels were within the normal range, with a creatinine clearance rate of 120 mL/min/1.73 m^2. Other laboratory assessments showed normal electrolyte, low serum albumin (1.3 g/dL), and high cholesterol (521 mg/dL) levels; complement proteins (C3 and C4) and immunoglobulins (Ig) A, G, and M were within normal limits. The patient's virology profile was negative for hepatitis B and C, and her immunologic status was negative for anti-double-stranded DNA, antistreptococcal antibodies, cryoglobulins, and rheumatoid factor. Urinalysis was 4+ positive for protein, with five to six red blood cells and five to nine WBCs per high-power field. The random, spot urinary protein-to-creatinine ratio (uPCR) was 16.6 g/g, and urine protein electrophoresis showed nonspecific proteinuria.

A light microscopic examination of renal biopsy specimens showed diffuse thickening of the capillary walls, focal, mild hypercellular mesangial cells, and normal-sized glomeruli. An electron microscopic examination revealed electron-dense deposits in some subepithelial areas and diffuse foot process effacement. An immunofluorescence microscopic examination revealed granular deposits of IgM, IgG, C3, and C1q in the mesangium (Fig. 2). Overall, the MN was diagnosed as Ehrenreich and Churg Stage III. Angiotensin receptor blocker or angiotensin-converting enzyme inhibitor was considered the first-choice treatment. However, the patient's blood pressure was too low to add antihypertensive drugs to the treatment