An Unusual Clinical Presentation of Kimura Disease Manifesting with a Typical Cephalocervical Lesion and an Atypical Subcutaneous Hip Mass Lesion

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Abstract

Kimura’s disease (KD) is a rare, lymphoproliferative inflammatory disorder, typically presenting as firm, painless and pruritic single or multiple subcutaneous lesions in the head and neck, especially in the parotid and submandibular regions. We herein report a case of a 39-year-old Japanese man presenting with a typical cephalocervical KD lesion around the salivary glands with a rare association with a distant subcutaneous mass at the hip. We also emphasize the radiologically and clinically important features in the differential diagnosis and management. To the best of our knowledge, this case is the first report of KD manifesting with a typical cephalocervical lesion and an atypical subcutaneous hip mass lesion.

Key words: Kimura disease, radiology, head and neck, parotid gland, hip

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Introduction

Kimura disease (KD) is a rare, chronic and benign lymphoproliferative inflammatory disorder of unknown etiology, which is typically seen in young Asian men in the second or third decade of life (1, 2). Painless subcutaneous masses, regional lymphadenopathy, eosinophilia and elevation of serum immunoglobulin E (IgE) levels are the characteristic clinical features of KD (3). Histologically, prominent lymphoid follicles, vascular proliferation and eosinophilic infiltration have been reported (4). Radiological findings described in the literature include subcutaneous head and neck nodules, which are often misdiagnosed as salivary gland malignancy (5). These lesions may also occur in the axilla, groin, trunk, abdomen and peripheral limbs (6). A PubMed and Medline search using the key words “Kimura’s disease or Kimura disease” revealed no reports of a rare association of typical head and neck findings with distant subdermal hip and buttock lesions. We herein, report a rare case of KD with such an association.

Case Report

A 39-year-old Japanese man presented with left buccal region and buttock swelling, which he had experienced for several years, while he was hospitalized for a perforated duodenal ulcer. His medical and family histories were unremarkable for any systemic, infectious or malignant diseases. His laboratory work-up at the time of admission revealed elevated serum IgE levels (7,000 IU/mL; NR ≤ 170 IU/mL) and eosinophil counts (3,693/μL; NR ≤ 510/μL). The neutrophil count was within normal limits (8,100/μL; NR ≤ 8,500/μL). Other blood test results including those for blood urea nitrogen and serum creatinine levels, and the urinalysis results were normal. On enhanced computed tomography (CT)
Imaging, a mass lesion centered in the left buccal region with an irregular shape and a poorly-defined margin was detected (Fig. 1A). The T2-weighted magnetic resonance (MR) image revealed a hypo-intense subcutaneous tissue mass with an irregular shape and a poorly-defined margin, and diffuse thickening of the buccal area of the left cheek. C: Another T2-weighted image showed subcutaneous lesions in front of the anterior wall of the left maxillary sinus (arrow), anterior to the left masseter muscle (arrow head) and anterior to the left parotid glands (thick arrow).

Figure 1. CT and MR images of the cephalocervical lesions. A: On enhanced CT, a mass-like lesion (arrow) centered in the left buccal region with an irregular shape and a poorly-defined margin was detected. The lesion was also accompanied by swelling of the perilesional subcutaneous tissues. B: A T2-weighted image showed a hypo-intense subcutaneous tissue mass (arrow) with an irregular shape, poorly-defined margins and diffuse thickening of the buccal area of the left cheek. C: Another T2-weighted image showed subcutaneous lesions in front of the anterior wall of the left maxillary sinus (arrow), anterior to the left masseter muscle (arrow head) and anterior to the left parotid glands (thick arrow).

Figure 2. An abdominal CT image of the hip lesion. A: A subcutaneous left hip mass (arrow) was detected by the abdominal CT during admission. B: The follow up CT scan one month after admission. Note the lack of change in the size of the mass (arrow).

Discussion

KD is a rare idiopathic entity that has been linked to allergic reactions and/or autoimmune responses characterized by peripheral blood eosinophilia and elevated serum Ig E levels (1, 6). A close relationship between the lesion size and the degree of eosinophilia was reported (1). It has been
Figure 3. Histopathology of the cervical and buttock lesions. A: A biopsy specimen of the neck lesion. Note the reactive large lymphoid follicles with enlarged germinal centers. B: A biopsy specimen of the neck lesion. Note the marked interfollicular eosinophilic infiltration. C: A biopsy specimen of the buttock lesion. Note the abscess formation with eosinophilic infiltration.

speculated that a viral or parasitic trigger may alter the T-cell immunoregulation or induce an IgE-mediated type 1 hypersensitivity, resulting in the release of eosinophilotrophic cytokines (1).

Clinically, the lesions are often misdiagnosed as salivary gland malignancy because of the diffuse nature of the disease (7). Other reported clinical associations include atopic dermatitis, allergic rhinitis, asthma, urticaria and renal disease (6).

Histopathologically, KD is characterized by the formation of multiple lymphoid follicles with germinal centers, many of which are infiltrated by plasma cells, lymphocytes, mast cells and especially eosinophils leading to folliculolysis (2). The lesions also demonstrate vascular proliferation and stromal fibrosis (1). The fibrosis can be present in the early stage of the disease and may be replaced later by hyalinization (1). Eosinophilic infiltration is usually considerable, with formation of eosinophilic abscesses as seen in the current case (2). Histologically, KD usually has three components including eosinophil infiltration with follicular hyperplasia, fibrocollagenous proliferation and arborizing vascular proliferation of the postcapillary venules (3, 6).

KD typically presents as firm, painless or pruritic single or multiple subcutaneous lesions with lymphadenopathy presenting in the head and neck, especially in the parotid and submandibular regions (1, 3, 4). Less common sites in the head and neck include the paranasal sinuses, orbits (including the eyelids, conjunctiva and lacrimal glands), tongue, epiglottis, ear lobe, tympanic membrane and parapharyngeal space (1, 6-8). Other less common sites outside of the head and neck include the axilla, groin, trunk, abdomen, peripheral limbs, chest wall and median nerve (1, 6, 8). In the literature, solitary hip/buttock lesions have been reported in just two cases (2, 4) and an unusual palatine presentation associated with an inguinal lesion was reported in one case (6). The current case manifested with a typical head and neck presentation coexisting with hip/buttock lesions. This is the first time such a concomitant presentation has been reported. A thorough clinical and laboratory examination excluded atopic dermatitis, allergic rhinitis, asthma, urticaria and renal disease. A biopsy of the head/neck lesions showed eosinophilic infiltration and helped to exclude salivary gland malignancy as a possibility. In the previously reported case, abdominal CT scans helped in the diagnosis of KD (4). In our case, an abdominal CT scan performed during the examination of the duodenal ulcer led to the detection of the hip/buttock lesions. Considering that KD is a systemic lymphoproliferative disorder resembling malignant lymphoma and IgG4 related diseases, we believe that systemic CT imaging in cases with a typical presentation around the head and neck soft tissues could help in the initial differential diagnostic work-up. This could be followed
by biopsies of the mass lesions to reveal the nature of the proliferative lesions and to differentiate them from salivary gland tumors as well as metastatic lymphadenopathy, lymphomas, angiolymphoid hyperplasia with eosinophilia, Langerhans cell histiocytosis, florid follicular hyperplasia, Castleman disease, drug reactions, dermatopathic lymphadenopathy, parasitic lymphadenitis and Churg and Strauss type of allergic granulomatosis (7, 9). It should be noted that the parotid tumors are usually encapsulated and are limited to the parotid gland while KD frequently manifests with irregular extensions into the adjacent subcutaneous tissues. Although lymphoma and metastatic lymphadenopathy may have similar presentations, the specific distribution of the lesions in KD and the irregularity of the lesion margin as well as the long clinical course definitely help in making a differential diagnosis (1).

There have been several reports suggesting that the radiologic findings of KD of the head and neck are nonspecific and variable (1, 5, 7). On CT scans, either well-defined nodular masses or ill-defined plaque like infiltrative masses in the subcutaneous tissues associated with lymphadenopathy have been reported as the typical findings (1, 6, 7). The attenuation of the masses varies from iso-to hyperdense to the muscle tissue (1). On MR images, the masses are reported to show variable signal intensity; low-to-high signal intensity on T1- and T2-weighted images (1, 7). On the T2 weighted image, prominent fibrotic tissues may appear as low signal intensity areas (7). Differing degrees of fibrosis and vascular proliferation lead to variable contrast enhancement and patterns of signal intensity in these masses (7). In the current case, all lesions showed heterogeneous hyperintensity on STIR and T2 weighted images.

Regarding the treatment of KD, conservative therapies such as radiotherapy and systemic steroids, or surgical resection have been advocated (10). The standard treatment for KD involves the use of oral steroids, although termination of the steroid therapy often results in recurrence of the disease (4). Systemic steroids effectively reduce disease activity and are indicated in cases of frequent recurrence, or when an associated nephrotic syndrome is present (10).

A surgical excision is usually successful when there is a single lesion or when the disease has been generally confined, and the patient is surgically amenable (10). However, as the disorder is benign, routine surgical excision cannot be recommended (10). Low-dose radiation therapy has been attempted, with varying degrees of success (3).

We herein described a case of KD manifesting with a typical head and neck presentation around the salivary glands with a rare association with a distant bilateral subcutaneous mass present at the hip. The diagnosis of KD entails a multidisciplinary collaboration employing radiological imaging to suggest a possible diagnosis and pathologic confirmation for a definite diagnosis.

The authors state that they have no Conflict of Interest (COI).

References


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