



Case Report

Kimura's Disease of the Pharynx - A Case Report of 'Unusual Granulation' At an Unusual Location

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ABSTRACT

Kimura's disease (KD) is a rare chronic inflammatory disorder, manifesting as deep subcutaneous mass lesions of head and neck with regional lymphadenopathy, affecting young men of Asian origin. When present at unusual locations these lesions tend to mimic neoplasia clinically. We report a case of Kimura's disease involving the pharynx, presenting with dysphagia and foreign body impaction. During laryngoscopy a 2x1cm mass was noted in the pharynx suspicious for malignancy. Biopsy of the lesion showed features of Kimura's disease, confirmed subsequently by raised serum IgE levels and high eosinophil count.

Problems faced in the differential diagnosis due to the unusual clinical presentation combined with certain atypical microscopic findings are discussed.

Keywords: Kimura's disease, pharynx, IgE

INTRODUCTION

Kimura's disease (KD) is a rare immune mediated inflammatory disorder, manifesting as deep subcutaneous mass lesions of head and neck with regional lymphadenopathy, affecting young men of Asian origin. The disease is associated with peripheral blood eosinophilia and elevated serum IgE levels. Characterized histologically by lymphoid hyperplasia, vascular proliferation and eosinophilic infiltration, the lesions of Kimura's disease, even in typical anatomic locations, present a diagnostic challenge. Distinguishing Kimura's disease from angiolymphoid hyperplasia with eosinophilia (ALHE) needs awareness and careful analysis of the diagnostic criteria. The problem is

compounded when the location is unusual and presentation, atypical.

We report a case of Kimura's disease of the pharynx illustrating these diagnostic difficulties. While its occurrence at unusual locations like oral cavity, hard palate, epiglottis and orbit [1-4] are on record, involvement of larynx and pharynx appears to be exceedingly rare, [5] and only one case of pharyngeal Kimura's disease has been reported so far in literature, to the best of our knowledge. [6]

CASE REPORT

A 65-year-old woman presented to our hospital in November 2012 with complaints of dysphagia and odynophagia of

four days duration. There was no history of change of voice, nasal obstruction or allergy.

Computed Tomography (CT) showed soft tissue thickening and asymmetry at the supraglottic level with involvement of pyriform sinus, left vallecula and hypopharynx. A linear hyperdense structure was seen in the pyriform sinus / hypopharynx, suggestive of foreign body, with possible inflammation and reactionary edema around.

Laryngoscopy was performed and a tiny piece of coconut shell was removed from the pyriform fossa. At the same time a 2x1 cm swelling was noticed extending from the lateral aspect of aryepiglottic fold into hypopharynx. Suspecting malignancy, a biopsy was taken from the swelling. The sample proved to be inadequate for assessment. A repeat biopsy was therefore done, after a gap of twenty days.

Laboratory investigations: complete blood count revealed eosinophilia of 20% (normal- 1 to 6%) and absolute eosinophil count of 2100/ μ L (normal- 20 to 500/ μ L). Renal function tests, diabetic profile and lipid profile recorded no abnormality. Testing for HIV and HBsAg gave negative results.

Histopathological examination: The second biopsy sample from the mass lesion showed pharyngeal mucosa with pronounced vascular proliferation, infiltration by numerous eosinophils and fibrosis. Lymphoid tissue was present focally [Fig.A]. The blood vessels were lined by plump endothelial cells without cytoplasmic vacuoles or hob-nail nuclei [Fig. B]. Eosinophils were found aggregating in foci with some karyorrhexis [Fig.B]. Interestingly, there was an additional component of pale histiocytic cells, infiltrating the interstitium, and forming vague collections with phagocytosed eosinophils and eosinophil granules [Fig.C]. An occasional giant cell

was also present [Fig. D]. These histiocytic cells expressed vimentin and the macrophage-monocyte marker CD68. Endothelial markers [CD31, CD34] were not expressed [Fig E&F]. Based on the above features a differential diagnosis of Kimura's disease/ALHE was considered. Serum IgE estimation was suggested, which turned out to be significantly raised - 448 IU/ml (normal- <100 IU/ml), confirming the diagnosis of Kimura's disease.

Treatment and Prognosis

Hydrocortisone injection was given for two days followed by oral corticosteroids for two weeks, tapered over a period of one month. She was symptomatically well on follow-up after 3 months.

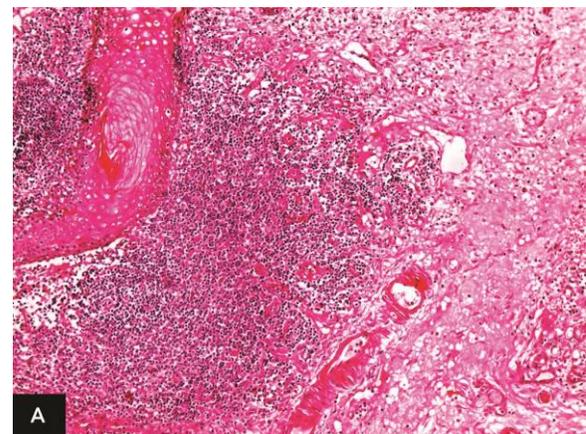


Fig A. Pharyngeal mucosa with lymphoid aggregates (hematoxylin-eosin, magnificationX100).

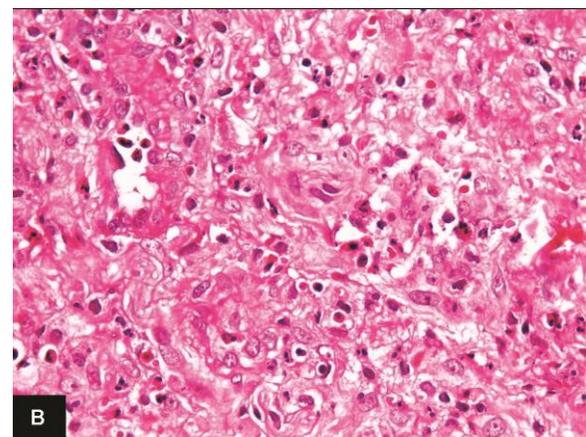


Fig B. Proliferating blood vessels are lined by plump endothelial cells. Numerous eosinophils and karyorrhectic debris are present in the interstitium (hematoxylin-eosin, magnificationX400).

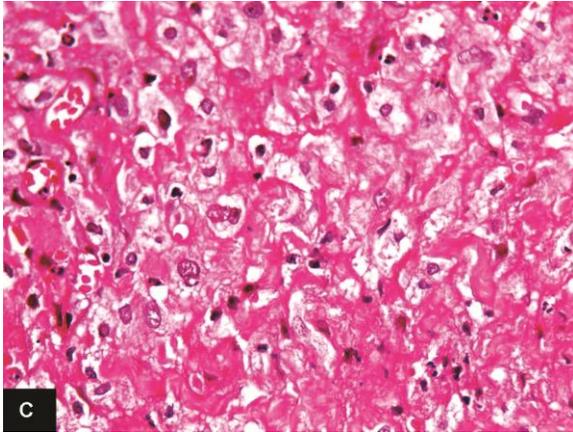


Fig C. Histiocytic cells with abundant foamy cytoplasm were conspicuous in parts of the sections (hematoxylin-eosin, magnificationX400).

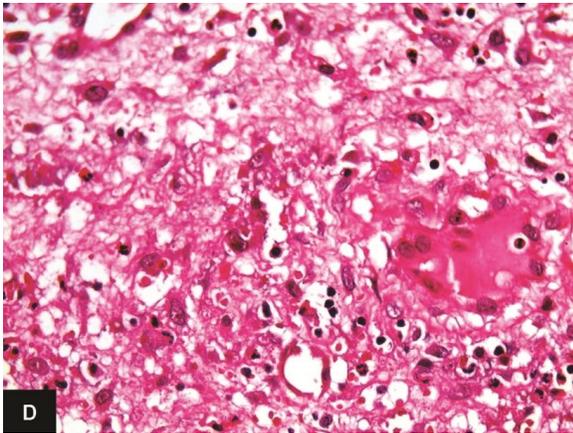


Fig D. Eosinophil phagocytosis by histiocytes and a giant cell (hematoxylin-eosin, magnificationX400).

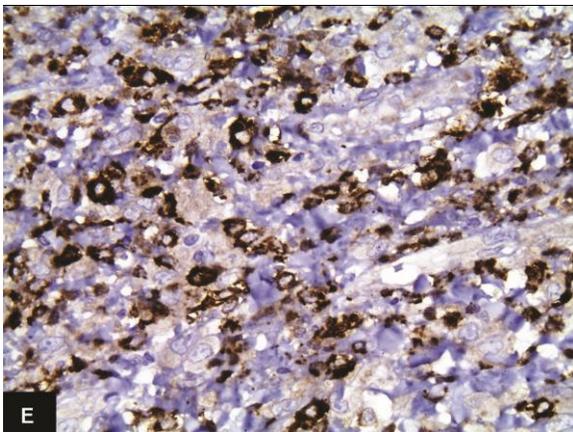


Fig E. Immunostain - The large cells are CD 68 positive confirming macrophage – monocyte lineage (magnificationX400).

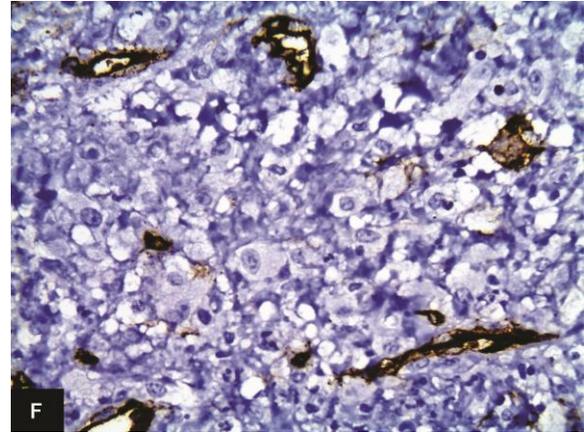


Fig F. Immunostain - CD34 highlights vascular endothelium. Large histiocytes are CD34 negative (magnificationX400).

DISCUSSION

Kimura et al, in the year 1948 described cases of “unusual granulation combined with hyperplastic changes in the lymphoid tissue”.^[7] The disease named after Kimura, presents as deep subcutaneous masses in the head and neck region, with associated lymphadenopathy, elevated serum IgE levels and peripheral blood eosinophilia.^[8] Kimura’s disease is endemic in parts of Asia but has been reported in non-Asian population as well.^[8] Histopathology of this chronic inflammatory disorder is a distinctive combination of hyperplastic lymphoid follicles, eosinophilic infiltrates, vascular proliferation and sclerosis. Other salient microscopic features are eosinophils within follicular centres, eosinophil abscesses, perivenular sclerosis and eosinophil granulomas. Presence of large vessels and epithelioid / histiocytoid endothelial cells are not features of Kimura’s disease.^[8]

Considerable confusion existed between angiolymphoid hyperplasia with eosinophilia (ALHE) and Kimura’s disease [KD], until the report by Rosai et al in 1979 separated the two conditions.^[9] ALHE is now considered an endothelial neoplasm, manifesting in the head and neck region as superficial mass lesions without lymph-

adenopathy, elevated serum IgE levels or peripheral blood eosinophilia.

Histologically, ALHE lacks the hyperplastic lymphoid follicles and eosinophilic abscesses of KD. Blood vessels tend to form lobules and are lined by “epithelioid” or “histiocytoid” endothelial cells, often with hob-nail nuclei. [8] Endothelium in KD, on the other hand, has been described as swollen, but not vacuolated or histiocytoid in appearance.

In the present case, biopsy of the mass showed proliferating small vessels lined by plump endothelium, numerous eosinophils and stromal sclerosis. Lymphoid aggregates, though present, were not overtly hyperplastic as one would expect in KD. A definite distinction between ALHE and KD could not be made at this point. It is to be noted that even in the first and perhaps only reported case of pharyngeal KD, Chong et al did not stress on the finding of lymphoid hyperplasia. Moreover, only a small biopsy sample was available to us for assessment which may not have been fully representative. Subsequent detection of elevated serum IgE levels along with peripheral blood eosinophilia favored the diagnosis of KD over ALHE. Review of literature revealed both KD and ALHE to be rare in this location. [10]

Apart from the absence of obvious lymphoid hyperplasia there were two other unusual features posing diagnostic challenge in the present case. The patient at the time of presentation had a foreign body in the pharynx and the mass was seen adjacent to it during laryngoscopy. The question we had to ask ourselves was whether the mass was the cause or result of foreign body impaction. Histologically, the lesion was not a foreign body granuloma, but ‘unusual granulation’ with eosinophils consistent with KD. The persistence of the mass at the time of second biopsy would also suggest that foreign body was not the cause, but got

impacted because of an already existing pathology. Even if we cannot trace the sequence of events with certainty, it may still be argued that trauma [in this case due to foreign body] is thought to play a role in the etiology of Kimura’s disease, thus ruling out the possibility of a nonspecific foreign body reaction.

The third unusual feature that intrigued us was the presence of large foamy cells in the mucosal interstitium. These cells formed ill-defined collections along with an occasional multinucleated giant cell and showed evidence of phagocytosis of eosinophils and eosinophil granules. They expressed CD68 and vimentin, consistent with monocyte-macrophage lineage. Review of literature showed that such unusual granulomatous reaction to apoptotic eosinophils has been uncommonly observed in Kimura’s disease. [11]

Subsequent finding of peripheral blood eosinophilia and raised serum IgE levels led to the diagnosis of KD in the present case. This was further confirmed by response to therapy.

Clinical presentation of Kimura’s disease may raise the suspicion of malignancy. Accurate diagnosis is important as the condition has a benign though protracted clinical course, with possible recurrences and variable response to different modes of therapy.

The present case of Kimura’s Disease of the pharynx is reported for its exceedingly rare location, interesting clinical presentation and unusual histological features, all of which combined to form a diagnostic challenge.

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