

Kimura's Disease of the Parotid: A Complete Clinical-Radiological-Pathology Report

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SUMMARY

Kimura's disease is rare chronic inflammatory disease with a distinct clinicopathological entity. It has three major components; inflammatory, vascular and fibrosis. It has to be considered as a differential diagnosis in young patient presenting with head and neck swelling. Although of unknown aetiology many hypothesis has been postulated. Inflammation is the most prominent and predominating characteristic in this disease. Although reported to be predominant in Asian literature regarding this disease is scanty. We report a complete clinical-radiological and pathological picture of this disease.

KEY WORDS:

Kimura's Disease, Parotid Gland Inflammatory Diseases, Chronic eosinophilic inflammation

INTRODUCTION

Kimura's disease (KD) was firstly described in a Chinese literature in 1937,¹ and its histological description was produced by Kimura et al in 1948 which has given its name². It is a disease of chronic eosinophilic inflammation, probably immune-driven, with angioplasty proliferation. The aetiology is unknown. Typically, the disease occurs as a tumour-like nodule in subcutaneous tissue of head and neck, antecubital fossa, inguinal region or parotid gland.³ It often accompanied by regional lymphadenopathy, raised serum eosinophil counts, and markedly elevated serum immunoglobulin E levels³.

Diagnosis of KD is based on histological characteristics, i.e. preserved nodal architecture with reactive germinal centres, eosinophil infiltration and proliferation of post-capillary venules.⁴ Malignant transformation of Kimura's disease has not been reported so far. Management of KD remained controversial with no definite guideline. Surgical excision is frequently performed. Other options include local radiation, systemic corticosteroids, cyclophosphamide, and pentoxifylline.

Kimura disease has a predilection for young Asian males. However, in Malaysia, report on its clinical manifestations, laboratory findings, therapeutic response and disease outcome are scanty. We report a case of KD in parotid gland with complete description of its clinical, pathological and radiological findings.

CASE REPORT

31-year-old Malay gentleman presented with 6 years history of painless right parotid mass. It was slow-growing but had become more prominent in previous 1 year. He denied any symptoms of fever, lethargy, weight loss or night sweats. There was no allergy and other systemic presentation.

Physical examination showed 8 x 4 cm right parotid mass, which is soft in consistency, not fluctuant, fixed and non-tender on palpation. There was no sign of local inflammation. No facial nerve involvement was noted. There was no cervical lymph node enlargement.

Reviewing his laboratory investigation, haemoglobin, total white cell count, and renal profile were within normal limits. However, the differential white cell count showed 10.2% eosinophilia. Immunoglobulin E (IgE) was not measured.

On ultrasonography, right superficial parotid gland was enlarged and appears heterogeneous. It measured 8.2cm (length) x 1.8cm (AP) and showed increased vascularity (Figure 1-A, 1-B, 1-C). Whereas, the right deep parotid lobe was thickened with homogeneous echogenicity. Multiple enlarged intra-parotid nodes with fatty hilum were detected (Figure 1-D). There were also multiple sub-centimetre cervical nodes. These were initially concluded as features of right parotid tumour, likely Warthin's tumour.

On the other hand, CT scan of neck showed a heterogeneous enhancing lesion at right parotid space, which consisted of solid and no fluid component. It measures 4.7 cm (AP) x 2.1 cm (W) x 7.9cm (Depth). There is no clear plane between the lesion and right deep parotid gland. No calcification noted. No surrounding fat streakiness. Right deep parotid lobe appears preserved. (Figure 1-E, 1-F) The result was not conclusive which correlation with histopathology report is mandatory.

We proceeded with fine needle aspiration cytology (FNAC) from the parotid swelling, which were inconclusive on two attempts. In view of progressively-increasing size of the mass and features concluded on ultrasonography as Warthin's tumour, the patient was treated by superficial parotidectomy.

The surgical specimen received as solid, tan brownish cut surface with surrounding fibrofatty tissue and salivary gland tissue, measuring 75x45x30mm. No area of haemorrhage, necrosis or any cystic space seen.

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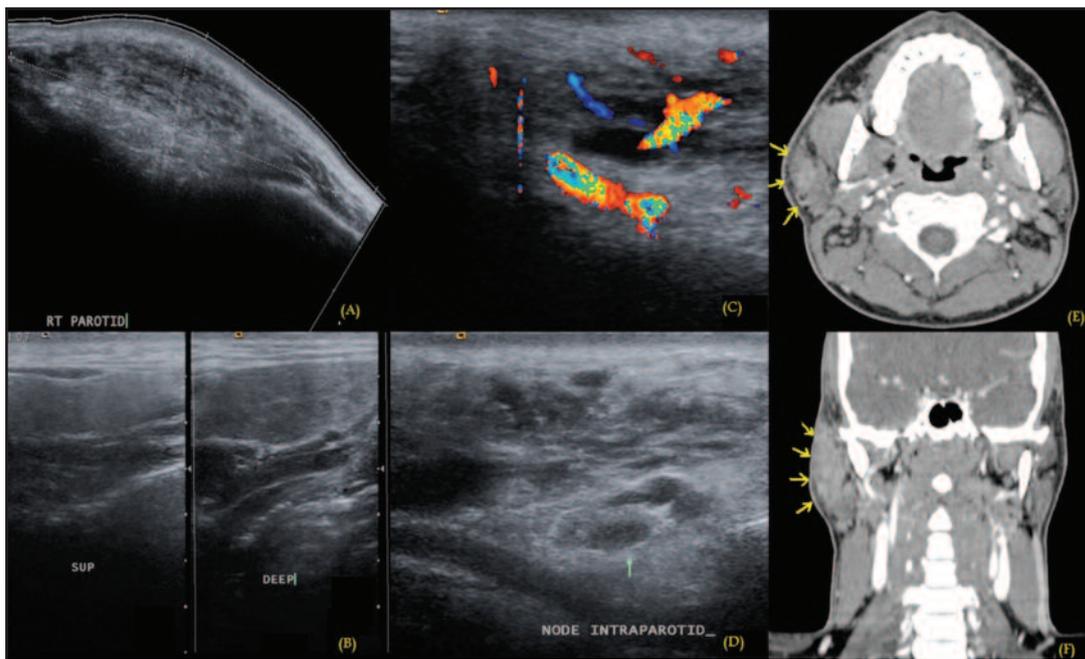


Fig. 1: Radiology

- (A, B) – US: Right parotid gland. Superficial lobe enlarged and appears heterogeneous. Deep lobe was thickened with homogeneous echogenicity.
- (C) – US: Right superficial parotid gland showed increased vascularity.
- (D) – US: Multiple enlarged intra-parotid nodes with fatty hila.
- (E, F) – CT: Heterogenous enhancing lesion at right parotid space, consisted of solid component. There is no clear plane between lesion and deep lobe.

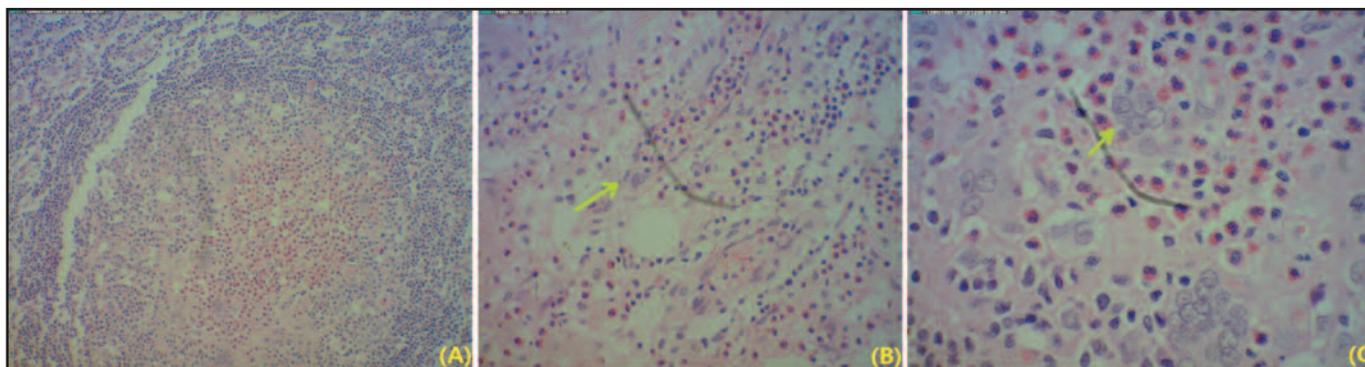


Fig. 1: Histopathology

- (A) Lymphoid follicles with prominent germinal centre and eosinophilic microabscess (H&E, x 10)
- (B) Eosinophilic infiltrates with proliferating capillaries (arrow) (H&E, x 20)
- (C) Prominent eosinophilic infiltrates with polykaryocytes (arrow) (H&E, x 40)

Microscopically, there was an intact parotid gland adjacent to the subcutaneous fat. The subcutaneous fat was replaced by numerous lymphoid follicles of various sizes (Figure 2-B), exhibiting prominent germinal centres frequently infiltrated by eosinophil's micro abscess (Figure 2-A). The lymphoid tissues are well vascularised and contain few polykaryocytes within the germinal centres (Figure 2-C). Surrounding oedema with lymphocytes, plasma cells and eosinophil infiltrate are seen. The features are consistent with Kimura's disease. The gross and microscopic findings confirmed that this is not a Warthin's tumour.

Post-operatively, recovery was good with no facial nerve palsy. A follow up of 1 year showed no recurrence. Renal profile remained normal.

DISCUSSION

Kimura disease (KD) is a chronic inflammatory lymphoproliferative disorder of unknown etiology and unsure pathogenesis. Many hypotheses were postulated, including autoimmune, allergic and infective factors. One of the most developed theory believed to be unknown stimulus

persistently activating the immune system, leading to clonal proliferation of Helper T cells, which may then lead to high expression of IL-4, IL-5, IL-13, potent stimulators of eosinophilic proliferation⁵. *Candida albicans*, human Herpes virus-8 and Epstein-Barr virus have been suggested as causative agents without any conclusive evidence. Up-to-date, no infective agent has been isolated from lesion of KD⁵. As in our patient, peripheral eosinophils are 10.2% of total white cells. No specific stimulus was identified from the HPE of the excised lesion.

Usually, KD presents as one or multiple soft tissue swellings along with lymphadenopathy in head and neck region with a preference for parotid and submaxillary glands³. Indeed isolated lymphadenopathy might be the only initial presentation of Kimura's disease. Our patient had a right sided parotid swelling with a few sub-centimetre cervical lymphadenopathies detected from ultrasound. Cases also were reported in other location such as axilla, trunk and groin. Usually these lesions are benign. Although it gradually increases in size, malignant transformation has not been reported. The overall prognosis is good.

The challenge will be in the preoperative diagnosis. Neither radiology nor cytology is specific for KD. Most diagnosis was made post operatively by the histopathological features. These lesions are usually well defined, not encapsulated and composed of three major components; the inflammatory, vascular and fibrous. As per our case these lesions are heavily infiltrated by lymphocytes and eosinophil.

KD closely resembles angiolymphoid hyperplasia with eosinophilia (ALHE) and they were earlier thought to be the same. They share similarity, i.e. proliferation of lymphoid follicles and blood vessels, as well as prominent eosinophil in the lesion as well as in blood. However, the site of occurrence is different where KD is commonly found in deep seated subcutaneous tissue or salivary gland tissue at head and neck, while ALHE are usually superficial in location and smaller in size.

The case discussed above was also initially thought to be Warthin's tumour pre-operatively. Generally, both KD and Warthin's tumour occur in parotid gland. Warthin's tumour is a multicystic lesion with separating septa which microscopically consist of prominent lymphoid tissue with germinal centres and eosinophilic microabscess is absent.

Other differential diagnoses of KD include acute lymphocytic leukemia, Hodgkin's disease and eosinophilic granuloma. There is no guideline for the treatment as there is only a few case series reported so far. Various modalities have been used including surgery, radiotherapy, steroids and immunosuppressive therapy with cyclosporine.

In conclusion KD disease of chronic eosinophilic inflammation with angioplasty proliferation. Although it is a rare disease it has to be considered as a differential diagnosis of the head and neck swelling especially involving the major salivary glands. It has a distinct clinicopathologic entity with characteristic histological feature and is important to separate from other entity. diagnosis.

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