International Journal of Research in Medical Sciences
Panchal A et al. Int J Res Med Sci. 2017 May;5(5):2243-2245
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: http://dx.doi.org/10.18203/2320-6012.ijrms20171878

Case Report

Kikuchi Fujimoto lymphadenitis- an uncommon entity in the surgical outpatients: a case report

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Received: 23 February 2017
Accepted: 27 March 2017

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ABSTRACT

Kikuchi Fujimoto lymphadenitis is a benign self-limiting disease with subacute necrotizing lymphadenopathy of unknown cause. The clinical, histopathological and immunohistochemical features point to viral etiology hence delaying diagnosis commonly. Clinicians and pathologists awareness of this disorder may prevent misdiagnosis and inappropriate treatment. We describe here a case of a young woman admitted under our care with fever and cervical lymphadenopathy. Cervical lymph node biopsy revealed the rare clinical disorder of Kikuchi Fujimoto lymphadenitis which is not thought of as a primary cause of lymphadenopathy in the Indian subcontinent where tuberculosis is widely prevalent.

Keywords: Kikuchi Fujimoto, Necrotizing lymphadenitis

INTRODUCTION

Kikuchi Fujimoto lymphadenitis is a benign self-limiting disease with subacute necrotizing lymphadenopathy of unknown cause. It is more common among young Asian women, usually affects cervical lymph nodes, and is characterised histologically by histolytic proliferation and necrosis of lymph nodes.[1]

The clinical, histopathological and immunohistochemical features point to viral etiology hence delaying diagnosis commonly. Clinicians and Pathologists awareness of this disorder may prevent misdiagnosis and inappropriate treatment. We describe here a case of a young woman admitted under our care with fever and cervical lymphadenopathy. Cervical lymph node biopsy revealed the rare clinical disorder of Kikuchi Fujimoto lymphadenitis which is not thought of as a primary cause of lymphadenopathy in the Indian subcontinent where tuberculosis is widely prevalent

CASE REPORT

A 30 year old female patient presented to our outpatients department with complaints of right sided painful cervical lymphadenopathy since 3 weeks and history of high grade fever with chills since 2 days. She gave a history of loss of appetite and loss of weight also since 3 weeks. She also complained of menorrhagia however, there was no history of cough or hemoptysis, hematuria or melena. On examination she had a 3x3cm firm mobile swelling in the right posterior triangle of the neck and 1.5x1.5cm palpable swelling in the right inguinal region. FNAC (cervical lymph node) was done on an outpatient that was suggestive of acute lymphadenitis and patient was given a course of oral antibiotics and analgesics.

On the subsequent visit she was admitted with the plan for a cervical lymph node biopsy. However on admission she developed fever with severe chills and rigor (104°F), and developed facial puffiness, malar rash and aphthous ulcers in the oral cavity. The complete blood picture showed pancytopenia (Hb 6.8 g/dl, TLC 1400, platelets 1,54000). Urine, throat and blood cultures grew no organisms. Patient was referred to dermatologists in view of the malar rash and was diagnosed with Acute lupus erythematosus (LE). Haematologist advised antinuclear (ANA) and antiphospholipid antibodies, rheumatoid factor, antineutrophil cytoplasmic antibodies (ANCA) and hepatitis B surface antigen which were all found to be negative. However, since the symptoms did not subside decision was taken to perform cervical lymph node biopsy in order to achieve a definitive diagnosis. Histopathology showed necrotizing histiocytic lymphadenitis of Kikuchi-Fujimoto type. Z-N stain for acid fast bacilli (AFB) was negative. Patient was then started on Prednisolone which was later tapered down. Patient recovered well and at a follow up visit 6 months later, the patient was doing well while still on prednisolone (25mg) with no evidence of relapse.

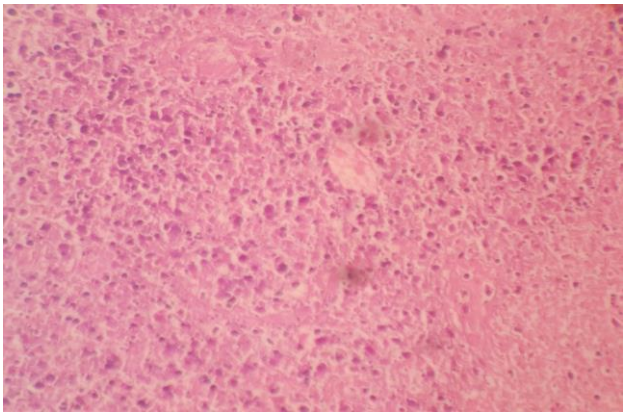

Figure 1: 40 X; H and E necrotizing histiocytic lymphadenitis of Kikuchi-Fujimoto type.

DISCUSSION

Kikuchi Fujimoto lymphadenitis was first reported in Japan in 1972.[2] It commonly occurs among young women with localised lymphadenopathy, mostly seen in the cervical region.[1] It is associated with fever and leukopenia in 50% of the cases.[3]

The differential diagnosis for enlarged cervical lymph nodes in the neck (posterior triangle) are lymphoma, tuberculosis, infectious mononucleosis, metastasis from the scalp or more distant sites, human immune deficiency virus (HIV), toxoplasmosis and rubella. In our patient there was no history of tuberculosis contact and neither history nor clinical findings suggestive of metastasis. Patient did not undergo any surgery or have viral infections in the past. Patient was investigated for viral markers which were all found to be negative.

The first cases of Kikuchi Fujimoto lymphadenitis or disease were reported in North America and Europe.[4] More commonly seen amongst the Asian population, however the disease is now found worldwide. The etiology though remains unclear with various causes postulated. Several infective agents such as EBV, parvovirus B 19 and HHV-6 have been thought to be causative although no relationship or link has been established.[5] Histological similarity seen between SLE and Kikuchi Fujimoto disease (KFD) led Dorfman and Berry to suggest that KFD could be an attenuated form of SLE.[6] Another hypothesis proposed that KFD might be a self-limiting SLE-like auto immune reaction to viral infected transformed lymphocytes.[7]

In contrast to most studies we observed a prolonged course with severe systemic and constitutional symptoms not responding to treatment with antibiotics or nonsteroidal anti-inflammatory drugs. Our patient finally was relieved of her symptoms on starting steroid therapy. The diagnosis and management of Kikuchi Fujimoto disease requires a multidisciplinary approach. However, it remains a matter of debate that steroid therapy may be needed only in those patients in whom the disease is associated with the hemophagocytic syndrome, SLE or other rheumatic disorders.[8-11] Patients with severe distressing symptoms and recurrence could benefit from corticosteroid treatment.

CONCLUSION

The aim of present case report was to highlight the diagnostic dilemma a patient with cervical lymphadenopathy can present to the clinicians in the advent of an uncommon diagnosis. Kikuchi Fujimoto lymphadenitis disease should be considered as a differential diagnosis when treating a patient with cervical lymphadenopathy in order to avoid delay in diagnosis, inappropriate treatment and prevent complications.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Panchal A, Gautam R, Mahadik A, Chougle Q, Shaffi MZ. Kikuchi Fujimoto lymphadenitis- an uncommon entity in the surgical outpatients: a case report. Int J Res Med Sci 2017;5:2243-5.