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CASE REPORT

Kimura's Disease Masquerading as Soft Tissue Sarcoma: A Case Report

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INTRODUCTION

Kimura's disease is a rare idiopathic chronic inflammatory disease that predominantly affects young Asian males with the head and neck region most affected.[1,2] Disease manifestation as upper extremity soft tissue masses is extremely rare[3] with only sporadic cases reported. As Kimura's disease is a benign entity, it is important not to misdiagnose these soft tissue masses as sarcomas. We present two pathologically confirmed cases of Kimura's disease affecting the elbows with focus on the ultrasound and magnetic resonance imaging (MRI) features.

CASE REPORTS

Case 1

An 18-year-old man with good past health presented with a 6-month history of bilateral painless soft tissue elbow swellings and increased right neck swelling. He had no constitutional symptoms, fever or itchiness. Physical examination revealed a firm subcutaneous non-tender mass over the medial aspect of each elbow up to 5 × 5 cm in size with no overlying skin changes or neurovascular deficit. Enlarged right neck lymph nodes were also palpable. Laboratory tests revealed elevated eosinophil count of 3.8 × 10^9/L (reference range, 0-0.6 × 10^9/L). Radiograph (Figure 1a) demonstrated soft tissue swelling at the medial aspect of both elbows with no internal calcification or underlying bone change. Ultrasound (Figure 1b and c) showed a poorly defined heterogeneous hypoechoic mass in the subcutaneous layer of the left elbow with curvilinear hyperechoic bands and moderate increased vascularity. MRI (Figure 2) showed ill-defined subcutaneous lesions at the medial epitrochlear region of both elbows with extension to mid humeri level. The lesions had an intermediate T1-weighted (T1W) signal, heterogeneous high signal on T2-weighted (T2W) imaging relative to skeletal muscle with moderate homogeneous enhancement and some intralesional tubular flow voids. A few enlarged discrete lymph nodes were identified and bilateral basilic veins were encased by the lesions although patency was maintained. There was no evidence of tumour necrosis or cystic degeneration. Mild adjacent subcutaneous T2W

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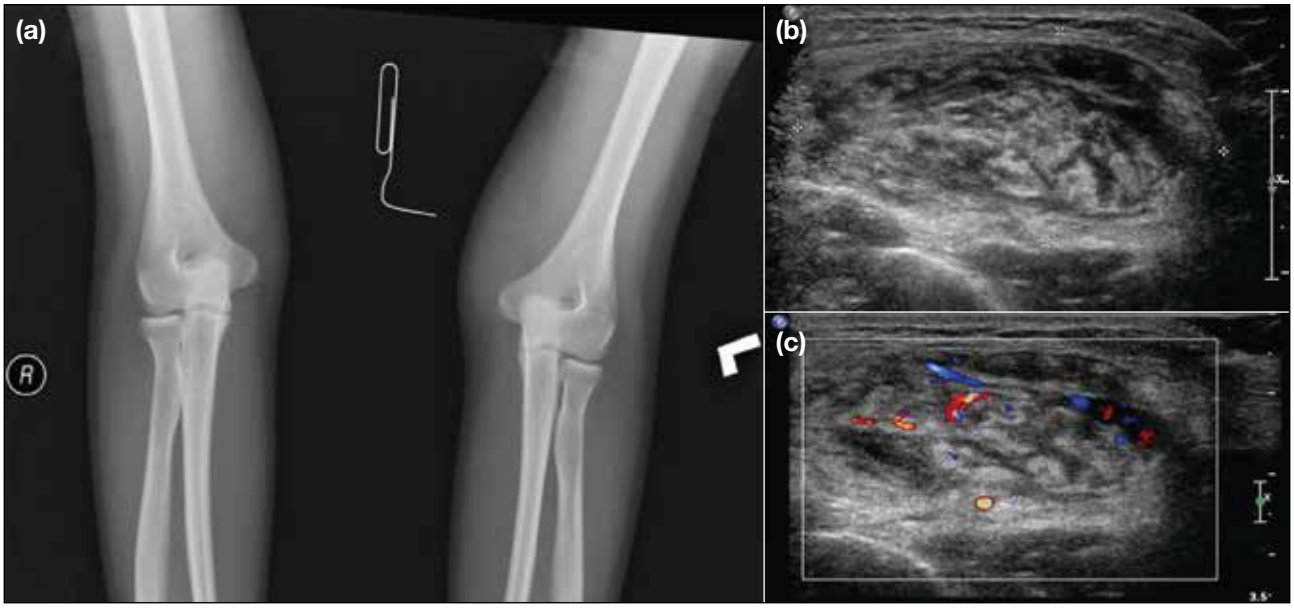

Figure 1. (a) Frontal radiograph of both elbows in case 1 showing subcutaneous soft tissue swellings without calcification or bone changes. (b) Ultrasound of left elbow in case 1 showed poorly defined heterogeneous hypoechoic lesion with internal hyperechoic bands. (c) In the ultrasound image of left elbow in case 1, the mass showed moderate vascularity on colour Doppler.

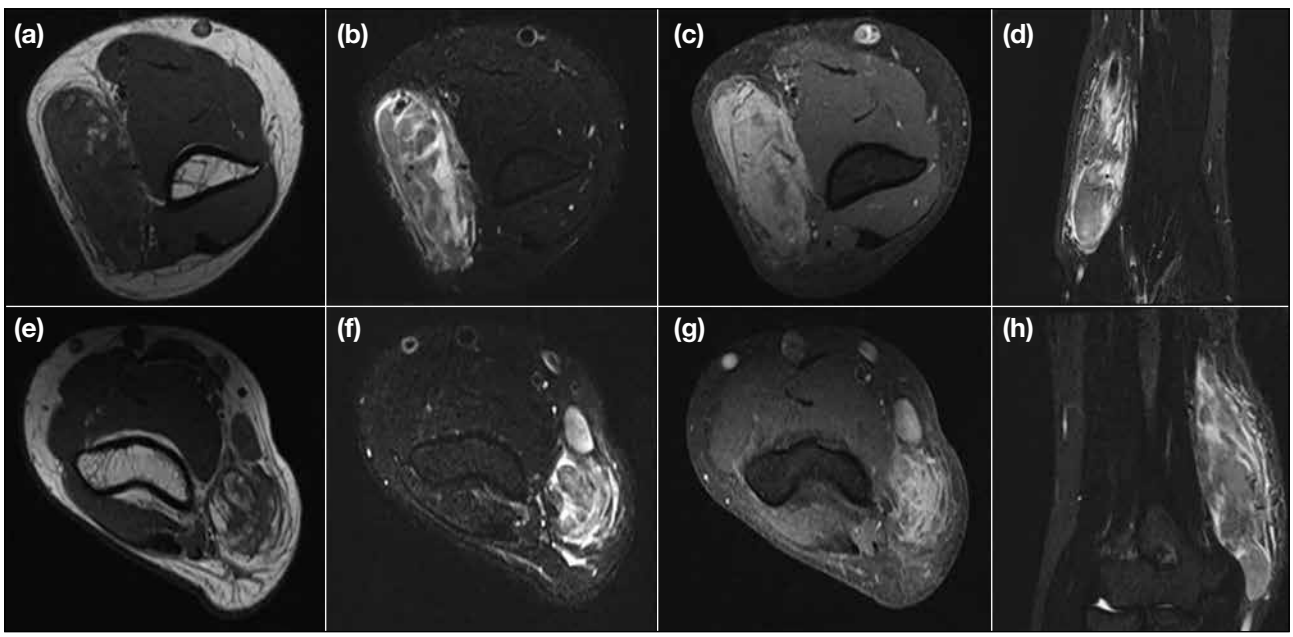

Figure 2. Magnetic resonance imaging of (a-d) left elbow and (e-h) right elbow in case 1. (a) Ill-defined subcutaneous mass with intermediate signal on axial T1-weighted image. (b) Heterogenous intermediate signal on axial T2-weighted fat suppressed image with encasement of basilic vein and internal flow voids. (c) Moderate homogeneous enhancement on axial T1-weighted fat suppressed image with contrast. (d) Medial epitrochlear location with mild adjacent subcutaneous oedema and strandings on coronal T2-weighted short tau inversion recovery image. (e-h) Similar findings at right elbow with additional discrete enlarged oval lymph node.

hyperintense strandings, oedema, and enhancement were also noted. No signs of muscular invasion or bone marrow oedema were evident. Ultrasound-guided core biopsy of both elbow masses revealed a large amount of lymphoid aggregates with eosinophilic infiltrates on a background of stromal fibrosis. Follicular hyperplasia, formation of rare reactive follicles, proliferation of endothelial venules, and perivenular sclerosis were seen. No malignant cells, granuloma or Reed–Sternberg cells were detected. Histopathology results (Figure 3)

were suggestive of Kimura's disease and the patient subsequently underwent local excision of the left elbow mass. Excisional biopsy result of the cervical lymph nodes was also in keeping with Kimura's disease.

Case 2

A 23-year-old man with good past health presented with a 1-month history of a firm painless mass over his left arm. He also complained of incidental left groin swelling. There was no history of trauma. Physical examination confirmed a 4 × 4 cm soft tissue mobile mass at left posterior arm with no skin changes and a 3 × 4 cm non-pulsatile mass of similar consistency at the left groin with lack of cough impulse. An elevated eosinophil count of 3.4×10^9/L (reference range, 0-0.6 × 10^9/L) was noted. Radiograph (Figure 4a) showed soft tissue swelling at the medial aspect of his elbow with no calcification or adjacent periosteal reaction. Ultrasound (Figure 4b) revealed an irregular hypoechoic subcutaneous mass over the left elbow with a few internal hyperechoic bands. MRI (Figure 5) showed an ill-defined T1W isointense, T2W hyperintense subcutaneous mass with homogeneous enhancement at the medial epitrochlear region of the left elbow extending up to mid humerus level. Marked adjacent subcutaneous oedema and fluid together with interlobular septal thickening and enhancement were seen. The basilic vein was encased with preserved flow. Nearby musculature showed normal signal and there was no bone marrow oedema. MRI of the left groin demonstrated a cluster of enlarged lymph nodes with marked subcutaneous strandings and oedema. Ultrasound-guided biopsy of the elbow mass revealed multiple reactive lymphoid follicles with germinal centre formation, heavily infiltrated by eosinophils and bland-looking lymphocytes. Occasional focal eosinophilic microabscesses were seen and vasculature was slightly prominent. Pathological features were suggestive of Kimura's disease. Core biopsy specimen taken from

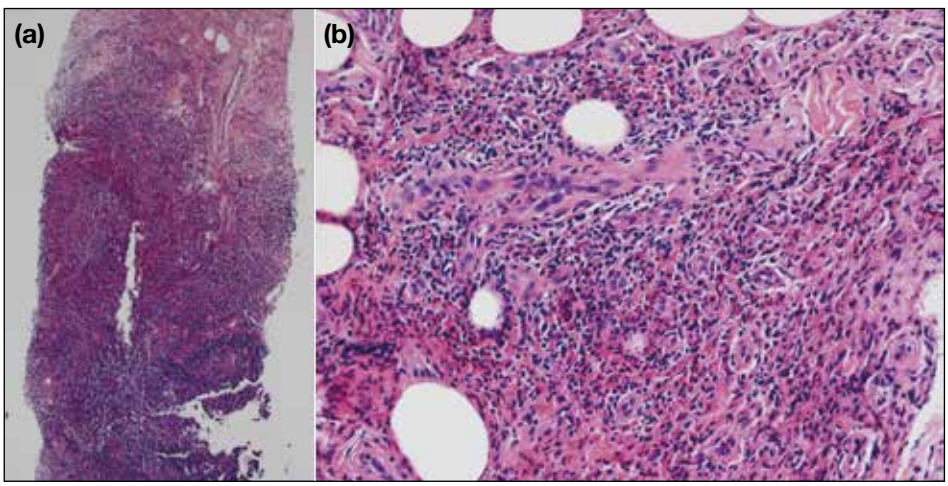

Figure 3. (a and b) Microscopic sections in case 1 showing cores of soft tissue with lymphoid aggregates, formation of rare reactive follicles, moderate amount of eosinophil infiltrates, and capillaries with bland-looking endothelial cells (haematoxylin and eosin staining, magnification unknown).

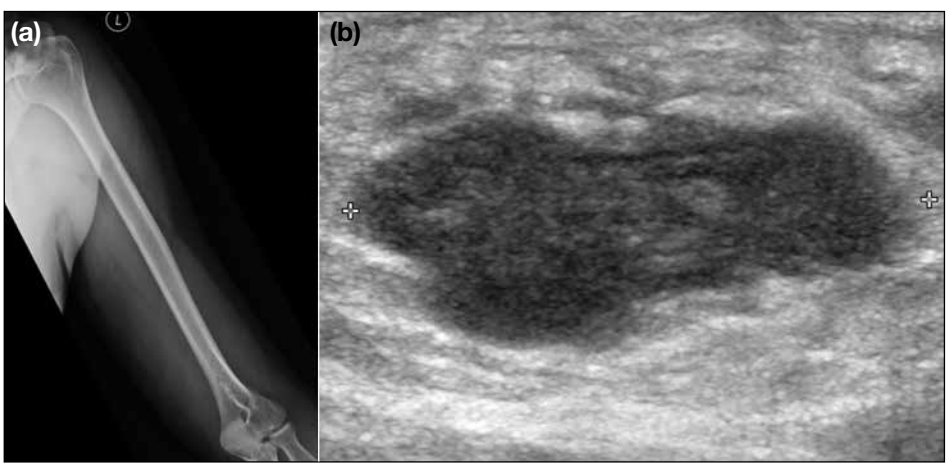

Figure 4. (a) Frontal radiograph in case 2 demonstrated soft tissue swelling at the medial aspect of the left elbow with no calcification or associated bone change. (b) Ultrasound of left elbow in case 2 revealed irregular hypoechoic mass with some internal echoic linear bands.

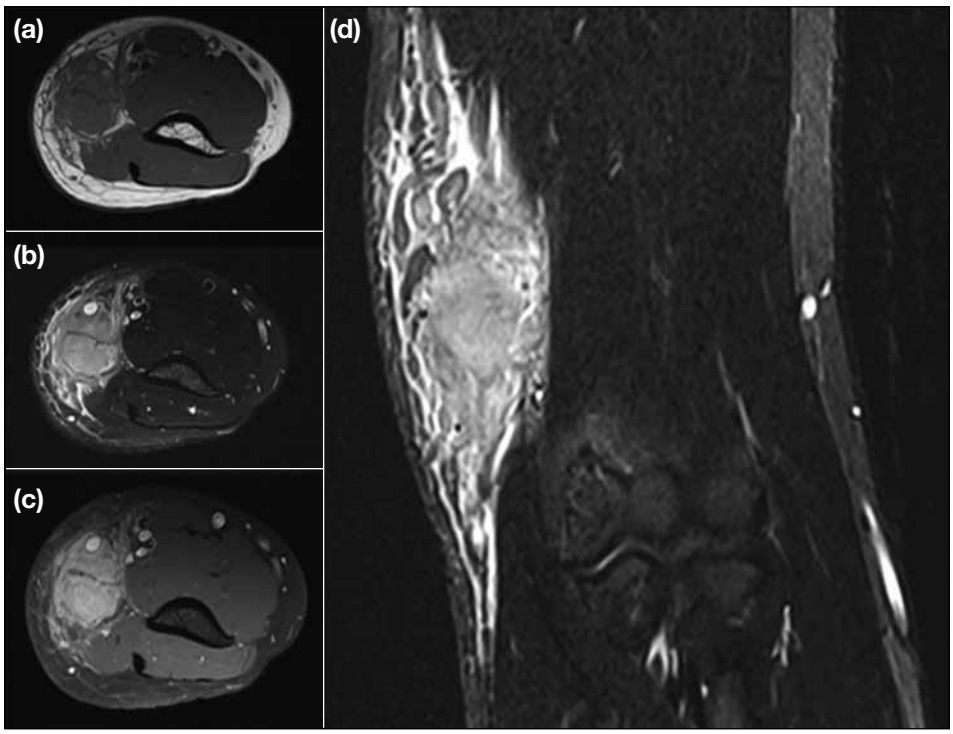

Figure 5. Magnetic resonance imaging of left elbow in case 2. (a) Ill-defined subcutaneous mass with intermediate signal on T1-weighted axial image, (b) hyperintense signal on T2-weighted axial image with encasement of basilic vein, (c) mild homogeneous enhancement on T1-weighted fat suppressed image with contrast, and (d) medial epitrochlear location with extensive surrounding subcutaneous fat strandings and oedema on coronal T2-weighted short tau inversion recovery image.

the left groin lymph node revealed similar findings. The patient was subsequently prescribed a course of oral steroid.

DISCUSSION

Kimura's disease is a rare chronic lymphoproliferative disorder[1] first described by Kim and Szeto in 1937[4] and later by Kimura et al in 1948[5]. It typically affects Asians in their second or third decade of life[6] with a higher incidence among males (male-to-female ratio 3:1). Typical presentation is painless subcutaneous masses in the head and neck, particularly in the parotid and submandibular regions. Other sites of involvement include the oral cavity, axilla, groin, extremities, and trunk. Symptoms have an insidious onset, may be vague or asymptomatic and fluctuate for several years.[7] Aetiology is unknown but there is speculation of an underlying abnormal immune response.

Although imaging findings of Kimura's disease in the head and neck region have been previously reported to be nonspecific and variable, consistent imaging features are reported in Kimura's disease of the upper extremity.[7-9] To date, the largest published study of imaging features of Kimura's disease in the upper extremity is by Choi et al[9] of nine cases. The authors reported partial-to-poorly defined subcutaneous soft tissue masses at the medial epitrochlear region adjacent to medial neurovascular bundles in all cases. The signal intensity on T1W images was similar to or slightly higher than that of muscle, while on T2W images, it was markedly elevated compared to muscle. Enhancement was at least moderately homogeneous. A variable degree of surrounding subcutaneous fat strandings and oedema as well as serpentine flow voids within the masses were also reported. The masses usually caused mass effect on surrounding muscles and encased neurovascular structures without abnormal signal change in the adjacent muscle, neurovascular bundles, bones or joints.[9] Radiographs revealed only nonspecific soft tissue thickening at the medial epitrochlear region with no periosteal reaction or erosion in the adjacent bone.[9] Shin et al[10] reported that ultrasound features of Kimura's disease in the upper extremities were of partially marginated subcutaneous masses with the presence of curvilinear hyperechoic bands and/or dots intermingled within the hypoechoic components. Moderate to severe vascular signals were observed in some of the hyperechoic bands and/or dots on colour Doppler ultrasound. The imaging findings and medial epitrochlear location in our two cases were compatible with previous studies.

On histopathological examination, Kimura's disease characteristically demonstrates lymphoid follicular

hyperplasia with prominent germinal centres, dense eosinophilic infiltrates on a background of abundant lymphoid and plasma cell infiltrates, eosinophilic microabscesses, increased postcapillary venules and perivenular sclerosis.[9] The vascular proliferation likely accounts for the moderate enhancement and flow voids in MRI, and the characteristic subcutaneous strandings and oedema were histologically proven to be proliferation of lymphoid follicles at subcutis.[9]

In both of our cases, there was concurrent involvement of either cervical or groin lymph nodes. Bilateral elbow involvement was also demonstrated in one case. To the best of our knowledge, there have been only four reported cases of bilateral upper limb Kimura's disease.[5,7,8,10] These suggest that Kimura's disease is a systemic disease rather than a locoregional disorder. Other reasons include frequent elevated level of serum immunoglobulin E, peripheral blood eosinophilia, generalised lymphadenopathy, and occasional renal involvement.[11]

Differential diagnoses include tuberculous lymphadenopathy, cat-scratch disease, angiolymphoid hyperplasia with eosinophilia (ALHE), nodular fasciitis, lymphoma, metastatic disease, and soft tissue sarcomas.[12] Tuberculous involvement of lymph nodes would give a matted appearance with central necrosis. Cat-scratch disease similarly affects the medial epitrochlear region but there would also be a contact history, painful swelling, necrotic nodes, positive serology, and more extensive subcutaneous oedema and strandings. It is difficult to differentiate ALHE from Kimura's disease on imaging but when examined through histopathology, ALHE does not show elements of fibrosis or sclerosis that are evident universally in Kimura's disease. Nodular fasciitis presents with rapidly growing painful nodules showing broad fascial contact but Kimura's disease is usually painless. Lymphoma often demonstrates bilateral enlargement of well-defined nodes without changes to subcutaneous fat. Metastatic lymph nodes classically show loss of central fatty hilum, internal necrosis, and capsular vascularity. Soft tissue sarcoma usually manifests as a large circumscribed heterogeneous mass with necrosis and some may show calcification. Kimura's disease tends to have poor margins, lack of necrosis, and absence of calcification.

Management of Kimura's disease is controversial and options range from medical therapy with steroids or cytotoxic drugs to radiotherapy or surgical excision.[13] Surgery is the treatment of choice for a single, well-defined lesion and has the benefit of pathological confirmation. Conservative management is reserved for those with recurrent disease and systemic involvement. Although Kimura's disease has an excellent prognosis with no risk of malignant transformation, the recurrence rate is around 25% to 75% after surgery.[7]

In conclusion, the diagnosis of Kimura's disease requires pathological confirmation and upper limb manifestation is rare. Nonetheless presence of the characteristic imaging appearance along with peripheral eosinophilia in a young Asian male should always raise a suspicion of Kimura's disease. The systemic nature of this entity warrants a meticulous physical examination and imaging to look for generalised lymphadenopathy and bilateral involvement.

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