Kimura disease

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Case report

A 14 year old adolescent boy presented with left parotid and left post-aural lumps of 1 year duration, associated with pain. His growth was normal and there was no fever. Abdominal examination did not show enlargement of the liver or spleen. His white cell count and peripheral blood film showed marked eosinophilia (35%). Roentgenograms of the chest and bones did not show any abnormalities. Skin tuberculin test was negative. Urinalysis was normal. Serum Ig E was 780 IU/ml (normal <200 IU/ml). Magnetic resonance imaging of neck soft tissue showed left parotitis and left cervical lymphadenopathy. The parotid and post-aural lumps were excised and sent for histopathology. Histopathology showed lobules of serous glands with many interspersed hyperplastic lymphoid follicles with prominent germinal centres and cuff of mantle zone, numerous eosinophils in stroma, infiltrating the lymphoid follicles and serous glands with focal clustering forming micro-abscesses and many thin walled vascular spaces with flattened endothelial cells, consistent with Kimura disease (Figures 1 and 2).

Left near total parotidectomy and left modified radical neck dissection were done. On follow up, child developed post-aural swellings bilaterally. As his disease is progressing we have planned to give him intravenous immunoglobulin. We are doing his urine analysis on follow up visits as Kimura disease patients are prone to develop nephrotic syndrome.

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The authors declare that there are no conflicts of interest

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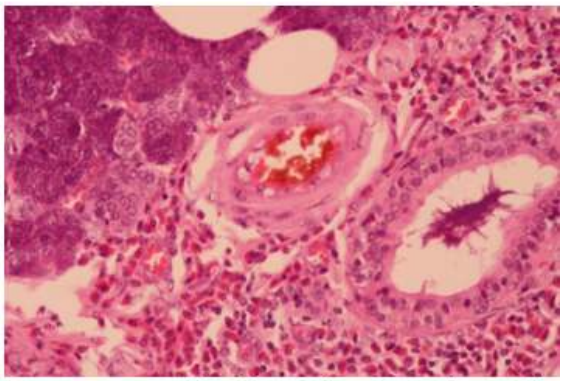

Figure 1: ***Parotid with periductal eosinophilic infiltrates***

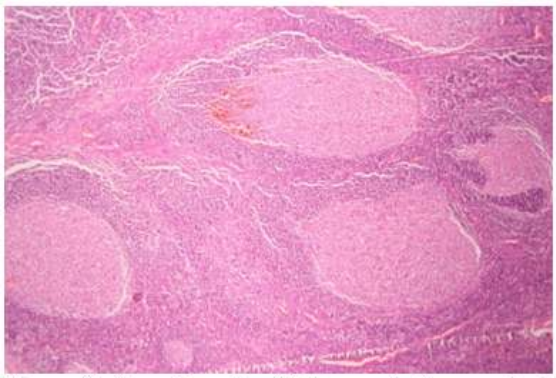

Figure 2: ***Lymph node with hyperplastic germinal centre***

Discussion

Kimura disease is a chronic inflammatory disorder of unknown aetiology, involving subcutaneous tissue, salivary glands and lymph nodes with characteristic tissue and peripheral eosinophilia and an elevated IgE level[1]. It is primarily seen in young Asian males[1]. The constant histologic features seen in this disease are preserved lymph node architecture, florid germinal centres, eosinophilic infiltration, and an increased amount of post-capillary venules[2,3]. Surgical excision of lesions is the initial mode of therapy but relapses are common[4]. Systemic steroid therapy is indicated for relapses but it is difficult to withdraw the steroids without another relapse occurring[4]. Malignant transformation has not been reported in Kimura disease[4]. Intravenous immunoglobulin (IVIG) can decrease enhanced IgE production both in vivo and in

vitro[5,6]. In fact, IVIG has been successfully used as a steroid sparing agent in the treatment of an 8 year old boy with Kimura disease[7]. Renal involvement can occur in over 50% of patients with Kimura disease and usually takes the form of nephrotic syndrome[8,9].

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