

Case Report

A RARE CASE OF KIKUCHI'S DISEASE – CASE REPORT

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ABSTRACT

Kikuchi's disease is a necrotizing lymphadenitis that is prevalent in Asia and is being increasingly recognized in other areas of the world. It usually occurs in women in their late 20s or early 30s and manifests as a posterior cervical adenopathy. It resolves spontaneously, usually over a period of several weeks to 6 months. Its initial clinical appearance is commonly similar to that of a lymphoma, and it can be pathologically misdiagnosed as such. Kikuchi's disease might be associated with systemic lupus erythematosus. We report a case of Kikuchi's disease that occurred in a 36-year-old woman. We discuss the clinical features, differential diagnosis, radiographic evaluation, and pathology of this case, and we review the literature in an effort to assist otolaryngologists in diagnosing this benign and uncommon entity.

CASES

A 36 year old female presented in YCRH Latur, with a two week history of fever, anorexia and weight loss. There were no other complaints. She had been previously fit and well and was on no medication. On examination She was lethargic, but otherwise looked well. She was afebrile and haemodynamically stable. Significant findings were generalised lymphadenopathy, palpable in the occipital, posterior auricular, cervical, axillary and inguinal regions. There was 2 cm smooth, tender hepatomegaly. Blood tests revealed neutropenia, hyponatraemia, raised alkaline phosphatase and C-reactive protein. Electrocardiogram and chest radiographs were normal. Her initial management consisted of fluid restriction and regular paracetamol, whilst results of further tests were awaited. These included blood and sputum cultures, autoimmune and viral screen.

Subsequently the axillary and inguinal lymphadenopathy resolved along with reduction in size and tenderness of the cervical lymph nodes. There were persistent intermittent temperature spikes and a two day episode of self-resolving diarrhoea.

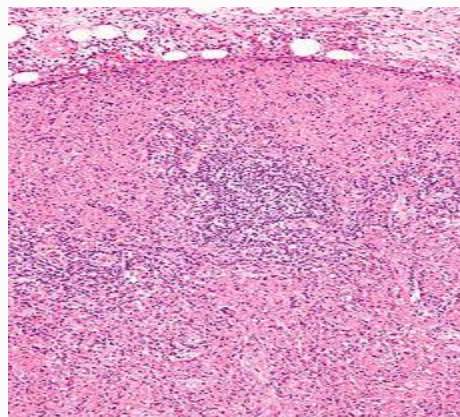


Figure 1: Showing Acute necrotizing Lymphadenitis

Blood, urine and stool cultures were negative. Sputum cultures grew respiratory tract flora and were negative for acid-fast bacilli. Paul Bunnell test was negative. The autoimmune screen was negative, as was toxoplasma and cytomegalovirus screens.

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Computerised tomography demonstrated generalised lymphadenopathy in the supraclavicular area; axillae, posterior mediastinum, para-aortic areas and deep in the pelvis and groin. The abdominal viscera were normal. Excisional biopsy of a cervical lymph node confirmed a diagnosis of Kikuchi-Fujimoto disease (KFD). Histological analysis showed histiocytic granulomatous infiltration with widespread necrosis of the lymph node extending beyond its capsule into the surrounding fat (Figure 1). No active treatment was instigated and the patient was discharged home after a two week in-patient stay. At follow-up she reports no symptoms, remains well and there are no abnormalities on clinical examination.

DISCUSSION

Kikuchi-Fujimoto disease (KFD) was described in 1972 in Japan. It is also known as Kikuchi disease, histiocytic necrotizing lymphadenitis, Kikuchi necrotizing lymphadenitis, phagocytic necrotizing lymphadenitis, subacute necrotizing lymphadenitis, and necrotizing lymphadenitis (Rammohan, Cherukuri, 2012).

It was first described in Japan by Dr Masahiro Kikuchi in 1972 and independently by Y. Fujimoto. Kikuchi-Fujimoto disease (KFD) is a rare, self-limiting disorder that typically affects the cervical lymph nodes. Recognition of this condition is crucial, especially because it can easily be mistaken for tuberculosis, lymphoma, or even adenocarcinoma. Awareness of this disorder will help prevent misdiagnosis and inappropriate treatment (Rammohan, Cherukuri, 2012). Kikuchi's disease is a very rare disease and mainly seen in Japan. Isolated cases are reported in America, Europe and Asia. It is mainly a disease of young adults (mean age, 20–30 years), with a slight bias towards females. The cause of this disease is not known although infectious and autoimmune etiologies have been proposed. Course of the disease is generally benign and self-limiting. Lymphadenopathy most often resolves over several weeks to six months. Recurrence rate is about 3%. Mortality is extremely rare and usually due to hepatic, respiratory, or cardiac failure. Some studies have suggested a genetic predisposition to the proposed autoimmune response. Several infectious candidates have been associated with Kikuchi's disease (Atwater, 2008).

There are many postulates about the etiology of KFD. A microbial/viral or autoimmune cause has been suggested. *Mycobacterium szulgai*, *Yesinia* and *Toxoplasma* have been implicated. More recently there has been growing evidence of the role of Epstein-Barr virus, as well as other viruses (HHV6, HHV8, Parvovirus B19, HIV- and HTLV-1) in the pathogenesis of KFD (Kaushik, 2004). But serologic tests including antibodies to a host of viruses have consistently proven noncontributory and no viral particles have been identified ultrastructurally. It is now proposed that KFD is a nonspecific hyperimmune reaction to a variety of infectious, chemical, physical and neoplastic agents. In addition to SLE, other autoimmune conditions and manifestations such as antiphospholipid syndrome, polymyositis, systemic juvenile idiopathic arthritis, bilateral uveitis, arthritis and cutaneous necrotizing vasculitis have been linked to KFD. It is possible that KFD may represent an exuberant T-cell mediated immune response in a genetically susceptible individual to a variety of non-specific stimuli (Bosch, 2006). The signs and symptoms of Kikuchi's disease are fever, lymphadenopathy, skin rashes and headache. Rarely hepatosplenomegaly and nervous system involvement resembling meningitis is seen. Differential diagnosis includes SLE, disseminated tuberculosis, lymphoma, Sarcoidosis, and viral lymphadenitis. Clinical findings sometimes may include positive results for IgM/IgG/IgA Antibodies. It is diagnosed by lymph node excision biopsy (Bosch, 2004).

CONCLUSION

Kikuchi's disease is a self-limiting illness which has symptoms which may overlap with Hodgkin's lymphoma leading to misdiagnosis in some patients so careful biopsy evaluation is mandatory for every case of cervical lymphadenopathy. ANA, APLA, Anti-dsDNA, RF are usually negative, and may help in differentiating from SLE. No specific cure. Treatment largely supportive. NSAIDs for tender lymphnodes

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and fever, corticosteroids are useful in severe extranodal or generalized disease. Symptomatic measures aimed at relieving the distressing local and systemic complaints have been described as the main line of management of KFD. Analgesics, antipyretics, nonsteroidal anti-inflammatory drugs (NSAIDs) and corticosteroids have been used. If the clinical course is more severe, with multiple flares of bulky cervical lymphadenopathy and fever, then a low-dose corticosteroid treatment has been suggested.

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