CASE REPORT

Kimura's disease revisited: report of a case with a clinical and cytohistological correlation

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SUMMARY

Kimura's disease (KD) is a rare chronic inflammatory disorder of unknown aetiology. The common mode of clinical presentation is in the form of painless subcutaneous nodules usually seen in the head and neck region and is associated with regional lymphadenopathy and occasional involvement of the major salivary glands. It is often accompanied by peripheral eosinophilia and markedly elevated serum IgE levels. Histologically, it is characterised by reactive lymphoid follicles with eosinophilic infiltration, sometimes forming eosinophilic abscesses and prominent postcapillary venules. There is no consensus about optimal treatment of this disease in the reported literature. This condition seldom resolves spontaneously but the prognosis is good. Malignant transformation has not been reported. Early diagnosis of KD could spare the patient unnecessary and potentially harmful diagnostic procedures. A high index of clinical suspicion can help in diagnosing this disease and aid in better management.

BACKGROUND

Although Kimura's disease (KD) is a rare chronic inflammatory disorder of unknown aetiology, it is important to keep this diagnosis in mind in young men with subcutaneous nodules in the head and neck region as the prognosis is good in spite of no spontaneous resolution. Malignant transformation has not been reported. Early diagnosis of KD could relieve patient anxiety vis-à-vis other malignant

neoplasms with similar clinical presentations and spare the patient unnecessary and potentially harmful diagnostic procedures.

CASE PRESENTATION

A 26-year-old man presented to the surgical outpatient department of our institute with bilateral postauricular swellings for the past 6 months (figures 1 and 2).

These swellings were gradually increasing in size, painless and associated with neither pruritus nor dermatitis. The patient had no history of prolonged fever, cough, loss of weight, ear discharge or contact with tuberculosis.

On examination, 4×2 cm lymph nodal masses were present in the bilateral postauricular region. These swellings were non-tender and firm in consistency, with normal overlying skin. Multiple sub-centimetric lymph nodes were palpable in the posterior cervical triangle of the left neck. There was neither axillary nor inguinal lymphadenopathy. Systemic examination was within normal limits.

Complete blood count of the patient showed peripheral eosinophilia (19.9% eosinophils). Chest X-ray was normal and Mantoux test negative. The erythrocyte sedimentation rate was 2 mm at the end of 1 hour. Renal function tests and routine and microscopic examination of urine did not show any abnormality. Serum IgE was markedly raised (2418.7 IU/mL, normal value ≤158.00 IU/mL). Ultrasonography of the neck showed multiple

Figure 1 Right postauricular swelling.

Figure 2 Left postauricular swelling.

CrossMark

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Figure 3 Leishman-stained smear (×4) showing markedly cellular smear with capillary channels.

Figure 5 Leishman-stained smear showing increased eosinophils (coarse red granular cytoplasm) (×10).

enlarged hypoechoic nodes in bilateral posterior triangles and bilateral parotid regions, with the largest measuring about 2.3×0.6 cm in the right postauricular region and 2.1×0.7 cm in the left postauricular region. A few enlarged nodes were also seen in the right side of neck (level II). Ultrasonography examination of the abdomen was within normal limits.

Fine-needle aspiration cytology findings

The patient was referred to the fine-needle aspiration cytology (FNAC) clinic. FNAC was performed on both postauricular masses as well as on the left posterior cervical lymph node, with a 23 G needle. A whitish, particulate to blood mixed aspirate was obtained from all sites. Smears were markedly cellular ([figure 3](#)) and showed a polymorphous lymphoid population with significant number of eosinophils, plasma cells, histiocytes, tingible body macrophages and several capillary fragments with prominent endothelial cells. Associated proteinaceous material was also noted ([figures 4](#) and [5](#)). No Reed-Sternberg cells and no parasitic fragments were present. An impression of Kimura's lymphadenopathy was given.

Subsequently, surgical excision was performed for both postauricular nodal masses. On gross examination, ovoid, elongated and encapsulated masses, each measuring 4×2×0.8 cm, were seen ([figure 6](#)). Microscopic examination of H&E-stained sections showed preserved lymph node architecture ([figure 7](#)) with

marked germinal centre hyperplasia and several eosinophilic abscesses ([figure 8](#)), numerous interfollicular eosinophils extending to the skeletal muscle, a few polykaryocytes of the Warthin-Finkeldey giant cell type ([figures 9](#) and [10](#)), extensive capsular and interstitial fibrosis and vascularisation of germinal centres. Increased paracortical plasma cells and mast cells and variably hyalinised blood vessels lined by prominent endothelial cells, were also present. Hence, a diagnosis of KD was established with relevant clinical details and specific histopathological features.

OUTCOME AND FOLLOW-UP

The patient was asymptomatic 1-year postsurgical excision.

DISCUSSION

KD is a rare chronic inflammatory disorder of unknown aetiology. It was first described, in 1937, in the Chinese literature, by ST Kimm and C Szeto, as an 'eosinophilic hyperplastic lymphogranuloma'.¹

This disease got its name after the histological description—'unusual granulation combined with hyperplastic changes of lymphatic tissue'—published by Kimura *et al*² in 1948.

The disease has a markedly male predominance with a male-to-female ratio of 3.5 to 7. Patients usually present in the third decade of the life.

Figure 4 Leishman-stained smear showing proteinaceous material and tingible body macrophages (×40).

Figure 6 Gross-examination showing node bisected along longitudinal axis.

Figure 7 Preserved lymph node architecture, H&E stain (×2).

The common mode of clinical presentation is in the form of painless subcutaneous nodules usually found in the head and neck region. These nodules may be associated with pruritus and dermatitis. The condition is commonly associated with regional lymphadenopathy and occasional involvement of the salivary glands (parotid and submandibular glands). It is often accompanied by peripheral eosinophilia and markedly elevated serum IgE levels.³

Primarily, it is a localised disorder, but sometimes may be associated with systemic manifestations. Renal involvement is the commonest systemic manifestation, affecting up to 60% of patients and presents as membranous glomerulonephritis, minimal change glomerulonephritis, diffuse proliferative glomerulonephritis, mesangial proliferative glomerulonephritis and also nephrotic syndrome.^{4–7}

Takao *et al* reported a case of a 60-year-old Japanese man who presented with severe numbness and coldness in his fingers, with gangrenous changes.⁸ This patient was diagnosed as having KD with thromboangiitis obliterans, which showed favourable response to systemic steroids.

This disease can affect the paediatric age group as well. Kumar *et al*⁹ reported KD in a 3-year-old child, manifesting in the form of painless axillary lymphadenopathy. Ranka *et al*¹⁰ reported KD in a 12-year-old child who presented with a unilateral cervical mass.

Figure 8 Eosinophilic microabscesses, H&E stain (×40).

Figure 9 Prominent post capillary venules with eosinophils and Warthin-Finkeldey giant cell, H&E stain (×40).

Histopathological confirmation includes presence of constant and frequent features as well as some that are rarely noted.⁶ Constant features are preserved nodal architecture, florid germinal centre hyperplasia, eosinophilic infiltration and postcapillary venule proliferation. Frequent features include sclerosis, polykaryocytes of the Warthin-Finkeldey giant cell type, vascularisation and necrosis of the germinal centres, proteinaceous and reticular IgE deposition in the germinal centres and eosinophilic abscesses within the germinal centres. Rarely, progressive transformation of the germinal centres may be seen.

The management of KD is, to date, mired in controversy. Surgical excision is the first line of therapy, although relapse may occur.⁶ A systemic corticosteroid is indicated for relapses and for renal involvement.¹⁰ Intralesional steroids have been tried with variable degrees of success, in the reported literature. In the present case, we excised both postauricular swellings, as the patient was cosmetically concerned by their presence and simultaneously started him on Cetirizine (H-1 blocker) for 1 month. The patient was asymptomatic at 1-year follow-up. Radiation treatment is usually administered for local control of lesions that are not responsive to steroids and a total dose of 20–30 Gy has been proven effective. Irradiation should be considered in patients resistant to steroids and also in young patients in whom long-term side effects of steroids may be more deleterious.¹¹

Figure 10 Proteinaceous pinkish material and Warthin-Finkeldey giant cell, H&E stain (×40).

Learning points

- ▶ Kimura's disease is a rare chronic inflammatory disorder of unknown aetiology seen in young Asian men; it manifests as subcutaneous nodules in the head and neck region and is associated with cervical lymphadenopathy.
- ▶ In spite of no spontaneous resolution, the prognosis is good. Malignant transformation has not been reported.
- ▶ Surgical excision is the first line of treatment, although relapse may occur, for which systemic corticosteroids are indicated.
- ▶ Early diagnosis of Kimura's disease spares the patient unnecessary and potential harmful diagnostic procedures.

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