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Case Series

Clinicopathological profile of Kikuchi-Fujimoto disease in children: an 18-child case series from a tertiary care center in South India and a reminder of its mimicry of tuberculosis

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ABSTRACT

Kikuchi-Fujimoto disease (KFD), or histiocytic necrotizing lymphadenitis, is a rare, self-limiting cause of fever and cervical lymphadenopathy in children that is frequently mistaken for tuberculosis, lymphoma or lupus lymphadenitis, and pediatric data from India are scarce. We describe the clinical, laboratory and histopathological profile, treatment and outcome of 18 children (mean age 10.7±2.8 years; male:female 8:10) with biopsy-proven KFD managed at a tertiary care center in South India between January 2020 and January 2026. All had cervical lymphadenopathy (bilateral in 55.6%) and fever occurred in 88.9%. Leukopenia was present in 83.3%, elevated ESR in 94.4%, and lactate dehydrogenase (LDH), tested in all 18, was raised in every child (median 822 U/l); antinuclear antibody, also tested in all 18, was positive in 6 (33.3%). Every lymph-node biopsy showed histiocytic necrotizing lymphadenitis, with negative GeneXpert and no malignancy, excluding tuberculosis and lymphoma. Three children (16.7%) had systemic lupus erythematosus and one additionally developed acute disseminated encephalomyelitis. All received NSAIDs and 4 (22.2%) required corticosteroids; none recurred. KFD should be suspected in a well-appearing child with prolonged febrile cervical lymphadenopathy, leukopenia and markedly raised ESR and LDH; early excisional biopsy rather than fine-needle aspiration is confirmatory and excludes tuberculosis, and long-term follow-up for systemic lupus erythematosus is advised.

Keywords: Kikuchi-Fujimoto disease, Histiocytic necrotizing lymphadenitis, Lactate dehydrogenase, Lymphadenopathy, Systemic lupus erythematosus

INTRODUCTION

Kikuchi-Fujimoto disease (KFD), or histiocytic necrotizing lymphadenitis, is a rare, benign and usually self-limiting disorder of fever and regional, predominantly cervical, lymphadenopathy of unknown etiology, first described in 1972.[1,2] Although reported worldwide, it has a predilection for Asian populations and is uncommon in children.[3,4] Because it has no pathognomonic clinical sign, KFD is frequently confused with tuberculous lymphadenitis, lymphoma and lupus lymphadenitis, and children often undergo extensive evaluation for pyrexia of unknown origin before diagnosis.[5] A definitive diagnosis rests on lymph-node histopathology.[6] Recognising KFD matters because it mimics serious disease, can recur, and may coexist with

or herald systemic lupus erythematosus (SLE).[7] Pediatric data from the Indian subcontinent are limited, and we therefore reviewed our experience to describe the clinical, laboratory and histopathological profile, treatment and outcome of children with biopsy-proven KFD at our center.

CASE SERIES

This case series included all children aged 18 years or younger with a biopsy-proven histopathological diagnosis of KFD on excisional lymph-node biopsy over a 6-year period (January 2020 to January 2026); the diagnosis rested on paracortical necrotic foci with karyorrhexis, plasmacytoid monocytes and histiocytes, without neutrophils or plasma cells.[6] Clinical features, laboratory investigations, histopathology, treatment and follow-up were retrieved from medical records. Lactate dehydrogenase (LDH) and antinuclear antibody (ANA) were tested in all 18 children; other investigations were performed as clinically indicated. Anemia was defined as hemoglobin <11 g/dl, leukopenia as a leukocyte count <4000/mm^3, elevated ESR as >15 mm/h and elevated LDH as >200 U/l (local laboratory reference ranges). Categorical variables are summarized as frequencies and percentages and continuous variables as mean±standard deviation or median with range. The study was approved by the institutional ethics committee (Ref: KKCTH-CTMRF: IEC-159/June 2024; IRB Min. dt. 20.06.2024); the requirement for individual consent was waived for anonymized, retrospective records.

Eighteen children with biopsy-proven KFD were included (mean age 10.7±2.8 years, range 6-18 years; 8 males, 10 females). All 18 (100%) had cervical lymphadenopathy, bilateral in 10 (55.6%) and unilateral in 8 (44.4%); nodes were tender in 11 (61.1%), firm in 9 (50.0%) and matted in 6 (33.3%); additional nodal sites were axillary in 3 and inguinal in 2. Hepatomegaly was present in 7 (38.9%) and splenomegaly in 3 (16.7%). Fever was the commonest symptom (16, 88.9%), followed by fatigue, weight loss and headache (5 each, 27.8%) and myalgia (3, 16.7%). The median illness duration at presentation was 4 weeks (range 2-12).

Leukopenia was the most consistent laboratory abnormality (15, 83.3%), with none showing leukocytosis; anemia occurred in 10 (55.6%) and thrombocytopenia in 5 (27.8%), with a bland peripheral smear in all. ESR was elevated in 17 (94.4%), and LDH, tested in all 18, was raised in every child (100%; median 822 U/L, range 465-3188); CRP was positive in 8 (44.4%). Transaminases were mildly elevated in a minority (AST >40 U/l in 7, ALT >40 U/l in 4) and proteinuria was detected in 8 (44.4%). ANA, also tested in all 18, was positive in 6 (33.3%) and the direct Coombs test in 6 (33.3%), whereas anti-dsDNA positivity and low complement were uncommon. Lymph-node histopathology showed histiocytic necrotizing lymphadenitis in every case; GeneXpert MTB/RIF was negative in all, with no granuloma or malignancy, excluding tuberculosis and lymphoma. Three children (16.7%) had associated SLE, one of whom also developed acute disseminated encephalomyelitis. All received NSAIDs and 4 (22.2%) required corticosteroids; no recurrence was documented (Table 1 and Figure 1).

Table 1: Clinical, laboratory and outcome profile of 18 children with KFD.

Parameters	N (%)
Mean age (years) / range	10.7±2.8 / 6-18
Male : female	8:10
Cervical lymphadenopathy	18 (100)
Bilateral/unilateral	10 (55.6) / 8 (44.4)
Tender/firm/matted nodes	11/9/6
Fever	16 (88.9)
Fatigue/weight loss/headache	5 each (27.8)
Anemia (Hb <11 g/dl)	10 (55.6)
Leukopenia (<4000/mm^3)	15 (83.3)
Thrombocytopenia (<1.5x10^5/mm^3)	5 (27.8)
Elevated ESR (>15 mm/h)	17 (94.4)
CRP positive	8 (44.4)
Elevated LDH (>200 U/l), n=18	18 (100)
ANA positive, n=18	6 (33.3)
Direct Coombs positive, n=18	6 (33.3)
Histiocytic necrotizing lymphadenitis	18 (100)
GeneXpert MTB/RIF	Negative in all
Associated SLE	3 (16.7)
NSAID/corticosteroid	18 (100)/4 (22.2)
Recurrence	0

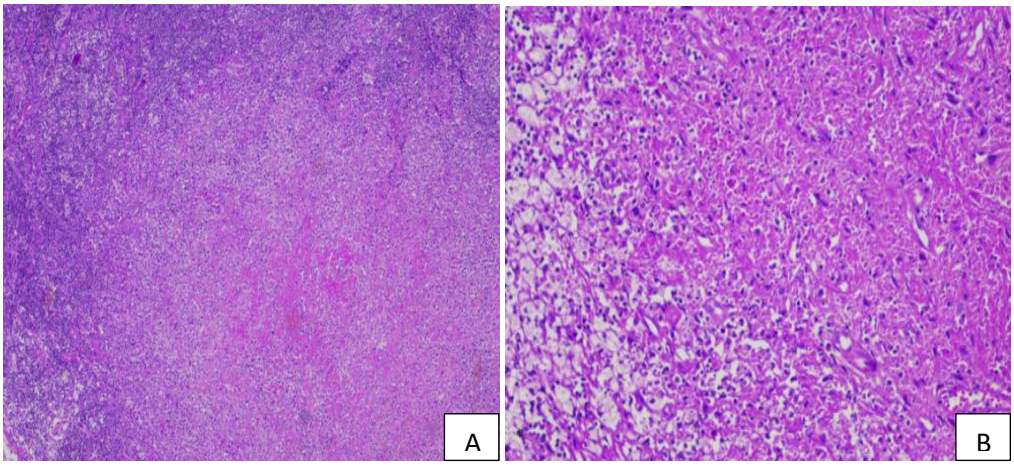


Figure 1: Lymph-node histopathology (hematoxylin and eosin); (A) H&E, x100: pale areas of geographic necrosis surrounded by phagocytic histiocytes; (B) H&E, x400: necrotic areas with karyorrhectic debris surrounded by histiocytes with crescentic nuclei; no intact neutrophils seen.

DISCUSSION

This series of 18 children adds to the limited pediatric KFD literature from South Asia. As elsewhere, febrile cervical lymphadenopathy dominated and the course was benign and self-limiting.[3,4] The mean age of 10.7 years matches other pediatric cohorts.8 In contrast to the classical adult female predominance and to some pediatric series reporting a male excess, our cohort showed a near-equal sex ratio (male: female: 8:10); such variability across age and sex is well recognized, including in large adult series and in analyzes of KFD stratified by age.[9,10] Our findings are also consistent with Indian reports.[11] All children had cervical involvement, more often bilateral, with tenderness and matting in a substantial proportion features that overlap with tuberculous lymphadenitis and reinforce the need for histopathological confirmation.

The laboratory signature was diagnostically useful: leukopenia in 83%, elevated ESR in 94% and raised LDH in every child, with a consistently bland peripheral smear. A markedly elevated LDH in a well-appearing child with prolonged febrile cervical lymphadenopathy should raise suspicion of KFD and prompt early biopsy rather than prolonged empirical antimicrobial therapy; leukopenia is likewise the commonest cytopenia reported in large pediatric series.[5,12]

The most important practical message is the mimicry of tuberculosis. In a tuberculosis-endemic country, KFD is commonly mistaken for tuberculous lymphadenitis both cause prolonged fever, tender or matted nodes, a raised ESR and necrosis on histology and children are frequently started on empirical antitubercular therapy before a tissue diagnosis. In our cohort GeneXpert was negative in all 18 and no biopsy showed granulomas or acid-fast bacilli. Recognising the histological signature of KFD (karyorrhectic necrosis rich in crescentic histiocytes, devoid of neutrophils and plasma cells) and distinguishing it from caseating necrosis is essential to avoid unnecessary treatment.[6]

This distinction cannot be made reliably on fine-needle aspiration cytology (FNAC), whose diagnostic accuracy in KFD is low (around 50%) because the diagnosis depends on architecture rather than individual cells; FNAC is often misread as tuberculosis, reactive hyperplasia or lymphoma.[13] Excisional lymph-node biopsy remains the gold standard and was the basis of diagnosis in all our patients; we recommend early excisional biopsy rather than reliance on FNAC when KFD is suspected.

KFD is associated with autoimmune disease, especially SLE, which it may precede, accompany or follow, and from which it can be histologically indistinguishable.[7] In a retrospective study of 91 adults with KFD, 11 had SLE.[14] Three of our 18 children (16.7%) had SLE and one developed acute disseminated encephalomyelitis; a recent review of 386 pediatric cases similarly documented progression to lupus and rare central nervous system involvement.[15] Because KFD may antedate SLE by months to years, a normal autoimmune screen at presentation does not exclude later lupus; we advise structured long-term follow-up with periodic ANA, anti-dsDNA and complement testing, particularly in children with cytopenias, proteinuria or a positive ANA. Consistent with its self-limiting nature, all children responded to NSAIDs, with corticosteroids reserved for the 22% who had severe or autoimmune-associated disease, and no recurrence was documented in our cohort a pattern mirroring other pediatric series, in which NSAIDs have increasingly replaced antibiotics.[8,12] Recurrent pediatric KFD, though uncommon, has been reported from the region, including a 10-year-old child in whom the disease mimicked infection and malignancy; the absence of recurrence in our larger series is therefore reassuring, though longer follow-up is needed for confirmation.[16]

Limitations include the retrospective, single-center design, the small sample, incomplete data for some optional tests, the absence of routine viral serology and variable follow-up, so the absence of recurrence should be interpreted with caution.

CONCLUSION

KFD should be considered in any well child with prolonged febrile cervical lymphadenopathy, leukopenia and markedly raised ESR and LDH. Early excisional biopsy not FNAC is confirmatory and excludes tuberculosis and lymphoma, and long-term follow-up for evolving SLE is recommended.

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