Kikuchi–Fujimoto disease in an adolescent girl with discoid skin lesion and high serum ferritin - A rare case report from Odisha

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ABSTRACT

Kikuchi–Fujimoto disease is a self-limiting disease which frequently appears with fever and lymphadenopathy, thus creating the need for differential diagnosis of persistent febrile lymphadenopathy. Although the cause of Kikuchi’s disease is unknown, some viral or bacterial infections or immunological conditions are attributed to it. It has occasionally been misdiagnosed as lymphoma or tubercular lymphadenitis; hence, clinicians should be made aware of this disease. We report a case of a 14-year-old female child who was presented with fever, rashes, multiple oral ulcers, and lymphadenopathy. The child was diagnosed as Kikuchi disease, on the basis of lymph node histopathology. There was a significant improvement of the clinical picture with oral prednisolone therapy for 2 weeks and tapering over the next 2 weeks.

Key words: Discoid skin lesion, Kikuchi–Fujimoto disease, Lymphadenopathy, Odisha, Serum ferritin

Kikuchi–Fujimoto disease (KFD) is a rare usually self-limiting disease that was originally reported from Japan. Cases are now described in all ethnic groups and all over the world and more so in the Asian population [1]. The disease most commonly occurs in people younger than 40 years of age with female preponderance. The characteristic clinical presentation of the disorder includes cervical lymphadenopathy and prolonged fever, but the presentation may be variable [2]. Its natural course is usually benign, and the clinical symptoms and signs disappear within a few months without any specific treatment. To establish the diagnosis of KFD histopathological examination of lymphnode is gold standard [3]. Here, we present a case of a 14-year-old female who was admitted to the hospital with complaints of fever.

CASE REPORT

A 14-year-old female child hailing from Kalarahanga, Bhubaneswar, presented with chief complaints of fever for 10 days. The child was a known case of seizure disorder on antiepileptic drugs. There was no history of weight loss, skin rash, arthritis, tuberculosis, or contact with tuberculosis. Although the child was on treatment as an outpatient case, the failure of improvement in her general condition and appearance of rashes needed hospitalization to evaluate the cause.

At the time of admission, her vitals were pulse rate - 100/min, blood pressure - 100/70 mmHg on the right upper arm, body temperature - 100°F, respiratory rate - 26/min, and SpO2 - 99% in room air. There was no pallor, icterus, cyanosis, clubbing, or edema. Head-to-toe examination revealed bilateral posterior cervical lymphadenopathy which was non-tender, and no palpable lymph nodes were seen in other parts of the body. Discoid skin lesions were present over ear pinnae (Fig. 1) and on the back side of the neck. Multiple oral ulcers were present over buccal mucosa, gums, tongue, and hard palate (Fig. 2). Erythematous rashes (maculopapular) were present over the face and body (neck, shoulder, chest, and back) (Fig. 3).

On routine investigations, it was found that total leukocyte count was 3000/cmm, neutrophils - 43%, lymphocytes - 54%, monocytes - 3%, hemoglobin - 10.1 gm/dl, total platelet count - 1.94 lacs/cmm, C-reactive protein - 7.1µg/dl, erythrocyte sedimentation rate - 10 mm at 1st h, procalcitonin - 0.47ng/ml, scrub typhus IgM was negative, routine microscopy of urine and culture sensitivity were normal, and blood culture sensitivity did not detect the growth of any organism. Comment on peripheral smear showed normocytic normochromic anemia with leukopenia, malaria parasite detection by quantitative buffy coat was negative, and ultrasonography of the abdomen and pelvis revealed no abnormality, and septic focus was excluded.

Biochemical parameters such as serum triglyceride - 121 mg/dl, serum ferritin - >2000 ng/ml, and serum complements C₃ and C₄ were within normal limit; antinuclear antibody and anti-dsDNA were negative; blood urea creatinine ratio and serum electrolytes were normal; and liver function test was under normal range except raised aspartate aminotransferase (AST) 66 units/L. The test for
On fine-needle aspiration cytology of cervical lymph node, acid-fast bacteria Gram-negative stains and necrotizing lymphadenitis were found with a possibility of KFD. Histopathological study of cervical lymph node revealed erosion of the lymphoid tissue, well-defined patchy foci of necrosis, and karyorrhectic and apoptotic debris with histiocytes, confirming the KFD (Table 1).

**DISCUSSION**

KFD, also known as histiocytic necrotizing lymphadenitis, is an uncommon, idiopathic, generally self-limited cause of lymphadenitis. The etiology remains unknown although an immune triggered mechanism by viral infections such as EBV, human T-cell leukemia virus type 1, parvovirus, CMV, and parainfluenza virus has been suspected [4]. KFD is typically reported to resolve within several months and with a recurrence rate of just 3–4% [5]. In this report, the patient was normal after treatment with prednisolone and was found to be healthy at 6 months of follow-up after her first manifestation. We are hypothesizing that the autoimmune mechanism may be the etiology due to its response to prednisolone treatment.

The diagnostic approach should be a collection of detailed medical history and a thorough physical examination. It is pertinent to take all the symptoms (such as prolonged fever, rashes, weight loss, night sweats, and fatigue); in consideration, their onset and duration, any insect bites, history of travel, exposure to animals, treatments (such as antibiotics), and response to them should be taken into account. To evaluate the cause, further workup for possible malignancy or chronic inflammatory condition is needed [6].

Extranodal involvement in Kikuchi’s disease is rare and has been documented in the skin, bone marrow, myocardium, and central nervous system. Cutaneous manifestations, mostly non-specific and variable in nature, have been reported in 16-40% of patients with Kikuchi’s disease [5]. In our case, maculopapular rashes on the face and body and discoid skin lesions on ear lobule and at the back of the neck were found. Blood markers for virus infections causing lymphadenitis (CMV, hepatitis B virus, herpes virus, HIV, and adenovirus) and bacterial infections (Toxoplasma gondii, Bartonella henselae, and Borrelia burgdorferi) are required [7]. Tuberculin skin test and interferon-gamma release assay should be done to exclude tuberculosis.

Raised serum ferritin in the patient of this case indicates ongoing inflammatory/autoimmune process. Although KFD with increase serum ferritin cases is rare in literature, one case of KFD with multisystemic involvement from Singapore and one case from India were reported [8,9].

The non-specific presentation of KFD requires numerous investigations, but excisional lymph node biopsy is needed to establish the diagnosis. Characteristic histopathologic features include irregular paracortical areas of coagulative necrosis, with abundant karyorrhectic debris which can distort the nodal architecture and a large number of histiocytes at the margin of the necrotic area. Karyorrhectic foci are formed by different cellular
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KFD is often misdiagnosed as tubercular lymphadenitis and started on antitubercular treatment. As it is a self-limiting disease, the patient gets cured in a few weeks, and it may be misinterpreted as a response to antitubercular medicines [10,11]. On the other hand, systemic lupus erythematosus (SLE) has developed in some patients who were thought to have KFD. The autoimmune origin has been suggested due to a significant number of cases in which SLE is diagnosed previously (30%), simultaneously (47%), or after KFD (23%) [12]. The patient will continue to be monitored closely for the next few years for any evolving symptoms of SLE or any other autoimmune disorder [11]. We feel that KFD is an iceberg phenomenon with many cases in the community which is misdiagnosed or not reported in the literature.

Conservative therapy with antipyretics is the mainstay of treatment, but oral corticosteroid therapy may be used in severe condition of disease or with cutaneous manifestation. The usual dose of steroid is 1 mg/kg/day for 5 days and tapering over the next 10 days. It is noticed that the use of steroid causes quick symptomatic relief and decrease the chance of relapse [13]. The severity of symptoms might justify high doses of methylprednisolone or use of intravenous immunoglobulin (0.4 g/kg, 2 days) [14]. Immunohistochemical analysis to know the type of T-lymphocytes, i.e., predominance of CD8+cells/CD68 is of significant value for definite diagnosis of KFD although it could not be performed due to non-availability of the facility. However, histopathological findings were characteristic of KFD in our case.

CONCLUSION

KFD should be suspected in young pediatric patients with prolonged fever and cervical lymphadenopathy, and they should be followed up for years for the development of recurrence. In our case, there is an extranodal manifestation in the form of skin rashes and oral ulcers were found along with high serum ferritin. More awareness among the pediatricians is required to suspect, diagnose, and report this disease.

REFERENCES


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