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Case Report

Unusual cause of cervical lymphadenopathy in post-partum patient with atypical presentation: Kikuchi disease

Siti F. Abdul Razak[1]*, Fong J. K.[2], M. R. M. Yunus[3], Elsie J. Anastasius[1]

[1]Department of Otorhinolaryngology, Queen Elizabeth Hospital, Karung Berkunci, Kota Kinabalu, Sabah, Malaysia
[2]Department of Pathology, Queen Elizabeth Hospital, Karung Berkunci, Kota Kinabalu, Sabah, Malaysia
[3]Department of Otorhinolaryngology Head and Neck Surgery, UKM Medical Centre, Kuala Lumpur, Malaysia

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***Correspondence:**
Dr. Siti F. Abdul Razak,
E-mail: anax.razak@gmail.com

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ABSTRACT

Kikuchi-Fujimoto disease, also known as histiocytic necrotising lymphadenitis, is an unusual type of painful lymphadenopathy. It is a self-limiting benign disease; usual management includes supportive therapy. We described a case of Kikuchi Disease in a childbearing-age woman who presented with painless and localised cervical lymphadenopathy during her postpartum period. Histopathology of the affected cervical node reveals areas of apoptosis and necrosis with abundant foamy histiocytes and plasmacytoid monocytes. The cervical node resolves spontaneously, and no subsequent new cervical lymph node was observed in this patient. Patient was given reassurance. It is vital to recognise Kikuchi disease as one of the causes of persistent cervical lymphadenopathy, to provide optimal management and to avoid incorrect treatment.

Keywords: Lymphadenopathy, Kikuchi disease, Necrotising lymphadenitis

INTRODUCTION

The Kikuchi-Fujimoto disease is a rare benign, self-limiting condition of the lymph nodes. It was first reported in 1972 by Japanese pathologists, Kikuchi and Fujimoto et al. Since then, the disease has been reported worldwide, particularly in the Asia continent. Although Kikuchi disease was initially thought to occur predominantly in young and healthy women, it has been reported in patients with all socio-demographics of age, gender, and ethnicity and tends to occur in various parts of the body. Typically, a patient will present with painful, localised lymphadenopathy with fever. Less commonly, other non-specific symptoms might also be reported, for example, nausea, lethargy, night sweats and loss of weight. As it has similar clinical features with other forms of lymphadenopathy including lymphoma, EBV mononucleosis, autoimmune, inflammatory conditions, and infectious disease such as tuberculosis, it is often misdiagnosed, with a rate as high as 40%.[10] Hence, it is crucial to have an accurate diagnosis thus leading to correct treatment.

CASE REPORT

A childbearing age, three months postpartum woman presented with painless neck swelling for three months duration, right after the delivery of her child but only seek treatment after three months. It was progressively enlarging. The patient denied any fever, no tuberculosis symptoms such as appetite loss, weight loss, night sweats and no blood upon coughing. She also had no contact with patients with an active disease of pulmonary tuberculosis. This was her first onset of neck swelling. On examination, there was a level two to level three cervical lymphadenopathy measuring approximately four-by-four cm and a smaller lymph node at level four one-by-one cm. The masses were mobile, firm, non-tender, non-pulsatile,

non-erythematous and had no other overlying skin changes. Nasoendoscopy noted no abnormality and indirect laryngoscopy was also unremarkable. Fine needle aspiration and cytology (FNAC) was performed, reported as aggregates of epithelioid histiocytes forming granuloma surrounded by small lymphocytes, histiocytes and plasma cells in a background of necrotic granular debris. Scattered multinucleated giant cells of Langhans type were present as well, consistent with granulomatous lymphadenitis. Given the FNAC suggestive of granulomatous lymphadenitis with the need to rule out tuberculosis and fungal infection, the patient was counselled for an excisional biopsy. An excisional biopsy of the level four lymph node was performed under local anaesthesia. Histopathology report showed adipose tissue and lymphoid tissue with widespread apoptosis and areas of necrosis (Figure 1). There were abundant foamy histiocytes highlighted by CD68 immunohistochemical staining and plasmacytoid monocyte (Figure 2), highly suggestive of Kikuchi disease. Serum anti-nuclear antibody was taken, and the titre was 1:80 while complement C3 and C4 factors were negatives. The patient had no recurrent cervical swelling up until this date.

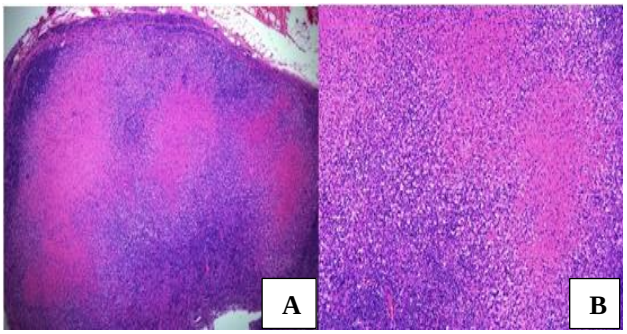

Figure 1: (A) Lymph node with paracortical areas of necrosis with widespread apoptosis and distortion of nodal architecture; and (B) coagulative necrosis with abundant karyorrhexis. Large amount of foamy histocytes are seen at the margin of the necrosis area (Haematoxylin and eosin, at 40X).

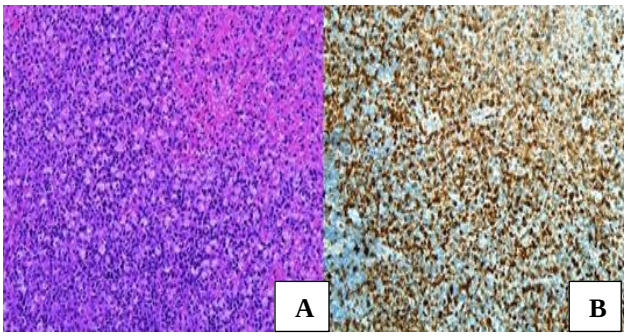

Figure 2: (A) There are abundant foamy haematocytes and monocytes admixed with karyorrhectic debris. No neutrophil is seen (Haematoxylin and eosin, at 200X); and (B) CD68 immunochemical stain highlights the abundant increased histocytes (at 200X).

DISCUSSION

Kikuchi disease, also known as histiocytic necrotising lymphadenitis, is a rare, benign, idiopathic cause of lymphadenopathy that is mostly seen in Asian women. It is reported higher in the Japanese population and East Asia, but more cases have been reported worldwide recently. It does not spare any age group; however, it is typically reported in young adults between 20 to 30 years old. There are also cases documented in pregnant women, but no reported case was seen in the postpartum period yet, making this case report the first seen in postpartum women. It was first described by Dr Kikuchi in 1972 following a case of lymphadenitis characterised by focal proliferation of reticular cells with numerous histiocytes and abundant nuclear debris. Dr Fujimoto presented similar a case within the same month, in a separate Japanese journal. The exact pathology of the disease is unknown, but it has been thought to have an infectious and immunological origin. Numerous infective agents have been reported including Epstein-Barr virus, human T-lymphotropic virus type one, parvovirus B19, human immunodeficiency virus, cytomegalovirus, hepatitis B, paramyxovirus, parainfluenza virus, rubella, Yersinia enterocolitica, toxoplasma and various fungal infections.[1,2] Systemic lupus erythematosus, Sjogren syndrome, Wegener granulomatosis, rheumatoid arthritis and Still disease are autoimmune condition that is associated with Kikuchi disease.[3]

Symptoms of Kikuchi disease may develop over a few weeks as an acute and subacute presentation with systemic B symptoms and painful unilateral posterior lymphadenopathy. Presented lymph nodes are usually smaller, between five to four mm, however, there was a reported case with seven cm lymphadenopathy.[4] Approximately, 1 to 22% of patients presented with generalised lymphadenopathy.[4] Affected lymph nodes are often described as typically painful, mobile, solitary, and non-suppurative. However, in this case, the patient presented with painless cervical lymphadenopathy, which is not a typical presentation of Kikuchi's disease. Systemic B symptoms include pyrexia presented in 35% of patients, erythematous rashes in 10% of patients and hepatosplenomegaly in 13% of patients.[4] Less common, non-specific symptoms include headache, nausea, fatigue, and arthralgia.[5]

There is no pathognomonic laboratory test for the ultimate diagnosis of Kikuchi disease. A full blood count may show leukopenia, with mildly raised erythrocyte sedimentation rate and C-reactive protein. Some literature has reported that leukopenia is observed in 25 to 58%, and leucocytosis is present in two to five per cent of Kikuchi lymphadenopathy patients.[6] Approximately, 25% of the patient has circulating atypical lymphocytes in the peripheral blood film.[4]

Although fine-needle aspiration and cytology are valuable tools for the diagnosis of certain diseases especially

tuberculosis of lymph nodes, confirmatory diagnosis of Kikuchi disease is through lymph node biopsy and histopathological analysis. A study by Das et al compared FNAC smears between patients with Kikuchi disease and reactive nodal hyperplasia, which reported overlapping cytological features seen in both conditions, therefore suggesting the limited diagnostic value of FNAC.[7]

Histopathology of the affected lymph node is characterised by partially preserved nodal architecture. The nodules are necrotic with abundant apoptotic karyorrhectic debris. The necrotic foci are either clumped together or isolated. Additional features include the presence of proliferating histiocytes, T lymphocytes (CD8), and immunoblasts with the absence of neutrophils. The minimum criteria for histopathological diagnosis are the presence of aggregated histiocytic with occasional crescent-shaped nuclei, plasmacytoid histiocytic and scattered karyorrhexis. Immunohistochemical staining will reveal histiocytes expressing CD68, myeloperoxidase CD4 markers and CD8 T-cells.[4] In a literature review by Kuo et al, Kikuchi disease can be categorised into 3 stages following histopathological features. The proliferative stage is made up of various histiocytes, plasmacytoid monocyte, and lymphoid cells containing karyorrhectic nuclear debris. Once necrosis is observed in the affected nodes, it will be categorised as a necrotising stage. It is categorised as the xanthomatous phase if foamy histocytes are predominant in the nodes.[8] The histopathology of this patient has the above criteria, making the diagnosis of Kikuchi lymphadenitis most likely.

Kikuchi disease is a self-limiting illness, resolution tends to occur within one to four months. There is no specific treatment due to its unknown aetiology. Treatment is targeted towards symptomatic relief in general with the use of antipyretics and analgesics. A corticosteroid may be given in relapsing and severe cases. It is recommended that patients receive a systemic survey as they tend to develop SLE in the future. Long-term follow-up is important as it tends to recur at a rate of three per cent.[9] Given the tendency to develop SLE, the patient was referred to the rheumatologist, for further autoimmune workup and long-term follow-up. Serum ANA was taken and showed positive with a low titre of 1:80, while ESR was not raised, and complement C3 and C4 factors were negative. Diagnosis of SLE was unlikely as the patient reported no clinical symptoms and was supported by low serum ANA titre. The patient did not complain of recurrent neck mass during subsequent follow-ups in the rheumatology as well as otorhinolaryngology clinic.

CONCLUSION

Kikuchi disease is a rare type of cervical lymphadenopathy with the exact pathophysiology remaining unknown. Although a majority of patients present with painful lymphadenopathy and pyrexia, a medical practitioner should have a high index of suspicion for Kikuchi disease in a young woman who presented with painless cervical lymphadenopathy, as seen in this case. Therefore, it is recommended to consider an autoimmune workup and to closely monitor and follow up with patients with Kikuchi disease.

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