Kimura’s Disease in the Lower Extremity: A Case Report Mimicking the Malignant Soft Tissue Mass

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We present a case of a 37-year-old woman who had Kimura’s disease involving the lower extremity mimicking malignant soft tissue mass. The diagnosis of Kimura’s disease would be considered if there is a subcutaneous solid mass showing the preservation of the nodal architecture with perinodal infiltrations and the laboratory examinations for peripheral eosinophilia and serum IgE level should be recommended although it occurs at the lower extremity.

Index words: Kimura’s disease
Lower extremity
Images

Introduction

Kimura’s disease has a benign clinical course with a triad of features: painless mass, eosinophilia, and raised serum IgE [1, 2]. In most of the previously reported cases, Kimura’s disease typically presents as single or multiple subcutaneous lesions in the head and neck, especially in the parotid and submandibular regions [3-6]. There are some studies that have demonstrated imaging findings of Kimura’s disease in other less common sites, such as thorax, abdomen and upper extremity [7-9], but to the our knowledge, there has no report about the imaging findings of Kimura’s disease in lower extremity without involvement of head and neck. Clinical findings of Kimura’s disease in an unusual location are occasionally suggestive of malignant soft tissue sarcoma due to its growing nature and associated regional lymphadenopathy without any infectious sign. When considering characteristic imaging findings in the diagnostic algorithm, radiologists may play an important role in alerting the clinician to Kimura’s disease. Here, we report an unusual case of Kimura’s disease with respect to the location.

Case Report

A 37-year-old woman presented with a history of a painless palpable mass in the right medial thigh for a month. There was no history of a precipitating trauma or of any increased or unusual activities. There was no
history of fever, night sweat, or weight loss. On physical examination, she appeared in good health. A non-tender, deeply located mass measuring $5 \times 3$ cm was palpable in the upper medial side of the right thigh. There was no discoloration or heating sensation overlying the skin. There were several lymph nodes measuring 1-2 cm in the right inguinal area. The clinical diagnosis of a soft tissue tumor with regional lymph node metastasis was made and she was examined with MRI. MRI demonstrated a poorly defined subcutaneous mass with eccentric fat and vascular signal void dots (Fig. 1. A-C) Peritumoral soft tissue infiltrations and diffuse enhancement of the mass were noted (Fig. 1. C). Several reactive lymph nodes were located in the right inguinal region just above the mass (Not shown here). The eccentrically located fat signal and signal void dots suggested pathology of a lymph node preserving nodal architecture or fatty mass with an unusual manifestation such as lipoma variants rather than another neoplastic process of soft tissue.

The US showed this mass as a large lymph node with preservation of nodal architecture and perinodal infiltrations, rather than soft tissue sarcoma or any other solid mass type (Fig. 1D). Several reactive lymph nodes in the inguinal area with the same architecture were also noted (Not shown here). As a diagnostic consideration, lymphoproliferative disorder would be highly indicative. Laboratory data revealed a white blood cell count of $11.84 \times 10^3$ /mm$^3$; differential count showed 41% eosinophils (normal range of 1-6%) and extremely high serum immunoglobulin E (IgE) level >68,300 IU/ml out of the normal range of 0-87.0 IU/ml. The associated peripheral eosinophilia and increased serum IgE levels made this lesion strongly suggestive for Kimura's disease. The patient underwent simple excision of the enlarged lymph nodes.

The pathological diagnosis confirmed Kimura's disease. The microscopic findings demonstrated a large lymph node with infiltrated mantle zone and perinodal fatty tissue by the numerous eosinophils (Fig. 1E-G). Several small lymph nodes from the right inguinal nodal station showed a similar manner of eosinophilic infiltrations (Not shown here).

**Discussion**

Kimura's disease is a chronic inflammatory disorder...
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Fig. 1. **d.** Ultrasonography showed an enlarged lymph node with a preservation of the central echogenic fatty hilum, thickened cortex and perinodal infiltrations.

**e-g.** Photomicrographs of specimen. Prominent eosinophilic infiltrates with hyperplastic follicles that were highly vascularized in a lymph node (e, hematoxylin-eosin, × 400). Dense lymphoid and eosinophilic infiltrations in the perinodal soft tissue (SC) crossing the capsule (C) of the lymph node (f, hematoxylin-eosin, × 200). Central fatty hilum with large hilar vessel (arrow) were identified (g, hematoxylin-eosin, × 12). Gross contour of nodal architecture of the largest enlarged lymph node was preserved on gross pathology mapping (Not shown here).

of unknown origin and benign clinical course. Kimura’s disease appear to be endemic in the Asian population and can occur at any age but commonly have occurred during the second and third decades of life. Men are more commonly affected than women, and the male-to-female ratio is greater than 3:1. The onset is insidious and the manifestations include enlarged nodular masses in the deep subcutaneous tissue of the head and neck region, most frequently intraauricular or retroauricular (1, 2). Kimura’s disease in almost all cases involves the regional lymph nodes and histologically, has three components: cellular [inflammatory infiltrate including increased eosinophils and follicular hyperplasia], fibrocollagenous, and vascular [arborizing vascular proliferation of the postcapillary venule, although endothelial cells are usually flat and lack cytologic atypia or vacuolization] (4). Painless slow growing palpable mass may mimic a malignant soft tissue mass.
in a case of unusual location of Kimura’s disease. Many imaging findings of head and neck involvement have been documented (3-6). Several case reports of unusual location, such as thorax, abdomen and upper extremity described CT or MR findings of Kimura’s disease (7-9). The imaging findings of Kimura’s disease in thorax and abdomen with CT showed extensive aggregated lymphadenopathy and in the upper extremity with MRI showed uniform internal signal intensity with iso- to slightly higher than that of skeletal muscle on T1-weighted images and high signal intensity on T2-weighted images, homogeneous enhancement, signal-void structures within the mass and surrounding edema. Despite the unusual location in our case, we discovered similar MRI imaging findings to Kimura’s disease in the upper extremity. We concerned the fat signal intensity umbilicating this mass on T1-weighted images and signal void dots traversing this fatty portion. The US showed this area as an echogenic fatty hilum of an enlarged lymph node with preservation of nodal architecture. Pathologic review according to specimen mapping revealed nodal hilum containing large afferent vessels in an enlarged lymph nodes. Therefore, signal void structure in the central area of the mass represents vascular structure of the nodal hilum. In our opinion, the fat signal intensity and signal void structure umbilicating the mass on T1-weighted images of MRI was very crucial in characterizing this lesion as a pathology in the lymph node. It may be an important MR finding in cases of soft tissue mass to differentiate the lymphatic pathology from sarcomatous lesion. Although a subcutaneous solid mass may have malignant looking appearance on the MRI, it should be carefully interpreted for eccentric fatty tissue containing the arteries on the MRI. In addition, US also has an important role in characterization of this subcutaneous mass. In addition to soft tissue sarcoma other differential diagnoses for Kimura’s disease in the lower extremity should include lymphoma, metastatic lymphadenopathy, infectious lymphadenopathy, such as tuberculous lymphadenitis and pyogenic lymphadenitis, Castleman’s disease and drug-induced lymphadenopathy. First, soft tissue sarcoma should be ruled out and then pathology in lymph nodes could be differentiated considering laboratory findings and biopsy.

In conclusion, Kimura’s disease in the lower extremity is a very rare benign disorder which could be confused with a malignant soft tissue mass. Concerning the characteristic US and MRI findings with preservation of the nodal architecture and perinodal infiltrations in cases of a subcutaneous solid mass, a preoperative diagnosis of Kimura’s disease could be possible in the presence of peripheral eosinophilia and increased serum IgE levels.

References

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