CASE OF THE MONTH

Head and neck lumps in an Asian male

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Case report

A 60-year-old Chinese male presented with a 1 year history of bilateral painless swelling over the cheeks and right upper neck. On physical examination, firm, mobile and non-tender subcutaneous masses were noted at these sites. Bilateral parotid tumours were suspected and a CT scan was performed for evaluation. A follow-up MRI was performed 1 year later because one of the masses at the lower lip was reported to have increased in size. The diagnosis was confirmed on histopathology and a characteristic blood picture. What is the most likely diagnosis? What is the characteristic blood picture in this condition?

Figure 1. Contrast-enhanced axial CT image of the neck.

Figure 2. Axial T_2 weighted MR image with fat saturation.

Figure 3. Gadolinium-enhanced coronal T_1 weighted MR image with fat saturation.

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Findings

The axial (Figure 1) contrast-enhanced CT image reveals several mildly enhancing, ill-defined plaque-like soft-tissue masses in the superficial lobes of the parotid glands along the outer table of the right mandibular angle and at the left lower lip. Small volume/enlarged intraparotid and upper cervical lymph nodes are also seen.

These masses are mildly hyperintense on the axial fat saturated T_2 weighted MRI (Figure 2). Moderate enhancement is seen in these masses on the coronal post-contrast fat saturated T_1 weighted MRI (Figure 3).

Diagnosis

Kimura's disease was confirmed on fine needle aspiration and cytology (FNAC). Peripheral blood eosinophilia is a typical feature of this condition.

Discussion

Kimura's disease is a rare chronic inflammatory disorder of unknown aetiology, prevalent in young Asian males. As these patients present with painless subcutaneous masses in the head and neck region, more sinister conditions like lymphoma, salivary gland tumours and tuberculous adenitis are often clinically suspected. A strong male preponderance has been noted, with most cases presenting in the third and fourth decades of life. Typically, peripheral blood eosinophilia and increased serum immunoglobulin E concentrations are accompanying features. Hence, an aberrant immune reaction to an unknown antigen has been postulated [1–3]. It largely occurs in the head and neck region; however, rarely the axilla, popliteal region, groin and forearm may be involved [1]. Concurrent nephropathy including nephritic syndrome has been described in some case reports [4]. Histopathological findings include prominent germinal centres with fibrous, vascular and cellular proliferation; and a cellular component consisting of dense eosinophilic infiltrates on a background of abundant lymphocytes, plasma cells, eosinophilic micro-abscesses with central necrosis and Warthin-Finkeldey-type polykaryocytes [2].

Soft-tissue diseases of the head and neck are routinely evaluated with CT at our institution while MRI is usually reserved as a problem-solving tool. However, MRI is widely accepted as the modality of choice in this regard. In Europe and some parts of Asia, ultrasound is used as a first line modality for evaluating neck masses. On ultrasound, the masses are mostly solid and hypo-echoic, with

homogeneous or heterogeneous internal architecture and high internal vascularity. The affected lymph nodes are often round, well-defined, and hypo-echoic with a preserved echogenic hilum. The masses are usually iso- or hypodense on CT scan, iso- or hypo-intense on T_1 weighted MR images and hyperintense on T_2 weighted MR images [5]. Variability in the signal intensities and in the degrees of enhancement on contrast-enhanced images has been attributed to varying degrees of fibrosis and vascular proliferation. Recently, in a retrospective study of 13 cases of Kimura's disease, Gopinathan et al [3] classified the lesions into two types; Type I lesions are well-defined nodular masses with homogeneous enhancement, frequent in a younger age group, and Type II lesions are ill-defined, plaque-like in configuration and heterogeneous in appearance with variable enhancement. These are prevalent in an older age group [3]. Well-defined rounded enhancing nodes with high vascularity have been described. [1]. The characteristic blood picture is supportive of Kimura's disease. FNAC may be diagnostic, but may be inconclusive and an excision biopsy may be necessitated to address the concerns of sinister conditions like parotid tumours, lymphoma, tuberculosis and angiolymphoid hyperplasia with eosinophilia, which are the clinical and radiological differentials of this condition.

The treatment options for Kimura's disease include steroids, radiotherapy and surgical excision. Steroids provide transient relief and the disease often recurs on cessation. Local irradiation is effective in a majority of cases. Excision is often associated with recurrence.

The imaging findings of subcutaneous masses in the head and neck region with or without infiltration of the major salivary glands and regional adenopathy in an Asian male should raise the suspicion of Kimura's disease.

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