CASE REPORT

Kimura's Disease Mimicking Hodgkin's Lymphoma: An Unusual Cause of Parotid Swelling

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ABSTRAK

Penyakit Kimura adalah penyakit benigna dan kronik yang puncanya tidak diketahui. Ia biasanya berlaku pada pesakit berbangsa Oriental yang datang dengan ketumbuhan di bawah kulit di bahagian kepala dan leher yang boleh disalahdiagnosa sebagai tumor malignan yang memerlukan pembedahan radikal dan kemoterapi intensif. Ia telah dilaporkan di dalam makalah berbahasa China dan Jepun. Ia sukar didiagnosa sebelum laporan biopsi tisu manakala sitologi aspirasi jarum (*fine needle aspiration cytology*) juga tidak banyak membantu. Justeru itu, jika pakar patologi kurang berpengetahuan tentang penyakit ini, ianya akan disalah anggap sebagai lesi yang malignan. Kami laporkan di sini satu kes pesakit Melayu yang datang dengan ketumbuhan parotid yang pada mulanya didiagnos sebagai limfoma Hodgkin. Penyakit Kimura sepatutnya disyaki apabila seorang pesakit lelaki muda dari Asia yang datang dengan ketumbuhan di bahagian kepala dan leher yang tidak sakit, dan disertai oleh peningkatan sel eosinofil (*hypereosinophilia).*

Kata kunci: Penyakit Kimura, ketumbuhan parotid, limfoma Hodgkin, eosinofilia, limfadenopati servikal

ABSTRACT

Kimura's disease (KD) is a rare, benign chronic inflammatory disease of unknown aetiology, typically presents in the Orientals as subcutaneous masses in the head and neck region that could be easily misdiagnosed as a malignant tumour, leading to unnecessary radical surgery or intensive cytotoxic therapy. It has been mainly reported in the Chinese and Japanese literature. It is difficult to diagnose before tissue biopsy and fine needle aspiration cytology (FNAC) has limited value. Hence, unless the pathologists are aware of this entity, it might be mistaken as a malignant lesion. We encountered a case of KD in a Malay patient presenting as a parotid mass that was initially diagnosed as Hodgkin's lymphoma (HL). This disorder should be suspected in young male Asian patients presenting with a painless unilateral mass in the head and neck region with associated hypereosinophilia.

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Key Words: Kimura's disease, parotid swelling, Hodgkin's lymphoma, eosinophilia, cervical lymphadenopathy

INTRODUCTION

Kimura's disease was first described in 1948 as an unusual granulation combined with hyperplastic changes in lymphoid tissue (Day 1995). Since then, series have been reported from China, Hong Kong, Singapore and Indonesia, all in patients of Mongoloid race.

The pathophysiology of KD may relate to a disturbance in the normal rate of production of eosinophils and IgE resulting in excessive elaboration of eosinophilotrophic cytokines such as interleukin 4 (Day 1995).

KD is quite a rare condition. However, it might in fact be more prevalent than it seems for two reasons. First, most of the patients afflicted with this condition are asymptomatic and so they might go undiagnosed. Secondly, the lesions might have been misdiagnosed as other conditions; clinically it is often confused with parotid tumours with lymph node metastasis.

KD has the ability to mimic a number of benign inflammatory and neoplastic conditions of the head and neck including, Langerhans cell histiocytosis, Castleman's disease, Kikuchi's disease and follicular lymphoma. The most common physical manifestation is a slowly enlarging subcutaneous mass, often in the head and neck region involving the salivary glands with regional lymphadenopathy. It is usually associated with peripheral blood and tissue eosinophilia with markedly increased serum IgE levels (Irish 1994). FNAC and diagnosis is usually established on histopathological examination.

We describe a young boy who presented with a parotid mass due to KD, which was initially thought to be HL.

CASE REPORT

A 16-year-old Malay boy presented with a painless swelling over the right parotid region, which was slowly increasing in size over the past one year. He had no constitutional symptoms. There was no previous history of tuberculosis (TB). Physical examination revealed a mass over the right parotid region measuring 6 x 6 cm that was firm and non-tender. There were enlarged lymph nodes over the right cervical region measuring 4 x 4 cm. There were no other significant physical findings. Full blood count revealed eosinophilia (eosinophil count 3.5 x 10^9/l). The serum lactate dehydrogenase level and renal profile were within the normal range. Viral serology and TB work-up were negative.

A computed tomography (CT) of the neck showed an enhancing ill-defined diffuse mass and enlarged lymph nodes over the right mandibular region. The underlying subcutaneous tissue showed increased density suggestive of an inflammatory lesion. CT brain, thorax and abdomen did not show any abnormality. FNAC of the parotid mass was reported as HL, while trucut biopsy of the cervical lymph node showed reactive hyperplasia.

A right superficial parotidectomy and lymph node excision biopsy were performed, as isolated parotid lesion is an unusual presentation of HL. The lymph node tissues contained multiple lymphoid follicles and reactive germinal centres with marked perinodal fibrosis. Benign parotid tissues were noted surrounding the lymph nodes with periductal fibrosis, acinar atrophy and proliferation of endothelial venules. There were prominent eosinophilic infiltration and eosinophilic microabscesses within the nodal parenchyma

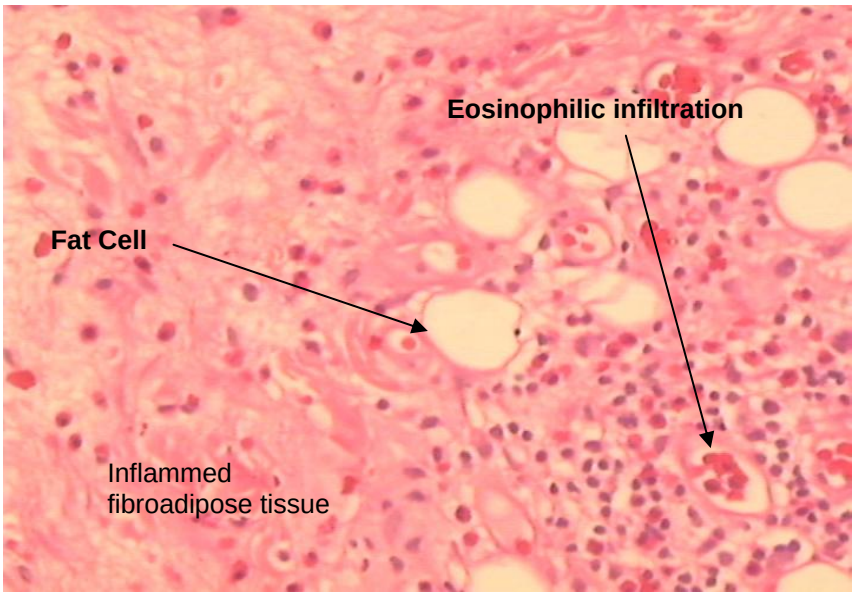

Fig. 1A: Low power view of section of right parotid mass showing dense eosinophilic infiltration within the nodal parenchymal and fibroadipose tissue.
(x 20 magnification; Haematoxylin & Eosin stain).

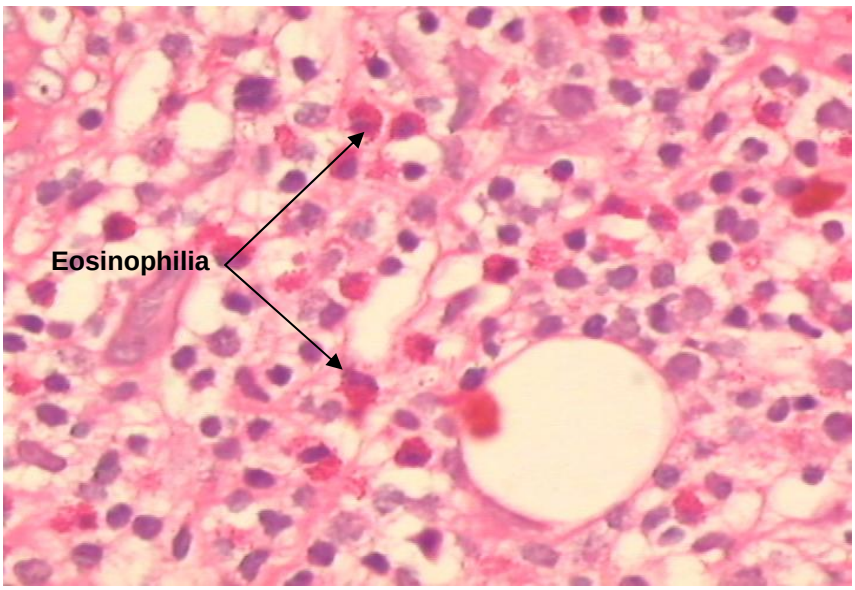

Fig. 1B: Higher magnification of Fig. 1A showing infiltration by eosinophils displaying orangeophilic cytoplasm and bilobed nucleus.
(x 100 magnification; Haematoxylin & Eosin stain).

and parotid tissue. Reed-Sternberg cells, giant cells and granulomas were not seen (Fig. 1A, 1B). The histopathological features were consistent with KD.

The patient was given oral prednisolone 60mg daily followed by a tapering dose over the next two weeks. At one-year follow-up (at the time of writing of this report), he remained disease free.

DISCUSSION

KD is an indolent, benign, but locally disfiguring disease. Its true importance lies in its ability to mimic a number of other benign inflammatory and neoplastic conditions of the head and neck. It is usually a localized process without systemic symptoms (Day 1995).

The most common causes of sub-acute or chronic cervical lymphadenitis, which should be considered in this case, are TB, Epstein Barr virus infection, toxoplasmosis, Kikuchi's disease and parotid tumours. The slowly enlarging mass and localised parotid involvement over a one year duration is rather unusual for HL. In addition, FNAC material alone is not conclusive for a definitive diagnosis of HL unless classical Reed-Sternberg cells are present. There were no radiological and microbiological evidence of TB and parotid involvement is unusual in TB. There was no laboratory evidence of viral or parasitic infection. The histological features which excluded Kikuchi's disease and malignancies.

Typical histological features in KD include preserved nodal architecture, florid germinal centre hyperplasia, eosinophilic infiltration, eosinophilic abscess, vascularization, necrosis of the germinal centres, proteinaceous deposition within germinal centres and fibrosis (Irish 1994). Specific histological features in HL include Reed-Sternberg cells and $CD30^+$ mononuclear cells were absent in the present case. Other differences between these two conditions are a) KD has no potential for malignant transformation where else HL is a haematological malignancy, b) KD occur in younger adults between 20-40 years old while HL has a bimodal age distribution, c) KD is commoner in Asians while HL is commoner in Caucasians, d) KD is normally a localized process with no systemic symptoms but HL has constitutional symptoms in 40% of patients, and e) IgE is elevated in KD but not in HL.

There are three major therapeutic options for KD. Resection of the tumour mass may be effective in permanently eradicating the mass if the entire lesion can be removed, but recurrence is common (Motoi 1992). Systemic and intralesional corticosteroids have been shown to reduce the size of the lesion, but the tumour tends to recur when these drugs are discontinued (Motoi 1992). In selected patients, it may be advisable to take a conservative approach, treating only if the mass continues to grow or causes significant deformity. Local irradiation has also been shown to be effective in shrinking the lesions, but it is generally not advocated in younger patients (Motoi 1992).

In summary, KD is an indolent, benign disorder, but may easily be mistaken as a malignant lesion. With strict histologic criteria, a correct diagnosis can be achieved, especially when combined with pertinent clinical information and laboratory studies.

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