SAS Journal of Surgery ISSN 2454-5104
SAS J. Surg., Volume-3; Issue-5 (May, 2017); p-138-140
Available online at http://sassociety.com/sasjs/

Review Article

Kikuchi Fujimoto Disease: A Rare Benign Disease

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Abstract: We describe a rare cause of cervical lymphadenopathy in a female, outline the clinical and histopathological features and discuss excision biopsy as the investigation of choice in this age group of fourth decade. A thirty - two year old female was referred to our institute with a three-month history of RIGHT cervical lymphadenopathy. She reported no other symptoms and haematological investigations were normal. Excision biopsy revealed extensive histiocytic necrotising lymphadenitis providing a diagnosis of Kikuchi-Fujimoto disease. Persistent cervical lymphadenopathy in the 16-40 year old age group warrants to rule out serious pathology such as metastatic cancer, lymphoma, Koch's or HIV. Once the examination and haematological work up is negative, we proceed to excision biopsy as the quickest way to obtain a diagnosis, which sometimes comes up with an unexpected Kikuchi-Fujimoto disease.
Keywords: Kikuchi-Fujimoto disease, cervical lymphadenopathy, Excision biopsy.

INTRODUCTION

The disease was first described in Japan in 1972. More recently, the disease has been reported throughout the world and in all races [1]. In adults over the age of 40, when lesions of thyroid and salivary glands are excluded, majority (75%) of neck lumps are malignant. Metastatic lymphadenopathy from a carcinoma of the aero digestive tract is also of particular significance, especially if there is a history of heavy smoking and drinking, and Localisation of the primary tumour must be made without delay. In contrast, cervical lymphadenopathy in patients aged between 16-40 years is mostly of infectious etiology; other causes in this age group include Koch's, rare inflammatory processes and neoplastic lesions such as lymphoma. We describe a 32-year-old lady who presented with cervical lymphadenopathy due to Kikuchi-Fugimoto disease (KFD, or subacute necrotising histiocytic lymphadenitis).

CASE REPORT

A 32-year-old female became aware of lump in her RIGHT side of neck in mid April 2012. She was an old treated case of Koch's a decade prior. On examining level II, III & IV nodes were enlarged, which were painless, firm but mobile, no axillary or groin nodes were palpable. She was apyrexic, haematological investigations at that time revealed a Hb - 11g/dl, TLC - 5700, ESR of 12 mm/hr. The patient was a non-smoker who denied any weight loss, itching or night sweats and was in otherwise good health. An excision biopsy was performed and submitted for histopathological assessment. The striking feature of the lymph node on microscopy was the presence of extensive areas of geographic necrosis bounded by relatively broad zones of histiocytes and activated lymphoid cells. Many apoptotic cell fragments were present being ingested by histiocytes but there was no neutrophil infiltrate, multinucleate giant cell formation or granulomatous inflammation. In other areas of the node, there was expansion of the Para cortical regions with many dispersed histiocytic cells; only a few small reactive lymphoid follicles were present. The histopathology suggested a necrotising lymphadenitis without neutrophil infiltration, diagnostic of KFD - occasionally such changes can be associated with systemic conditions, in particular systemic lupus erythematosus (SLE). The patient was tested for autoantibodies including ANA, the most commonly detected autoantibody in SLE; the results were negative. Patient received symptomatic treatment for the same for a week and had a spontaneous uneventful recovery. The patient is followed-up for four years.

DISCUSSION

Kikuchi Fujimoto disease is a subacute necrotizing lymphadenopathy of unknown cause that is more common among young Asian women, usually affects cervical lymph nodes, and is characterized histologically by histiocytic proliferation and necrosis of lymph nodes [2].The differential diagnosis of enlarged cervical lymph nodes is a discussion beyond

the scope of this article but the principal conditions to be distinguished in cervical lymphadenopathy are lymphoma, metastatic tumour, Koch's, reactive conditions such as infectious mononucleosis, human immunodeficiency virus (HIV),. In this case there was no history of previous surgery or clinical findings suggesting metastasis from tumour elsewhere; there were no oral or head and neck manifestations to suggest systemic viral infection, including HIV. There was no skin lesion in the drainage area to indicate a local inflammatory cause for the lymph node enlargement. KFD was first described independently in 1972 by Kikuchi [3] and Fugimoto [4]. In 1982 the first cases of KFD were reported in North America and Europe [5] and the disease is now recognized worldwide. KFD affects young women peak age being the third decade [6], rare below 16 years. The ratio between females and males is 4:1, gender differences being less apparent in Asian populations. The aetiology of KFD remains unclear. Infectious agents include upper respiratory tract infections and several viral infections caused by cytomegalovirus, Epstein-Barr virus [7, 8]**;** have been postulated as causative although no relationship has yet been established. Due to the similar histology seen in KFD and the lymphadenitis of SLE and that both diseases most often occur in young females, Dorfman and Berry [9] suggested KFD could be an attenuated form of SLE. Another explanation [10] proposed that KFD might be a self-limiting SLE-like autoimmune reaction to viral infected transformed lymphocytes. It is likely that the development of KFD is a multifactorial process involving environmental, biological and genetic influences. The majority present with cervical lymphadenopathy, the posterior cervical triangle being the most commonly affected site [11]. Other less common signs and symptoms include splenomegaly, fever and weight loss – one third of patients have a rash at presentation, findings that can heighten the clinical overlap with infectious mononucleosis, SLE and lymphoma. A number of non-specific haematological abnormalities may occur in KFD. Approximately 50% of cases show a mild neutropenia [12] with leucopenia also present in 25-40% of cases [4]. Other non-specific findings include a raised CRP and ESR but their absence does not exclude KFD. Fine needle aspiration cytology (FNA) can diagnose KFD but exclusion of non-Hodgkin's lymphoma may not be possible on FNA. Excision biopsy confirms the diagnosis. The histopathological features of KFD are quite distinctive and the only mimic is SLE lymphadenitis. The lymph node changes in Kawasaki disease (mucocutaneous lymph node disease), cat scratch disease and atypical mycobacterial infection are quite different, being characterised by intravascular fibrin thrombi and neutrophils, stellate micro abscesses with neutrophils and necrotising stellate granulomatous inflammation respectively [13]. KFD is a self-limiting condition that rarely requires specific treatment in most cases. Management is, therefore, based on supportive therapy, such as analgesia and anti-inflammatory medication. In patients with neurological symptoms or in those cases where KFD is found with another medical condition, immunosuppression with corticosteroids appears to improve the patient's condition rapidly. A high initial oral dose of prednisolone with a subsequent reducing dose is the advocated regime [14, 15]. There are reports of excellent responses to hydroxy chloro quine [16, 17], immunoglobulins [18], and minocycline [19]. Recurrence of KFD is approximately 3% [20].

CONCLUSION

We describe a case of KFD presenting as an unusual cause of cervical lymphadenopathy that mimicked lymphoma. The histopathological findings were classical and the diagnosis of KFD could be achieved without difficulty.

The authors have no conflict of interest.

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