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An Atypical Presentation of Kikuchi-Fujimoto Disease Illustrating Overlapping Features of Systemic Lupus Erythematosus: A Case Report

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Abstract

Purpose: To report a case on atypical presentation of kikuchi-Fujimoto disease (KFD) that mimics multiple features with SLE.

Case Report: A 19 year old female patient with an uncommon presentation of kikuchifujimoto disease (KFD) is described.the patient was presented with tacypnoea,fever, abdominal pain and weight loss.

Keywords: Kikuchi-Fujimoto, Systemic lupus erythematosus (SLE), India

Introduction

Kikuchi-Fujimoto Disease (KFD) is known to occur both in the juvenile and adult population. The first case of reported Kikuchi Fujimoto disease was in Japan in 1972 and since then this disease has been described worldwide, with most cases reported in Asia [1, 2] Due to its low incidence rate and non-specific clinical features, KFD has not been well understood. Although unidentified infectious agents and an autoimmune response have been considered as the prime causes, the KFD pathogenesis remains unclear Typically, this clinicopathologic disease is a benign and self-limited condition. It is known to be uncommon in the United States, with most cases occurring in Asia, although there have been cases described worldwide in the literature. There is a higher incidence of KFD in women, aged 20 to 35 years. Because of its shared clinical features, the disease can be easily mistaken for other forms of lymphadenitis with a misdiagnosis rate up to 40% [5] Generally, KFD is diagnosed via excisional lymph node biopsy and histopathological analysis. KFD shares many characteristics with other causes of lymphadenopathy including lymphoma, inflammatory disorders, autoimmune conditions, and infectious causes of lymphadenopathy like tuberculosis infection; therefore, it is important consider KFD in cases of persistent lymphadenopathy and must be differentiated from these conditions (1). We also performed a literature review of KFD to provide an evidence-based understanding of this disease.

Case Report

A 19-year-old female was admitted to the hospital because she had been breathing difficulty since one week worsened yesterday, H/O fever and right abdominal pain since one week along with breathing difficulty patient diagnosed as SLE, Psoriasis, myocarditis with moderate biventricular dysfunction since 6 months symptoms started.

The patient's vital signs were a respiratory rate of 34 breaths/ min, temperature of 98.6°F, blood pressure of 120/80 mmHg, and a heart rate of 80 beats/min. Physical examination revealed multiple soft, tender masses on the right side of her neck, which measured approximately 2 cm in diameter; no changes were evident in the skin overlying them.The patient's oral cavity, ears, nose, and throat, and thyroid gland were examined first because acute cervical lymphadenopathy in those areas is common with an infection, but no lesion was found. Cardiovascular, neurologic, and respiratory testing was normal. The patient's abdomen was soft with normal bowel sounds.

Results from laboratory tests (complete blood count, creatinine, electrolytes, C-reactive protein, and erythrocyte sedimentation rate, creatinine, sodium). Here Hb, sodium BUN creatinine was depleted and LDH, Potassium, CRP, ESR was also elevated. Anti nuclear anti body shuld be positive, ANA Positive(3+)in 1:100 dilution nuclear granular pattern is seen in antibody against Ku, Nrnp/Sm, SS-A, SS-B, CENP ds DNA, Nucleosomes, histones, spindle fibre and DFS70.

We then performed noninvasive imaging, including whole-body magnetic resonance imaging (MRI) and a thoracic-abdominal computed tomography CT scan mild bacterial pneumothorax noted, Loculated pleural fluid collection with air fluid levels in the left inferior hemithorax suggestive of hydropenumothorax, mild peri cardial effusion, peri bronchial and sub plural chronic infiltrates noted in the left lower and upper lung lobe milder changes are also noted in the right middle lung lobes. She was treated with inj. Meropenem 1g iv, Inj:Azithromycin 500mg iv, Inj:pantoprazole 40mg iv, T. Prednisolone 20 mg and other supportive measures was given.

Discussion

KFD or histiocytic necrotizing lymphadenitis is an unprecedented cause of lymphadenopathy [1]. It for the most part happens in patients in Asia and seldom within the United States [1]. The precise frequency of KFD remains vague since this infection is effectively mixed up for other causes of lymphadenopathy. A survey by Kucukardali *et al.* [6] in 2007 recognized 330 cases of KFD since 1991. A report by Feder *et al.* [7] in 2014 revealed that 10 cases had been detailed within the Joined together States. The etiology and pathogenesis of KFD stay vague; be that as it may, irresistible specialists and an immune system reaction are considered the 2 primary causes [8]. Contaminations that have been proposed as conceivable etiologic operators of KFD incorporate EBV, varicella-zoster infection, human herpesviruses 6, 7, and 8, parvovirus B19, paramyxovirus, parainfluenza infection, rubella, and human T-lymphotropic infection sort 1 [4]. An affiliation moreover has been found between immune system conditions and KFD, particularly SLE [1].

Clinically, the onset of KFD can be intense or subacute and it as a rule settle suddenly inside a few weeks [9]. Lymphadenopathy is watched in 100% of patients; in up to 90% of cases, it is back cervical, and in less than 22% of cases it is generalized [4, 5]. Lymph hubs extend in measure from 0.5 cm to 4.0 cm [1]. Fever and delicate and difficult lymph hubs too are common in patients with KFD [1]. Up to 90% of them display with fever [4] extending from 38.6Â°C to 40.5Â°C, which keeps going from 1 week to 7 weeks [1]. Delicate lymph hubs are show in 82% of patients with KFD [4]. Other side effects incorporate weight misfortune, sickness, heaving, shortcoming, migraine, night sweats, and upper respiratory indications [9]. The skin is the foremost commonly influenced organ aside from lymph hubs [5]. Patients with KFD can have papules, facial palmar erythema, plaques, or knobs [1].

Comes about of research facility testing for KFD are non-specific, counting discoveries of iron deficiency, leukopenia, leukocytosis, thrombocytopenia, and rises in liver chemicals, lactate-dehydrogenase levels, and erythrocyte sedimentation rate [4, 10]. On imaging, the lymph hubs in patients with KFD have an unpredictable external edge and they are less circular than is seen with lymphoma [5, 11]. Ultrasound uncovers a hypoechoic center with a hyperechoic edge [11]. CT appears homogeneous improvement and no critical rot [10].

KFD is analyzed based on histopathologic discoveries gotten from fine-needle goal or biopsy of the influenced lymph hub after other illnesses have been avoided [4]. Histologic discoveries incorporate protected nodal engineering, shifting degrees of coagulative rot within the paracortical locales with copious karyorrhectic flotsam and jetsam, and an nonappearance of eosinophils or neutrophils [10]. Crescent-shaped histiocytes and plasma-cytoid monocytes are regularly seen encompassing necrotic ranges [5]. In patients with CKD, histiocytes express CD68, myeloperoxidase, and CD4 [5].

Differential determination of lymphadenopathy incorporates irresistible specialists, such as TB, toxoplasmosis, Bartonella henselae, HIV, and EBV; irritation; SLE; lymphoma; and metastasis [1]. Serologic testing can propose other causes of lymphadenopathy, such as cat-scratch infection or toxoplasmosis, and histological comes about can be utilized to prohibit cancer [1].

Since KFD is self-limiting and ordinarily settle inside a couple of months, perception is the foremost common approach to administration [5, 8, 9]. NSAIDs and corticosteroids are utilized to treat patients who have indications or extranodal lymphoid organ inclusion [5, 8]. No rules exist for dosing and term of corticosteroids in administration of KFD [1, 5]. In 4% of patients, KFD repeats at up to 8 a long time after their introductory introduction [5]. Therefore, long-term follow-up plays an important role in assessing the recurrence rate.

Conclusions

The clinical introduction is variable and it can mirror other infections. The imaging highlights for KFD are non-specific. The conclusion ought to be considered in youthful ladies who have cervical lymphadenopathy for a brief time. Long-term follow-up is essential to survey these patients for improvement of immune system malady or a repeat of KFD. In spite of the fact that a uncommon malady, KFD ought to be kept in intellect within the differential conclusion of cervical lymphadenopathy. Due to the striking likeness within the bundle of non-specific signs and indications and introductions covering with a few conditions such as TB, SLE, and lymphoma, a thorough workup and strongly examinations are ordered to distinguish the over conditions from KFD. The conclusion is affirmed by lymph hub biopsy and immunohistochemistry. A long-term follow-up is required in patients with KFD to discover determination and location of repeat.

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