LETTER TO EDITOR (VIEWERS CHOICE)

KIMURA DISEASE IN A 12 YEAR OLD BOY

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A 12 year old male presented with a history of painless right sided cervical lumps for 2 months duration. There was no history of febrile illness, pharyngitis, cough, rash or diarrhea. He had never received blood transfusions. Physical examination showed an apparently well boy with cluster of non-tender, firm lumps in right posterior triangle of the neck largest being 3.5 x 2 cm. Overlying skin was not inflamed. Laboratory investigations revealed hemoglobin 11.4g/dl, white cell count 19100 cells/cumm with 15% of them being eosinophils, platelets 220,000 cells/cumm and peripheral smear showed marked eosinophilia with reactive lymphocytes. Computed tomography of head and neck showed no soft tissue masses. There were slightly prominent lymph nodes in the level II, III in the neck and supraclavicular fossa. Chest radiography and renal functions were normal. Urine examination was normal. Excision biopsy of the largest lump was taken to rule out malignancy. Microscopy of the sample revealed a lymph node with preserved nodal architecture. The cortex showed reactive follicular hyperplasia and the paracortex showed heavy eosinophils infiltrate associated with eosinophil abscess formation with perinodal infiltration rich in eosinophils. There was no malignancy. Based on the peripheral hypereosinophilia and histology, a clinical diagnosis of Kimura disease was made.

Kimura disease is an uncommon idiopathic chronic inflammatory disorder that most commonly presents as painless lymphadenopathy or subcutaneous masses in the head and neck region. (1) The first report of Kimura disease was from China in 1937, in which Kimm and Szeto described seven cases of a condition they termed "eosinophilic hyperplastic lymphogranuloma." (1) The disorder received its current name in 1948, when Kimura et al noted the vascular component and referred to it as an "unusual granulation combined with hyperplastic changes in lymphoid tissue". (1) Kimura disease usually affects young and middle-aged Asian males. This benign condition is characterized by a triad of painless subcutaneous masses in the head and neck, prominent eosinophilia and markedly elevated immunoglobulin E (IgE) levels. (2) The solitary lesions are usually in deep subcutaneous tissues, frequently associated with regional lymphadenopathy and salivary gland involvement and clinically may mimic a neoplasm, including acute non-lymphocytic leukemia, Hodgkin disease and follicular lymphoma. (2) Untreated, these masses tend to slowly enlarge and may eventually become disfiguring. (3) Local or generalized pruritus and subacute or chronic dermatitis may occur. Proteinuria may occur in 12% to 16% of cases. Nephrotic syndrome is the most common presentation of renal involvement with a wide spectrum of histologic lesions being described. (2) Our patient had normal renal function and no proteinuria.

Pathophysiology of Kimura disease is not better understood but may relate to a disturbance in the normal rate of production of eosinophils and IgE, currently believed to be a product of an interaction between types 1 and 2 T helper cells which could result in excessive elaboration of eosinophilotrophic cytokines such as interleukin 4. Allergic or parasitic etiologies have been actively sought, but not identified. (3) The clinical course of Kimura disease is generally benign and self-limited.

Treatment of Kimura disease is problematic. At initial presentation, surgical biopsy is the most frequent diagnostic procedure and excision may be curative. (4) The recurrence is common, occurring up to 25% of cases treated with surgical excision alone. Other therapeutic options, including radiation, systemic corticosteroids and cytotoxic agents have all been tried with variable responses. (5)

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