ORIGINAL RESEARCH PAPER
Volume - 13 | Issue - 10 | October - 2024 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

A RARE CASE REPORT OF A YOUNG ASIAN FEMALE WITH KIKUCHI FUJIMOTO DISEASE AND SYSTEMIC LUPUS ERYTHEMATOSUS

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ABSTRACT

Kikuchi Fujimoto disease (KFD) also known as histiocytic necrotizing lymphadenitis is a benign and self-limiting disease, which presents as lymphadenopathy and fever. It has been observed that KFD is associated with Systemic lupus erythematosus (SLE) and is suspected to be an unusual presentation of SLE. Here we describe a patient with a final diagnosis of KFD with SLE who had both features of KFD and SLE who recovered with supportive therapy and immunosuppressive drugs.

KEYWORDS

Kikuchi Fujimoto disease, Systemic Lupus erythematosus, aseptic meningitis, lymphadenopathy, autoimmune disease.

INTRODUCTION

KFD also known as histiocytic necrotizing lymphadenitis is a benign and self-limiting disease and patients usually present with complaints of lymphadenopathy [commonly in the cervical region] and fever. It most commonly occurs in young women. [1] Clinical manifestations also include weight loss, skin rash, arthromyalgia and aseptic meningitis. [2] Clinically KFD may mimic other diseases such as SLE, infectious lymphadenitis and malignant lymphomas and careful evaluation and histopathological diagnosis is required to differentiate between them. Etiology of KFD is uncertain, but various viral diseases such as Epstein Barr Virus, Herpes Simplex Virus, Varicella Zoster Virus, and autoimmune disorders including SLE and Sjogren's syndrome has been associated with the disease.[3][4] Diagnosis is based on Histopathological examination. Diagnosis of the disease depends on the lymph node biopsy, and histologically, 3 patterns of KFD are visualized; proliferative, necrotizing and xanthomatous.[5][4] Immunohistochemistry of KFD shows myeloperoxidase-positive histiocytes. Proper diagnosis is important as the clinical feature of this disease points to various differential diagnosis such as infectious lymphadenitis, autoimmune lymphadenopathy [SLE] and malignant lymphoma.[6]

Poor familiarity with physicians and pathologists toward this disease poses diagnostic and treatment challenges. Treatment of KFD is mainly supportive provided, other causes of similar clinical presentations are ruled out.[7] Here we are presenting the case of a patient with KFD and SLE, with clinical features such as aseptic meningitis, rash and weight loss.

Case Report

A 25-year-old female with no known comorbidities presented with headache since the past 5 months, which was acute in onset, bilateral temporal and severe in intensity associated with occasional episodes of vomiting, which was non-projectile in nature with no bile or blood staining. Headaches were associated with photophobia and phonophobia. There is history of an episode of loss of consciousness 2 weeks back with up rolling of eyes, deviation of angle of the mouth, tongue biting during episodes of unconsciousness, but no history of jerky movements of limbs or involuntary micturition or defecation. This was associated with the gradual regaining of consciousness without any residual confusion. The patient had a similar episode 1 week after the first episode. The patient had a history of fever for 2 weeks, which was high grade associated with chills and rigors, which was a continuous type.

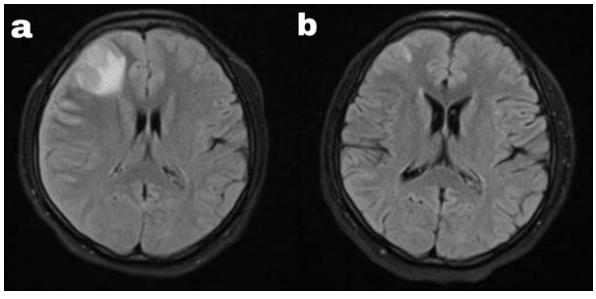
Patient gives a history of hair loss, rashes all over the body, weight loss, muscle cramps and recurrent oral ulcers for the past 6 years. Patient has had cervical lymphadenopathy for 6 years for which Fine needle aspiration cytology (FNAC) was done in previous hospital visits. FNAC was normal and the lymph node did not change in size and hence was not evaluated further.

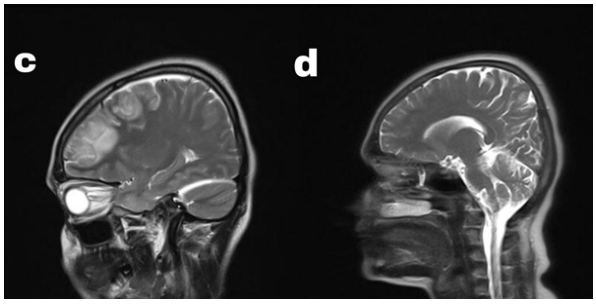
On general examination, the patient had pallor and there was an enlarged left cervical lymph node which was non-tender. Patient had normal higher mental functioning and the motor system was intact. All cranial nerves were normal on examination. Suspicion of tubercular lymphadenitis and lymphoma was made.

On investigations, the patient had bicytopenia (hemoglobin 10.8mg/dl and Total leukocyte count of 2460/mm^3). Normocytic normochromic anemia was observed. Mantoux's test was negative. Fundus examination showed early papilledema changes. C3 and C4 levels were low. Since the patient had bicytopenia and had clinical features of SLE an ANA IFA was done, which showed fine speckled cytoplasmic pattern with a titer of 1:1000. MRI brain showed T2 and FLAIR hyper-intense areas in right frontal and temporal regions with no significant post-contrast enhancement. Pachymeningeal and leptomeningeal enhancement was also noted in the right frontotemporal region suggestive of early cerebritis with meningitis. MRI Brain Spectroscopy was done which ruled out infection and Tuberculosis.

A cervical lymph node biopsy was performed. Grossly, the lymph node looked pale brown and was irregular measuring 1.2x0.5x0.4cm. The Cut section was pale to pale brown and firm. Microscopic examination showed incompletely effaced architecture composed of histiocytes, plasmacytoid cells and karyorrhectic debris. Some of the histiocytes show crescent-shaped nuclei and foamy cytoplasm. No areas of necrosis or granuloma were seen. All these features are suggestive of KFD in the proliferative phase. The absence of hematoxylin and neutrophils indicate KFD rather than SLE. A final diagnosis of KFD with SLE was made.

The patient was started treated with inj. Mannitol, inj. Ceftriaxone, inj. Dexamethasone, tab. Levetiracetam, tab. Domperidone and tab. Paracetamol and was relieved symptomatically. Repeat mri brain was done after a course of steroids and was normal.





a) MRI Brain (Axial view) showing T2 hyper-intense areas in right frontal region. b) MRI brain (Axial view) after a course of steroids showing resolution. c) MRI brain (Coronal view) showing T2 hyperintensities in right frontal and parietal regions. d) MRI Brain (Coronal view) after a course of steroids showing resolution.

DISCUSSION

KFD was first reported in Japan in 1972 and is characterized by lymphadenopathy (mostly involving the cervical lymph nodes) and fever[1]. Other symptoms of the disease include weight loss, skin rash, arthromyalgia and aseptic meningitis.[2]

The differential diagnosis for the presenting complaints are broad and includes diseases such as tubercular lymphadenitis, lymphoma etc. all of which have different treatment modalities and were initially suspected in our patient. Understanding the possibility of KFD via histopathological features becomes necessary as no effective treatment has been established and the treatment given is conservative[1,7]. Lupus lymphadenopathy can mimic KFD in morphology and should be considered in patients to prevent delay in treatment[3].

Treatment is non-specific and often managed with analgesics and antipyretics. Corticosteroids are given for relapsing and severe cases and those with aseptic meningitis.[6] The disease is usually self-limiting, and most patients recover with a short dose of steroids. KFD with SLE can have a more aggressive course and treatment is recommended to prevent relapse.

Autoimmune manifestations have been noted in KFD. KFD may precede, follow or coincide with the diagnosis of SLE. KFD is suspected to be an unusual presentation of SLE. Sopeña et al has reported an incidence of 23% SLE in 20 cases with KFD.[7] Kucukardali et al has reported that SLE-associated KFD is more common in Asian patients than European patients.[8]

Patients with KFD with other features suggestive of SLE should be screened for SLE.

Our patient had other features associated with autoimmune conditions and positive SLICC criteria and a diagnosis of SLE with KFD was made. Differentiating between KFD and SLE is challenging and is mainly based on histopathological evidence. Our patient was also found to have decreased levels of C3 and C4. Low C4 is associated with the development of autoimmune disorders such as SLE.[9] The patient had cervical lymphadenopathy for 6 years and was not evaluated for KFD as FNAC was negative and the enlarged node showed no morphological changes.

CONCLUSION

Many of the symptoms, patient characteristics and histopathological features of KFD are similar to other autoimmune diseases, particularly SLE and differentiating between the two is challenging and is mainly based on histopathological evidence. We present this case to highlight the occurrence of SLE and KFD together and the need for long-term follow-up and screening of patients with KFD for SLE.

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