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Radiological Features In A Patient With Kimura's Disease

W E H Lim, N G Tan, K P Tan

ABSTRACT

Kimura's disease is an immune mediated inflammatory disorder that usually involves the head and neck region, primarily affecting the salivary glands, adjacent muscle and regional lymph nodes. Peripheral blood eosinophilia is the norm. Clinically and radiologically, it is difficult to differentiate Kimura's disease from salivary gland malignancy, lymphoma or haemangioma. The radiological findings of a patient who presented with a left facial mass involving the left parotid gland and the adjacent muscles are discussed. We also propose that the difference in the degree of enhancement between the initial and subsequent computed tomography study done two and a half years later may be due to the development of increased fibrosis and sclerosis as the disease progresses.

Keywords: computed tomography, magnetic resonance imaging, angiography, cervical lymphadenopathy, parotid gland disease

INTRODUCTION

Kimura's disease is a rare, chronic inflammatory disorder that is almost exclusive to Orientals, with rare cases having been reported in Caucasians[1-4]. The first report of this entity was in the Chinese literature in 1937, by Kim and Szeto, who documented 7 cases which they designated histologically as 'eosinophilic hyperplastic lymphogranuloma'. Kimura is however, credited with first describing the disease in 1948. Most of the literature on this unusual entity is found in journals of histopathology, with only limited published reports of its radiological features[5-7]. We present a case in a 50-year-old Chinese man who was initially diagnosed clinically to have a venous haemangioma in the left parotid region, and discuss the clinical presentation and radiological findings on angiography, computed tomography (CT) and magnetic resonance imaging (MRI).

CASE REPORT

A 50-year-old Chinese man presented with a 6-month history of a painless and non-tender mass in the left malar region, anterior to the ear which was noted to be enlarged and subsided spontaneously. The patient was otherwise well and had no constitutional symptoms. Physical examination revealed the presence of a soft, 6 cm diameter mass in the left temporal and malar region. No other significant clinical findings were noted. CT scan showed the presence of a densely enhancing, ill defined mass in the subcutaneous tissue lateral to the left parotid gland, which extended into the superficial lobe of the parotid gland and the masseter and temporalis muscles(Fig 1). The attenuation values prior to contrast enhancement averaged 70 to 80 Hounsfield units (HU). After contrast this increased to 130 HU. No enlarged regional lymph nodes were reported. The scan findings were interpreted as being consistent with a haemangioma. A carotid angiogram (Fig 2) was performed and this showed that the lesion exhibited mild vascularity with contrast staining noted in the late phase of the angiogram. The patient was provisionally diagnosed to have a venous haemangioma with a differential diagnosis of a soft tissue tumour. Blood investigations at that time were essentially unremarkable, although there was no record of a differential eosinophil count. The patient defaulted follow-up, only to return two and a half years later as the mass had increased in size. This was confirmed on clinical examination, which also revealed the presence of enlarged nodes at the angle of the jaw. Repeat CT scan examination showed that the mass had enlarged (Fig 3), with further involvement of the left parotid gland and also the left submandibular gland. Enlarged lymph nodes were seen in the region of the parotid gland and the left carotid sheath. There was extension of the mass into the left masseter and temporalis muscles as reported

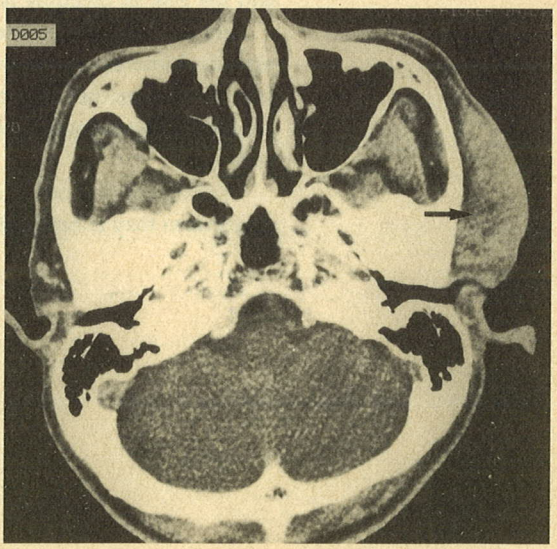

Fig 1 - Initial contrast enhanced axial computed tomography scan (CT) demonstrating the dense contrast enhancement (130 HU), and the involvement of the temporalis muscle by the mass (arrow).

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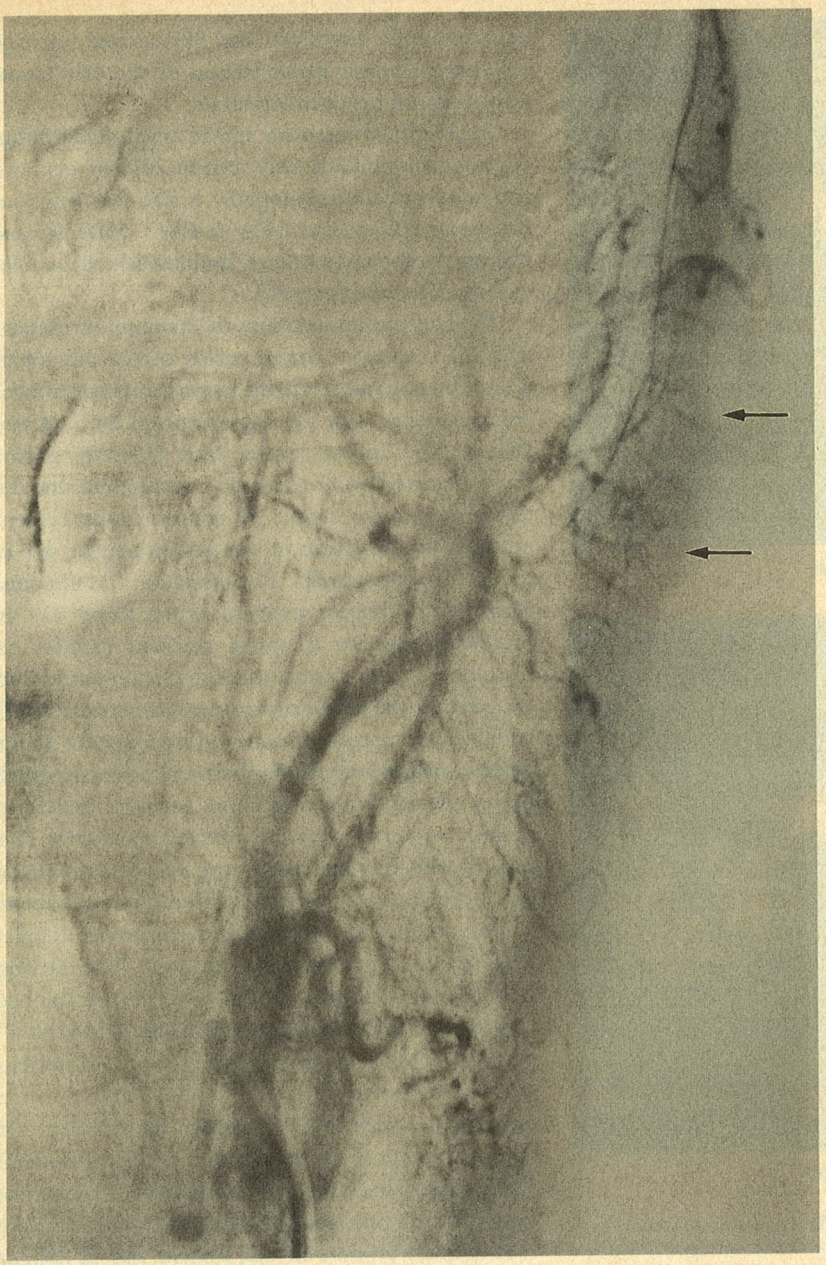

Fig 2 - Selective external carotid angiogram showing the mildly vascular nature (arrows) of the left parotid and malar mass.

previously. This time, the enhancement with intravenous contrast was minimal, in contradistinction to the previous CT scan. The contrast dose administered and radiographic factors were similar in both examinations. The attenuation values averaged between 70 to 80 HU on the unenhanced study and 80 to 90 HU after contrast administration. The regional lymph nodes demonstrated mild, uniform enhancement. The MRI scan done at 1.0T (Fig 4) showed that the mass and the intraparotid nodes were mildly hyperintense to the parotid gland on T2 weighted (T2W) scan. On the proton density (PD) sequence, the mass was hypointense relative to the parotid gland, while the nodes remained hyperintense. On the T1 weighted (T1W) studies, both the mass and the nodes were hypointense to the parotid gland. Strong contrast enhancement was exhibited by the mass on administration of Gadolinium DTPA (GdDTPA), while the lymph nodes showed mild enhancement. A diagnosis of a slow growing soft tissue tumour of the salivary gland with nodal extension was made based on the clinical and imaging findings.

The patient underwent a fine needle aspiration, which demonstrated the presence of lymphoid cells, possibly due to reactive hyperplasia. An excision biopsy was then performed. The patient's peripheral blood eosinophil count was 30% during this admission. The pathology of the parotid mass and the cervical nodes showed features of Kimura's disease. There were aggregates of lymphoid cells and eosinophils in the subcutis, with inflammatory infiltrates noted around thin walled blood vessels which had prominent endothelial cells. Thin slivers of fibrous tissue with lymphoid and eosinophilic infiltrates centred around the blood vessels were seen in the specimen of tumour resected from the left parotid gland.

DISCUSSION

Kimura's disease is extremely uncommon. The aetiology is unknown, although it has been proposed that it represents an immune inflammatory response to a yet undetected stimulus[2,3]. Its radiological features have been reported previously in English radiological literature as case reports[5,6] or in small series of cases[7]. This may be because it primarily occurs in patients of Oriental extraction. The disease affects predominantly males in the second to third decade of life, with a greater male to female ratio of 3:1.[2,3]. The clinical onset of disease is insidious, and a prolonged clinical course is usual. Most patients present with painless discrete nodules or a localised swelling in the head and neck region. The subcutaneous and deep soft tissue, major salivary glands and regional lymph nodes are the usual sites of involvement[2,3]. Involvement of the axilla, groin and other sites of the body have also been reported[3]. Peripheral blood eosinophilia is invariably present. Although a benign condition, the clinical course is often progressive and recurrence after surgery or local

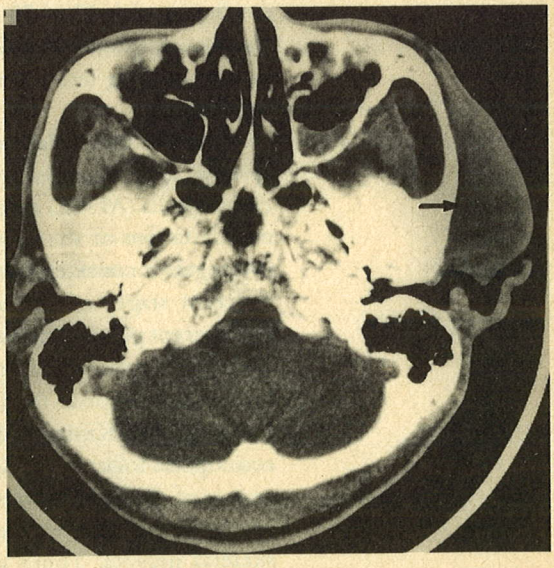

Fig 3 - Contrast enhanced axial CT images obtained 2½ years after the initial CT study. The mass (arrow) had enlarged, with only mild contrast enhancement seen (80 to 90 HU).

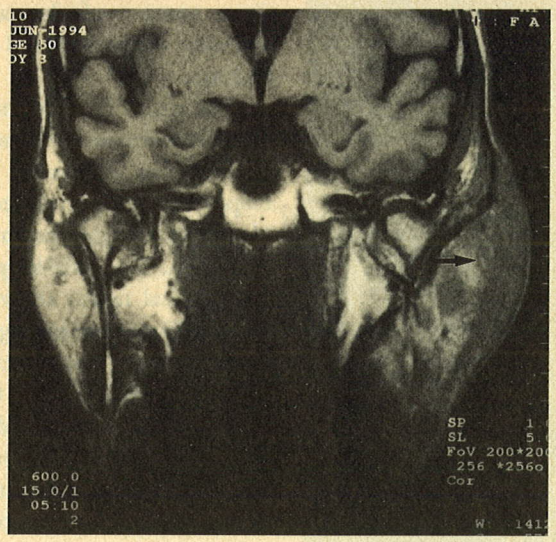

Fig 4a

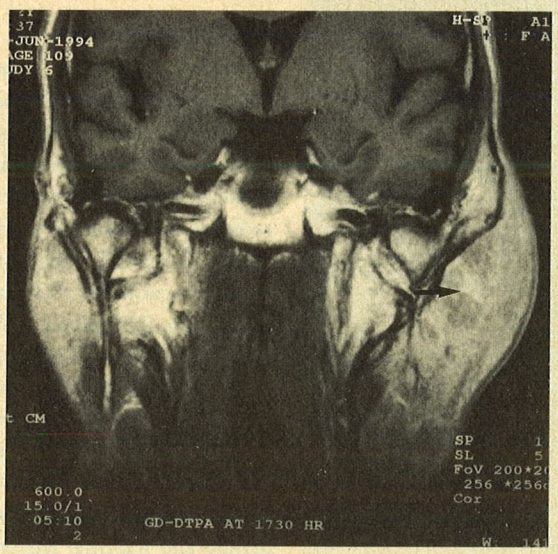

Fig 4b

Fig 4a - (unenhanced) and **4b** - (GdDTPA enhanced) coronal T1W MRI images. The mass (arrow) and the enlarged nodes are hypointense to the parotid gland precontrast. Marked enhancement is noted in the mass, post contrast administration.

irradiation with 25 to 30 Gy[8] is common. There is also usually good initial response to steroid therapy, but the swelling tends to recur with the cessation of treatment[8].

There is much debate as to whether Kimura's disease and Angiolymphoid Hyperplasia with Eosinophilia (ALHE) are distinct entities or different manifestations of the same disease[4,9]. Most Western investigators consider them to be the same disease or different stages of the one disease[3,4,9]. However, more recent papers originating from Hong Kong and Taiwan[1-3], present clinical and histopathological points that appear to distinguish both entities. Histologically, Kimura's disease is characterised by eosinophilic folliculolysis, IgE deposits in the germinal centres of the lymph nodes and frequent involvement of the salivary glands, regional lymph nodes and skeletal muscle. There are also florid lymphoid infiltrates with prominent lymphoid follicles, vascularization of germinal centres and proliferation of high endothelial venules. There may or may not be eosinophilic abscesses, and fibrosis and sclerosis are also a feature. Vacuolization of the endothelial cells does not occur in Kimura's.

In ALHE, there is no sex or racial predilection, blood eosinophilia is only seen in 20% of patients, and cervical lymphadenopathy is also less common (less than 20% versus 50% to 66%). Sclerosis and fibrosis are also not a feature. Endothelial cell vacuoles can also often be found[1-3].

We had the opportunity to examine our patient with CT scan twice over a period of two and a half years. On the earlier scan, the lesion exhibited stronger enhancement, as in the case reported by Som and Biller[5]. The findings on the later CT study mirror those reported by Ahuju et al[7]. We propose that the difference in the degree of contrast enhancement between the two scans in our patient may be due to the fact that the initial CT examination was obtained early in the disease process, when there was an increased number of post capillary venules on histology[2,3]. The vascularity of the lesion would thus account for the dense contrast enhancement on CT, and the mild vascularity noted on angiography. As the disease progressed, the fibrosis and sclerosis which began around the postcapillary venules, resulted in gradual obliteration of the affected venules[2,3]. This may explain the decrease in the degree of contrast enhancement on the second CT scan in our patient; and the absence of significant flow on colour doppler imaging, as reported by Ahuja[7]. The strong enhancement noted on the GdDTPA enhanced studies probably reflect the sensitivity of MRI to T1 shortening. This is because there was also marked contrast enhancement of the lesions in the 2 earlier case reports with MRI[5,7], despite the different CT scan observations in their patients. The ultrasound findings have been reported by Ahuja in 2 cases. The masses being described as being hypoechoic; with well-defined borders in one patient and ill defined borders in the other. The regional lymph nodes were also hypoechoic, solid and had preserved hila[7].

The findings on both CT and MRI in our patient are non-specific. A salivary gland tumour with lymph node extension or a lymphoma of the salivary glands with nodal involvement are just two of the differential diagnoses. Imaging however, can exclude acute inflammatory conditions, confirm the presence of a discrete mass and guide biopsy procedures.

CONCLUSION

Kimura's disease is rare in comparison to the more common salivary gland tumours and lymphoma. The varied and non-specific imaging findings make diagnosis based on imaging alone difficult. However, it may be suggested as a diagnosis, in an Oriental male presenting with a salivary gland mass that is associated with both cervical lymphadenopathy and blood eosinophilia.

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