Kikuchi Disease in a Child

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Abstract
Kikuchi disease, also called histocytic necrotizing lymphadenitis or focal histiocytic lymphadenitis, is a rare, idiopathic and generally self limited cause of lymphadenitis. It was first described in 1972 in Japan[1]. The most common clinical manifestation is cervical lymphadenopathy with or without systemic symptoms & signs. It almost always runs a benign course and resolves in several weeks to months.[2]

Case Report
A 12 year old girl presented with a six week history of swelling on the right side of her neck which had gradually increased in size. She had some pain on movement of her neck. There was no history of fever, rash, pallor, itch, bruising or any other systemic symptoms. A 5 day course of Flucloxacillin had no effect. Examination showed a healthy, afebrile child, with a 5˜»4 cm mobile, non-tender right sided cervical swelling. There was no overlying inflammation or other abnormal clinical findings. Routine laboratory investigations were normal apart from leucopoenia (total WBC 3.1, lymphocytes 1.2). An excision biopsy of the lymph node was performed. Normal lymph node architecture was preserved but the node showed multiple discrete areas of necrosis containing eosinophilic material and karyohectic debris. There was no significant neutrophil, eosinophil or plasma cell infiltrate in the areas of necrosis.Cells present were predominantly macrophages and T lymphocytes. This was confirmed by appropriate immunohistochemistry .These appearances are typical of histiocytic necrotizing lymphadenitis (Kikuchi Disease). The patient recovered completely without further intervention.

Discussion
Kikuchi Disease has been recognised worldwide and is well discussed in pathologic literature, but reports in children are very rare. The youngest age reported is 19 months old.[3] The etiology of Kikuchi Disease is unknown. A viral pathogenesis has been long favoured, because of the self limiting clinical course, lack of neutrophil response and failure to response to antibiotics. However a specific virus has not been identified. The histologic and immunologic findings suggest a hyper immune reaction to unidentified antigens. An autoimmune contribution to the pathogenesis is suggested by observations that the disease may precede or occur in association with systemic lupus erythematosus. Cervical lymphadenopathy and fever are the most common presentations.The posterior cervical lymph nodes are the group usually affected but axillary, epitrochlear, mediastinal inguinal, intraparotid and iliac nodes can also be involved. The size of the affected lymph node is usually 2-3cm. Although nodes larger than 5cm have been described. Some patients with lymphadenopathy complain of malaise, fatigue, night sweats, weight loss and gastroenteric symptoms. Transient rash similar to rubella or drug eruption may be seen in some patients. eucopoenia has been reported in up to 58% of the patients and was described as characteristic in a Japanese report.[4] ESR and CRP may be elevated or mildly abnormal LFTâ S may occur.

The diagnosis of Kikuchi disease is confirmed by excision biopsy and microscopic examination. Histologic findings include Paracortical necrosis which may be patchy or confluent. Histiocytes with crescent shaped nuclei. Histiocytes and macrophages containing phagocytised debris from degenerated lymphocytes. ther cells -lymphocytes, plasmacytoids, monocytes, macrophages and immunoblasts (predominantly T-cells). Kikuchi Disease usually remits within 1-6 months.[4,5] Treatment is symptomatic using NSAIDâ s to relieve fever, flu-like symptoms or lymph node tenderness. Our patient was symptom free within a month.

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