CASE REPORT

Kikuchi-Fujimoto disease with multiple extra-nodal features - A clinical mimic

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SUMMARY

Extranodal involvement in Kikuchi's disease is uncommon. A 31-year-old previously healthy Indian woman was admitted with high grade fever, multiple joint pain and skin rash for 3 weeks. She had negative anti-nuclear antibodies and had features of Kikuchi's disease on lymph node biopsy. She also had multiple extranodal manifestations including erythematous maculopapular rash, symmetric polyarthritis and hepatosplenomegaly. Kikuchi's disease with extranodal involvement can clinically mimic diseases like hematological malignancies, connective tissue disorders and certain infections. A lymph node biopsy plays a crucial role in making an accurate diagnosis by excluding other diseases. A discussion on the importance of differentiating Kikuchi's disease from systemic lupus erythematosus is included.

Key words: *Kikuchi's disease;Extra nodal features;clinical mimic.*

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INTRODUCTION

Kikuchi's disease (KD) is an uncommon, benign, self-limited condition of unknown etiology which mainly affects young adults with a higher prevalence among Asians. It is clinically characterized by tender regional lymphadenopathy, fever, and occasional systemic involvement. Lymphomas, systemic lupus erythematosus (SLE), and certain infections have similar clinical and histopathological features, which makes it important to exclude such conditions to make a proper diagnosis of KD.

CASE REPORT

A 31-year-old previously healthy Indian woman was admitted with high grade fever, multiple joint pain and skin rash for 3 weeks. She had pain involving large and small joints of upper and lower limbs with swelling and redness. She denied history of weight loss, had no sick contacts and had no history of addictions. She had no increased frequency of micturition, dysuria, loin pain, vaginal or urethral discharge, bleeding manifestations or mucosal ulcers. Examination showed palpable posterior cervical lymph nodes which were firm in consistency, mobile, non-tender, and not matted. She had erythematous, non-scaly maculopapular rash over bilateral cheeks, forehead, anterior aspect of neck, central chest and extensor aspect of bilateral upper limbs.

Hemoglobin was 10.6 g/dL (12-15 g/dL), erythrocyte sedimentation rate 65 mm in 1 h (normally less than 20 mm/hr) and C-reactive protein was 16 mg/L (normal range 0.8 mg/L to 3.0 mg/L). Peripheral smear was normal. Urinalysis was normal. Biochemical parameters were normal. Chest radiography and electrocardiogram were normal. Etiological workup including Dengue, Leptospira, Brucella, Rickettsia, Epstein Barr virus, HIV, Hepatitis B and C serologies and antinuclear antibody was negative. The test for malarial parasite was negative and blood cultures did not reveal any growth. C3 and C4 levels were normal. Serum ferritin was 5000 microg/L (10-200 mcg/L). Ultrasonography of abdomen showed mild hepatosplenomegaly. Lymph node biopsy showed lymphoid follicle with germinal center, paracortical

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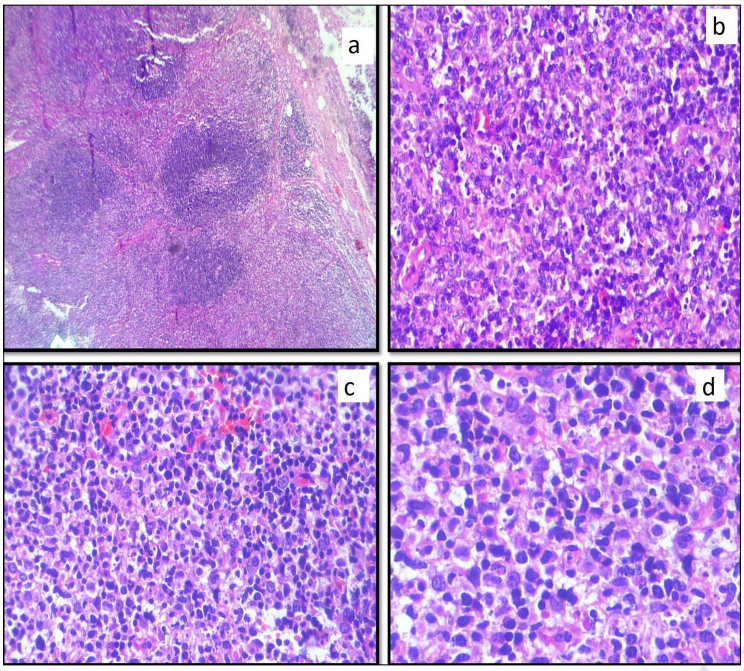

Figure 1 - Lymph node biopsy showing partially preserved architecture of lymphoid follicle with germinal center and paracortical area expansion (A), paracortical area expanded by histiocytes and many apoptotic bodies (B), crescent shaped nuclei of histiocytes (C), and patchy areas of cortical necrosis (D).

area expansion with histiocytes and many apoptotic bodies, crescentic histiocytes and patchy areas of cortical necrosis without neutrophils (Figure 1). Immunohistochemistry showed CD68 and Myeloperoxidase positive histiocytes characteristically seen in KD (Figure 2). A diagnosis of KD was made based on the investigations. She was started on oral prednisolone 1 mg/kg, which was tapered and discontinued slowly over the next 8 weeks. When reviewed after 8 weeks she was afebrile, had no joint pain, skin rash or lymphoadenopathy, and serum ferritin was normal. She was followed up for one year and did not have any further complications.

DISCUSSION AND CONCLUSIONS

KD is a rare lymphohistiocytic disorder that affects young women of Asian descent more frequently than other ethnic groups. It is an enigmatic, benign, and self-limited syndrome characterized by regional lymphadenopathy and mild fever. Differential diagnosis of fever with lymphadenopathy and skin rash include infections, lymphoma and connective tissue diseases such as SLE. Extranodal involvement in KD is uncommon. Involvement of skin, bone marrow, nervous system, eye and liver dysfunction have been reported (1, 2). Dermatological manifestations reported previously with KD include; erythematous rashes and nodules, crusted papules; scattered, indurated, erythematous lesions; erythema multiforme; and erythematous maculopapular eruptions, predominantly involving the face and upper body (3). Generalized papulopustular skin lesions and lip edema with desquamation and erosions were also reported previously (4, 5). Hepatosplenomegaly may be associated with KD (6). KD with small and large joint polyarthritis has been described previously (7).
Our patient had KD with extra nodal mani-

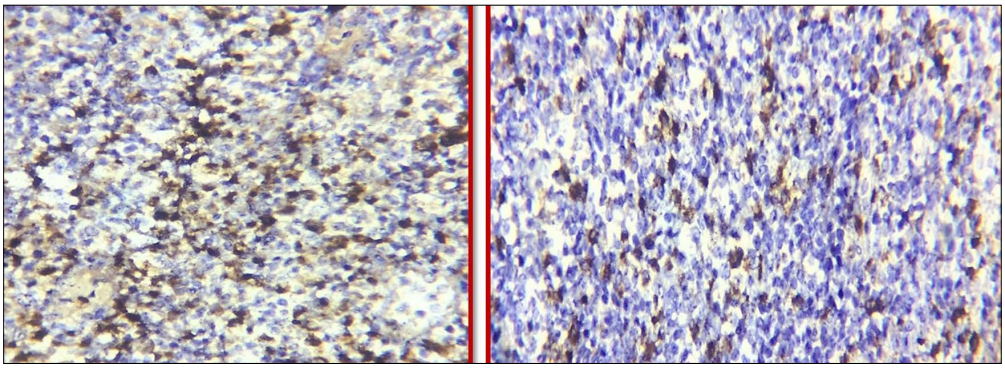

Figure 2 - Immunohistochemistry showing CD68 (A), and myeloperoxidase positive histiocytes (B).

festations including erythematous maculopapular rash, symmetric polyarthritis and hepatosplenomegaly. The presentation of KD with predominant extranodal features makes it difficult for the physician to differentiate it from other disorders such as connective tissue disorders, especially SLE, lymphomas and certain infections. Lymph node biopsy is necessary to make an accurate diagnosis excluding other clinical mimics. SLE has been seen to either coincide, precede, or follow a diagnosis of KD. Among 35 cases of SLE-associated KD reviewed by Santana et al., seven patients were diagnosed with SLE prior to KD diagnosis, fourteen patients were diagnosed with KD and SLE simultaneously, and fourteen patients were diagnosed with SLE after a diagnosis of KD (8). Our patient did not have SLE during the diagnosis of KD or the follow-up period of one year. Lymph node biopsy and immunohistiochemistry in our patient showed classic features of KD without features of SLE (increased neutrophils, eosinophils, or plasma cells, and vasculitis and hematoxylin bodies).

This case is important for the following reasons. Firstly, young Asian women presenting with clinical features in keeping with a diagnosis of systemic lupus erythematosus or lymphoma should have a lymph node biopsy in order to rule out more benign conditions such as KD. Secondly, KD has been described in association with multiple extranodal manifestations including erythematous maculopapular rash, symmetric polyarthritis and hepatosplenomegaly.

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