A Case Of Multiple Relapses And Remissions Of Cervical Lymphadenopathy; Kikuchi-Fujimoto Disease

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ABSTRACT

Once thought to be an east Asian disease, predominantly Japanese, cases of Kikuchi's disease have been reported from other parts of Asia, Europe and America.The major clinical finding is painless cervical lymphadenopathy, mostly unilateral and involves posterior cervical lymph nodes. Routine laboratory investigations doesn't help in diagnosis. Accurate diagnosis is established by excisional biopsy of the effected lymph node. . It is usually self-limiting and treatment is mainly symptomatic. Recurrence has been reported in some cases. Our case appears to be an interesting variant as multiple relapses and remissions were observed with are always self-limiting and resolve without any specific medication or treatment.

Key words: Cervical lymphadenopathy, Histiocytic necrotizing lymphadenitis, Kikuchi – Fujimoto disease

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Conflict of interest: No

Case report is Original: YES

Whether case report publishes any where? NO

INTRODUCTION

Kikuchi's disease was previously known as subacute necrotizing lymphadenitis. It was first described by Kikuchi1 and Fujimoto et al2 in 1972, hence sometimes called as Kikuchi-Fujimoto disease. The first case reported outside Japan was reported in 1982.3 Most reports of Kikuchi's disease have been published in the pathology literature. Very few reports have come from Pakistan and there appears to be a general lack of awareness about the condition among the surgeons.4,5 It is a self-resolving, benign cause of cervical lymphadenopathy, usually posterior cervical, and is predominantly seen in young Asian women, who are in their late 20s or the early 30s. Initially Strong female Predominance reported, but recent studies from Eastern countries suggest male to female ratio closer to 1:1.3This report describes Kikuchi's disease in a 21 years old man.

CASE REPORT

The patient reported here is a 21 years old man of normal built who presented to our out-patient department with multiple large swellings on the right side of neck in April' 2015. There were no associated symptoms as pain, fever, cough, night sweats, weight loss, anorexia, or history of any recent illness. There was history of similar swellings on the same

side of neck in the past and this was reported to be the fourth recurrence, all spaced at an interval of approximately 10 – 12 months. The Previous 3 times he neither took any proper medical advice nor was investigated, but every time due to his beliefs received some "spiritual therapy" from somebody in the village and as the swellings completely used to resolve in 8 to 10 weeks he attributed the resolution to that. This time his friend forced him to visit hospital.

On clinical examination only posterior group of right cervical lymph nodes was enlarged. The nodes were non-tender, discrete, firm in consistency and freely mobile. No other lymph node group was involved. Scalp and ENT examination was unremarkable. Chest, abdomen and genitalia examination was also normal. Baseline laboratory investigations including CRP and ESR were all in normal range. X-ray Chest and abdominal sonography did not show any pathology. CT scan of head and neck was performed and the only positive finding was multiple enlarged right posterior cervical lymph nodes ranging from 0.5 to 4 cm in diameter.

FNAC was non-conclusive so excision biopsy of a lymph node was performed and sent for histopathology which surprisingly reported that it was Kikuchi-Fujimoto disease, a very rare condition in our part of the world. Eosin and hematoxylin staining showed disrupted architecture with paracortical expansion, cellular debris and multifocal areas of necrosis. There were no prominent Neutrophils but plasma cells and numerous histiocytes were identified. Immunohistochemical staining showed a predominance of T cells with very few B cells. The findings were consistent with histiocytic necrotizing lymphadenitis i.e., Kikuchi's disease.

The patient was reassured and discharged on a short course of glucocorticoid therapy. Follow-up showed regression at 7 weeks and complete clinical resolution at 12 weeks.Follow-up after 1 year has not shown any signs of recurrence and he is now on long term follow-up for future possible recurrence.

DISCUSSION

Once thought to be an east Asian disease, predominantly Japanese, cases of Kikuchi's disease have been reported from other parts of Asia, Europe and America.

The onset is acute or sub-acute, mostly evolve during a period of 2 to 3 weeks. The major clinical finding is painless cervical lymphadenopathy, though occasionally physicians come across patients with painful and generalized lymphadenopathy. However clinical features may vary from patient to patient. Some Patient of KFD with lymphadenopathy complain of fatigue, malaise, weight loss, night sweats hepatomegaly, skin lesions, headache and rarely constitutional neurological symptoms.7,8 Lymph nodes may vary in size from 0.2cm to 4cm, mostly unilateral and involves posterior cervical lymph nodes9 as was observed in our case. Typically lymph nodes are smooth, firm, discrete and mobile. Occasionally, some patients present with generalized lymphadenopathy such as inguinal, axillary, celiac and mediastinal lymph nodes.10

Most of the time patients are diagnosed as a case of viral infection due to little lymph node swelling or pain.4 The etiology of Kikuchi's disease remains a mystery, though on the basis of clinical manifestation infectious and autoimmune etiologies have been proposed.5 The viral agents associated based on immunological tests include Human Herpes virus (HHV) 6 and 8, Epstein Bar virus (EBV) and Parvovirus B19.6 However the association with SLE has been stated with a frequency probably more than that expected by a chance alone thus patients once diagnosed with Kikuchi- Fujimoto disease should be kept on long term follow-ups for early detection of SLE.3

Routine laboratory investigations doesn't help in diagnosis except for CRP and ESR may be raised in some cases and many patients have neutropenia. About 25% to 31% of patients have atypical lymphocytes in their peripheral blood smear.11 Most of the investigations are carried out to rule out the differentials.

FNAC has limited role with overall diagnostic accuracy estimated at 56%.4,12MRI and CTare not able to distinguish Kikuchi's disease from other diseases that commonly cause lymphadenopathy, such as lymphomas, tuberculosis and tumor metastasis.13 Accurate diagnosis is established by excisional biopsy of the effected lymph node.14 Three main histological patterns seen are proliferative, necrotizing and xanthomatous. Proliferative pattern is approximately one third and has dominant inflammatory infiltrates but no neutrophils. Half cases are of necrotizing type and xanthomatous pattern is rare and has abundant foam cells.15

No specific treatment is available. Treatment is highly supportive according to sign and symptoms. It is usually self-limiting and resolves within 1 to 4 months. Prednisolone may help in advanced cases to accelerate resolution.16Corticosteroids and immunosuppressant drugs have also been advocated in resistant cases. A possible recurrence rate of 3% to 4% has been reported. 3, 17 A few deaths are also reported.18 Our case appears to be a different variant as multiple relapses and remissions were observed with are always self-limiting and resolve without any specific medication or treatment.

CONCLUSION

Doctors particularly surgeons should be aware of atypical presentations of lymphadenopathies so that they are able to think out of the box for prompt and appropriate management of such diseases which may otherwise be overlooked. Moreover lymph node biopsy is inevitable for diagnosis of these atypical conditions and one should not be reluctant to carry it out if diagnostic uncertainty exists.

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