ORIGINAL RESEARCH PAPER
Volume - 13 | Issue - 08 | August - 2024 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

A RARE CASE OF KIKUCHI-FUJIMOTO DISEASE IN 10 YEARS BOY AT DEPARTMENT OF PEDIATRICS AND NEONATOLOGY, ETERNAL HOSPITAL, JAIPUR

Pediatrics

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ABSTRACT

Kikuchi-Fujimoto Disease (KFD), also known as histiocytic necrotizing lymphadenitis, is a rare cause of cervical lymphadenopathy. Patients usually present with localized lymphadenopathy, fever and fatigue. Because of the poorly understood etiology, it can be mistaken for an infectious disease or even malignance. Here we discuss a case of KFD that initially presented with left sided cervical lymphadenopathy that later progressed to left supraclavicular lymph nodes. Due to its characteristic overlap with other disorders like tuberculous lymphadenitis and lymphoma, KFD remains an arduous diagnosis for physicians. Therefore, one should be made aware of symptoms that can lead to misdiagnosis in patients.

KEYWORDS

Lymphadenitis, Cervical lymphadenopathy, Tuberculosis, Lymphoma

INTRODUCTION

Kikuchi-Fujimoto disease (KFD), also known as histiocytic necrotizing lymphadenopathy, is a rare, benign (non-cancerous, non-malignant) disorder of the lymph nodes, predominantly of young adults and school aged children. While slightly more common in females, it is also seen in males. This disorder presents with similar signs and symptoms as lymphoma, including enlarged lymph nodes (lymphadenopathy). The exact cause is not known. KFD shows up as painful swelling of lymph nodes, often in the neck, along with general symptoms like fever and fatigue. (1)

CASE-REPORT

Here we are presenting a case report of 9 year 11 months old boy presented with complaints of neck swelling in left submandibular region for last 1 month, which was gradual in onset and associated with intermittent pain. He also had complaint of high-grade fever for last 1 day, associated with chills not responding to oral medications. On examination HR-104/ minute, rhythmic, regular, without radio-radial or radio-femoral delay, RR-20 per minute regular and rhythmic, BP-102/74 mmHg, Temp-104°F, SPO2-96%. On local examination of neck left submandibular matted lymphadenopathy, soft, mobile, tender, measuring approx. 2.5 cm X 3 cm, warm to touch. Systemic examination- Per abdomen- Hepatic tenderness, Spleen Tip Palpable, Epigastric tenderness present. On investigations he was diagnosed as Dengue fever with warning signs and treated as per protocol with IV fluids, IV antibiotics and supportive management. USG neck was done before admission at some other center which showed necrotizing granulomatous matted lymph node for which he underwent FNAC at that center which revealed non necrotizing granulomatous inflammatory pathology with hemorrhagic change. Here at our hospital due to Dengue fever and thrombocytopenia we avoided invasive test for Lymphadenopathy. So we waited till he recovered from dengue. During the treatment of Dengue fever his swelling appears to reduce in size so we repeated USG neck after recovery from dengue fever which showed similar findings and same size of swelling. So we decided to go for excision biopsy in order to ruled out malignant and other causes of disease. In our work up for tuberculosis, CBNAAT, TB QuantiFERON- were negative. Biopsy showed typical findings of Kikuchi-Fujimoto disease. Post excision biopsy he is doing well and there is no recurrence after 3 months of follow-up.

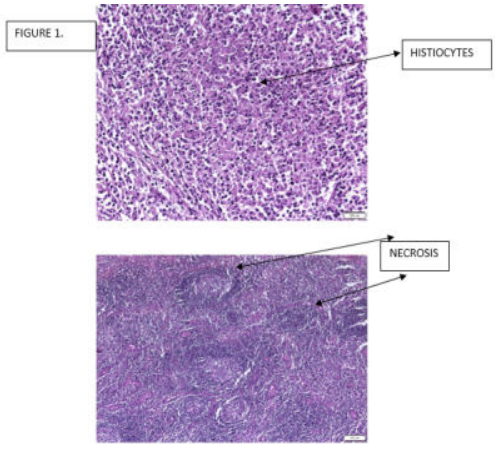
DISCUSSION

The actual cause of KFD is still unknown but it has been proposed to have infectious and immunological etiologies (2) . This disease is thought to be a hyperimmune response to infectious, physical or chemical agents. Some of the unidentified agents may include toxoplasmosis, *Brucella*, *Bartonella henslae*, *Yersinia enterolitica*, human herpes virus, Epstein-Barr virus, parainfluenza, paramyxovirus, parvovirus B19, cytomegalovirus and human immunodeficiency virus (2-5) . However, serological and molecular studies have been unable to identify a single specific pathogen. Due to this reason, KFD diagnosis is markedly limited to invasive procedures like excisional biopsy (to observe cellular changes) and not just physical examination and history.

Prevalence of Kikuchi disease has been seen highest amongst the Japanese population and people from East Asia but more recently this disease has been reported all over the world (6) Our case is from Department of Pediatrics, Eternal Hospital, Jaipur, Rajasthan.

Typically young adults (aged 20–30) are affected, but it does not seem to spare any age group as cases have been reported in the pediatric population as well, which can be seen in the reports of Byun JH (7) . However, the case report by Byun JH shows that, when Kikuchi disease occurs in children, it often involves the central nervous system leading to meningitis and encephalitis.

As reported by Deaver *et al.* (8), clinical course of this disease has some specific and non-specific features with the specific one being unilateral cervical lymphadenopathy. Although lymphadenopathy is commonly found in cervical lymph nodes other groups such as the axillary and mediastinal lymph nodes may also be involved. Unexplained fever and night sweats are also among the common clinical presentations (9) Our patient also presented with all the above listed common complaints. Less common complaints include headache, fatigue, arthralgia, myalgia, night sweats, weight loss, rash and abdominal pain (9). Our patient experienced none of them except fatigue. Although rare, patient may present with the involvement of central nervous system and peripheral nervous system (10).



Confirmation of diagnosis is done by lymph node biopsy and histopathological analysis which shows distorted nodal architecture. The nodules are mostly necrotic and have debris from nuclear fragmentations due to cellular apoptosis. These necrotic foci are either isolated or clumped together. In addition, there is presence of proliferating histiocytic, T lymphocytes (CD8) and immunoblasts (8).

The minimum criteria for KFD diagnosis is presence of aggregated histiocytic with occasional crescent-shaped nuclei, plasmacytoid histiocytic, and scattered karyorrhexis (8) . The biopsy results of our patient were quite similar, making KFD our primary diagnosis. Due to similar clinical characteristics, KFD is often mistaken for lymphoma, tuberculosis, systemic lupus erythematosus and even metastatic adenocarcinoma. Therefore, any physician who comes across a case of lymphadenopathy, should keep KFD in mind when consider differential diagnoses. KFD is self-limiting and resolution occurs is one to four months. There are no specific drugs for KFD and usual treatment is symptomatic, consisting of antipyretics and analgesics.

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