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Case report

An unexpected asymptomatic epiglottal site of Kimura disease

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ABSTRACT

Introduction: Kimura disease (KD) is a chronic lymphoproliferative disorder of unknown etiology that affects the skin and lymph nodes, mostly observed in males of Asian descent. The natural history of asymptomatic epiglottal KD remains unknown. This rare site of KD is often only diagnosed when tumor growth starts to obstruct the upper airways.
Observation: A 34-year-old North African male presented with fatigue and multiple, slowly progressive, fluctuating skin nodules in the right mandibular and retroauricular regions. Computed tomography of the head and neck revealed a large soft tissue tumor close to the right mandibular body and unexpected thickening of the epiglottis. Transnasal laryngoscopy confirmed the CT findings and showed thickening of the epiglottis. The diagnosis of KD was based on histological examination of biopsy specimens taken from the right mandibular tumor, a cervical lymph node, and the epiglottis.
Discussion: Most cases of KD with epiglottal involvement present with dysphonia and dysphagia. No consensus guidelines are available concerning the complementary investigations that should be performed. This case report raises the question of whether patients with suspected KD should be systematically screened for lesions in unusual and potentially dangerous anatomic sites.

1. Introduction

Kimura disease (KD), first described by Kimura in 1948, is a chronic lymphoproliferative disorder of unknown etiology that affects the skin and lymph nodes. The usual clinical features are skin nodules in the head and neck and regional lymphadenopathy. Laboratory findings often include peripheral blood eosinophilia and elevated serum immunoglobulin (Ig)E levels, which are probably due to intense secretion of interleukins-4 and -5 by activated CD4+ (Th2-type) T lymphocytes. Pathological examination always shows deep dermis lymphoid infiltrate with germinal centers, proliferation of small capillary venules, and fibrosis [1]. Kimura disease mostly affects males of Asian descent between the ages of 20 and 30 years. Renal involvement is observed in 30 to 40% of cases, and is associated with poor prognosis [2]. We report a case of slowly progressing, multifocal KD with asymptomatic involvement of the epiglottis.

2. Case report

A 34-year-old North African male presented with fatigue and multiple, slowly progressive, fluctuating skin nodules in the right mandibular and retroauricular regions that had first appeared in adolescence. Episodes of nocturnal fever were concomitant with swelling of the skin nodules.

Histological examination of a skin nodule specimen was compatible with KD. The patient was initially treated with oral corticosteroids, but this treatment had to be discontinued because of limited efficacy and corticosteroid-induced mood disorders.

After having been lost to follow-up for several years, the patient consulted again for recurring inflammation of the skin nodules in the right mandibular region (Fig. 1). A blood test showed fluctuating peripheral blood hypereosinophilia (~1000 eosinophil cells/mm^3) and greatly elevated total serum IgE levels (1514 kU/L; normal range: 0 to 114 kU/L). Immunophenotyping results ruled out a lymphocytic hypereosinophilic syndrome. Immunoglobulin G subclass levels were normal (IgG4 level: 0.285 mg/mL; normal range: 0 to 0.83 mg/mL). Computed tomography (CT) of the head and neck revealed a large soft tissue tumor (greatest dimensions: 48 × 36 × 24 mm) close to the right mandibular body. The

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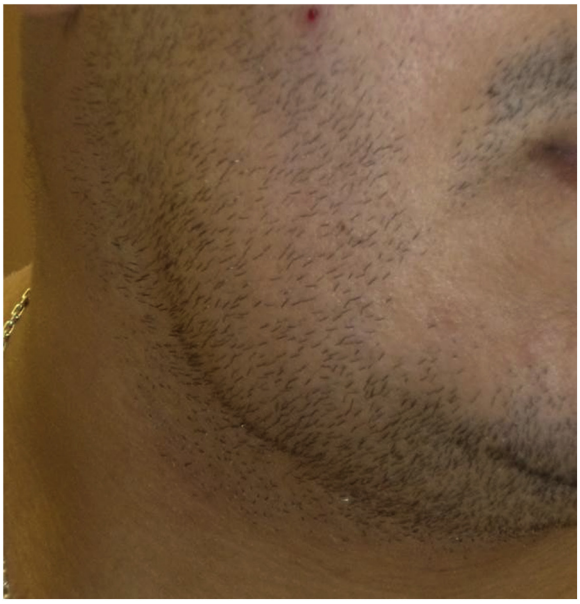

Fig. 1. Slowly progressive, fluctuating skin nodules in the right mandibular region.

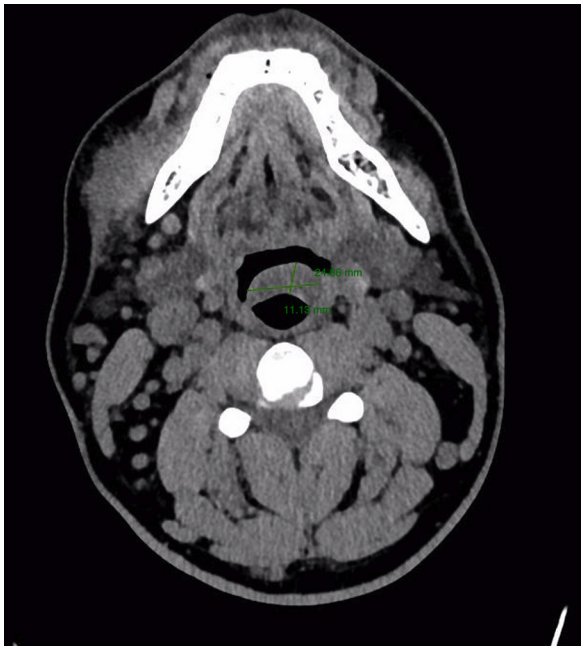

Fig. 2. CT scan of the neck showing a large soft tissue tumor (greatest dimensions: 48 × 36 × 24 mm) in the right mandibular region, together with unexpected thickening of the epiglottis (11 × 25 mm).

tumor had invaded the right masseter muscle, but not the adjacent bones. CT scan also showed unexpected thickening of the epiglottis (11 × 25 mm) and several enlarged cervical lymph nodes (Fig. 2).

Transnasal laryngoscopy confirmed the CT findings and showed thickening of the lingual aspect of the epiglottis (10 × 20 mm) (Fig. 3).

The diagnosis of KD was based on histological examination of biopsy specimens taken from the right mandibular tumor, a cervical lymph node, and the epiglottis. Massive lymphoid infiltration and secondary follicles were observed in the epiglottal sample. The follicles had a hyperplastic germinal center with normal thickness of the mantle zone. Isolated or grouped eosinophilic cells, lymphocytes and plasma cells were observed between the follicles, together with vascular hyperplasia. Eosinophil infiltrate was also

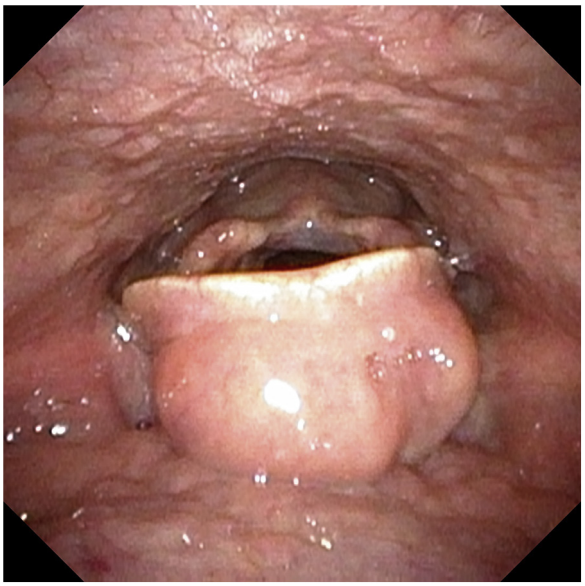

Fig. 3. Transnasal laryngoscopy confirmed thickening of the lingual aspect of the epiglottis.

found in the thickest part of the follicles and in the germinal centers (Fig. 4). These histological findings were consistent with the angiolymphoid hyperplasia described in KD [1,3]. There was no histopathological evidence of IgG4-related disease. The extramembranous glomerulonephritis and proteinuria frequently associated with KD were not present [4]. Given that the patient was not overly troubled by his symptoms, we opted for close surveillance and did not prescribe any treatment.

3. Discussion

Although KD is most commonly observed in Asians, rare cases in non-Asian patients have been reported [5]. In this context, the retrospective study by Kottler et al. of 22 cases found that subcutaneous nodules are the most common feature at presentation [6]. Furthermore, only two patients in Kottler's case series were of North African descent, and three were diagnosed following histological examination of a lymph node biopsy. Kimura disease of the epiglottis is rare, and is often only diagnosed when tumor growth starts to obstruct the upper airways. Consequently, most cases of KD with epiglottal involvement present with dysphonia and dysphagia, and treatment with low-dose oral corticosteroids is usually effective [7]. Of the 11 cases of KD with epiglottal involvement reported in the scientific literature, only two were asymptomatic and were diagnosed after routine CT for cervical lymphadenopathy. Both of these cases were Asian men with elevated peripheral blood eosinophil counts. To the best of our knowledge, the present report is the first to describe early-onset KD (during adolescence) in a patient of North African descent [7]. No consensus guidelines are available concerning the complementary investigations that should be performed in KD, as this disease rare and usually indolent. Mucosal involvement is also uncommon.

In the absence of guidelines, surgical removal of lesions (when feasible) appears to be the best option. Treatment with systemic corticosteroids is also commonly proposed, but raises the issue of long-term adverse events.

In the present case, the epiglottal lesions were not resectable; in the absence of any clinical impact for the patient, we opted for surveillance with no active treatment. If the epiglottal lesion becomes symptomatic or even life-threatening (due to airway obstruction), high-dose intravenous methylprednisolone treatment or local (laser) resection could be considered.

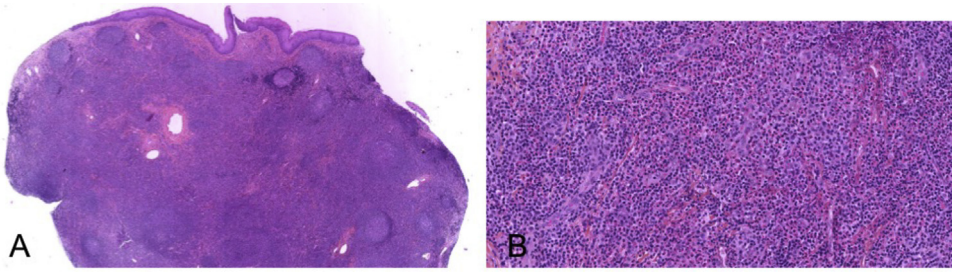

Fig. 4. (A) Histological findings of the epiglottal lesion. Hematoxylin-eosin-stained samples (magnification: × 20) show lymphoid tissue containing secondary follicles (B), eosinophil infiltration and vascular network hyperplasia (magnification: × 200).

The natural history of asymptomatic epiglottal KD remains unknown. The low incidence of KD epiglottal lesions may be due to incorrect or missed diagnoses in asymptomatic cases. The present case of epiglottal KD was diagnosed unexpectedly on CT performed to screen for regional lymph involvement, and could otherwise have gone unnoticed.

4. Conclusion

We report a case of KD with rare, asymptomatic, epiglottal involvement revealed by routine CT scan and confirmed by histological examination. This case raises the question of whether patients with suspected KD should be systematically screened for lesions in unusual and potentially dangerous anatomic sites. However, surgical resection is not always feasible and other treatment options may need to be considered.

Disclosure of interest

The authors declare that they have no competing interest.

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