Case Report

Kikuchi–Fujimoto disease: a case report

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ABSTRACT

Kikuchi–Fujimoto disease (KFD) or histiocytic necrotizing lymphadenitis is a rare, benign, self-limiting disorder that typically presents with cervical lymphadenopathy and fever. Because of its clinical similarity to infectious, autoimmune or malignant conditions, misdiagnosis is common. We report a case of a 41-year-old male presenting with unilateral cervical lymphadenopathy, fever and night sweats. Excisional lymph node biopsy and histopathology revealed features consistent with KFD, including necrotizing lymphadenitis with karyorrhectic debris and crescentic histiocytes without neutrophilic infiltration. Awareness regarding KFD is essential for clinicians and pathologists to avoid unnecessary interventions such as prolonged antibiotic therapy or chemotherapy. Early histopathologically confirmation can significantly reduce patient morbidity and healthcare costs.

***Key words**: Kikuchi–Fujimoto disease, necrotizing lymphadenitis, cervical lymphadenopathy.*

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INTRODUCTION

Kikuchi–Fujimoto disease (KFD), first described in Japan in 1972, is an uncommon cause of lymphadenopathy, predominantly affecting young adults, especially women.[1,2] Clinically, it presents as tender cervical lymphadenopathy often accompanied by fever, fatigue and weight loss. The disease is self-limiting, usually resolving within 1–4 months but its resemblance to lymphoma, systemic lupus erythematosus (SLE) and tuberculosis often complicates diagnosis.[3] Here, we report such a case.

CASE REPORT

A 41-year-old male presented with a 6-week history of progressively increasing left sided neck swelling associated with intermittent fever, night sweats and fatigue. There was no history of weight loss, recent infection or exposure to tuberculosis patient. His past medical history was unremarkable and he was not on any regular medication. He worked as a journalist and denied any high-risk exposure.

On examination, there was a firm, tender lymph node measuring approximately 2.5 cm × 2.5 cm in the left posterior cervical chain which was mobile and not fixed to underlying structure with no overlying skin change. No hepatosplenomegaly or generalized lymphadenopathy was noted.

Initial laboratory evaluation revealed normal blood picture with normal erythrocyte sedimentation rate (ESR) and Mantoux test revealed 12 mm induration at 72 hours. Serologic tests for human immunodeficiency virus (HIV), Epstein-Barr virus, cytomegalovirus, toxoplasmosis and anti-nuclear antibody (ANA) were negative. Chest X-

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ray was normal. Excisional biopsy of the lymph node demonstrated patchy necrotizing areas with karyorrhectic nuclear debris, abundant crescentic histiocytes, plasmacytoid dendritic cells and absence of neutrophils. These findings were consistent with KFD.

The patient was managed conservatively with non-steroidal anti-inflammatory drugs (NSAIDs) and supportive care. Symptoms improved gradually and lymphadenopathy resolved within 6 weeks.

DISCUSSION

KFD is rare but increasingly recognized worldwide, not only in Asian populations but also in Europe and North America.[4,5] The condition shows strong predilection to young women with female: male ratio ranging from 2 to 4:1 and peak incidence in 3rd decade of life.[6-8] The exact etiology remains uncertain, though viral and autoimmune mechanisms have been proposed.[3,9] HLA-DPB1*0202 allele has been noted as an association.[10] The underlying pathophysiology of KFD involves cell-mediated immune destruction. Current evidence suggests that cytotoxic CD8-positive T lymphocytes mediate apoptotic cell death, representing the primary mechanism of tissue damage.[10,11] Histiocytes appear to amplify this response through phagocytosis of apoptotic debris. The apoptotic process is thought to involve the Fas-Fas ligand pathway, with transmission electron microscopy revealing characteristic features including nuclear chromatin condensation, fragmentation and preservation of cellular organelles.[11,12]

Several inflammatory mediators have been implicated in disease pathogenesis. A study demonstrated elevated serum interferon-gamma and interleukin-6 during the acute phase, with normalization during recovery.[13,14] Immunophenotyping studies reveal T-bet-expressing CD4 and CD8 cells, along with B cells, within affected lymph nodes.[15] More recently, genomic approaches using high-throughput sequencing have identified candidate single-nucleotide polymorphisms and altered gene expression patterns that may serve as disease biomarkers and provide insights into pathogenesis.[14] These molecular findings may ultimately lead to improved diagnostic techniques and targeted therapies.

Clinically, KFD presents with tender cervical lymphadenopathy, often accompanied by constitutional symptoms. In a retrospective study, lymphadenopathy was universally present, while fever occurred in 35% and extra nodal manifestations including rash, arthritis and hepatosplenomegaly were less common.[3] Although KFD typically presents with localized lymphadenopathy, uncommon systemic manifestations have been increasingly recognized. Neurological complications, while rare, are clinically significant and include aseptic meningitis (the most frequent neurological finding).[15,16] Other reported manifestations include pulmonary (effusion, infiltrates and nodules),[17,18] musculoskeletal (polyarthritis, myositis), ocular (uveitis), endocrine (thyroiditis, parotid enlargement), hepatic (autoimmune hepatitis), renal and hematological (hemophagocytosis)[19,20] involvement.

Differential diagnoses include tuberculous lymphadenitis, lymphoma, SLE and viral infections. Histopathological examination remains the gold standard for diagnosis, as clinical and radiological findings are nonspecific.[5]

Histology shows characteristic biphasic evolution; proliferative phase shows follicular hyperplasia, paracortical expansion with mixed inflammatory infiltrate (lymphocytes, blasts, plasmacytoid monocytes, histiocytes) and prominent apoptosis. Preserved architecture, polyclonal pattern and negative viral studies exclude lymphoma and infection. In necrotizing phase, coagulative necrosis with CD68+ histiocytes (crescentic nuclei, phagocytosed apoptotic body), karyorrhectic nuclear debris and predominant CD8+ T cells. Absence of neutrophils distinguishes this from SLE and drug-induced lymphadenopathy.[20]

Key histologic differentials include, SLE: distinguished by presence of hematoxylin bodies, plasma cells and C4D immunohistochemistry,[21] Herpes simplex lymphadenitis, shows fewer mononuclear cells with neutrophilic infiltrate, Hodgkin lymphoma shows necrosis, contains neutrophils; Reed-Sternberg cells are CD15+/CD30+/CD45- and non-Hodgkin lymphoma, excluded by polyclonal infiltrate and architectural preservation.Distinguishing Features of KFD includes abundant CD8+ cytotoxic T cells around necrotic areas differentiate from SLE and reactive hyperplasia, increased plasmacytoid dendritic cells favor Kikuchi disease over reactive lymphadenitis or lymphoma[3,5] and digital CD123 quantification aids in diagnosis.[22]

Treatment is supportive; corticosteroids may be considered in severe or recurrent cases. Prognosis is generally excellent, with most patients recovering without sequelae. KFD typically occurs as an isolated condition; however, case reports have documented associations with several other disorders. Co-occurrence with Still's disease,[23] cryptogenic organizing pneumonia and SLE has been described. Of particular note, KFD has been reported following B-cell lymphoma, highlighting the importance of comprehensive evaluation and long-term follow-up in affected patients.[24]

Conclusion

This case of a 41-year-old man with KFD underscores two main clinical lessons. First, even though KFD most often occurs in young women, it should still be considered when assessing cervical lymphadenopathy in men and in individuals older than their early 30s. Second, the presentation can closely mimic tuberculosis, making accurate diagnosis particularly challenging in areas where tuberculosis is common. KFD should be considered in the differential diagnosis of cervical lymphadenopathy, particularly in young patients. Early recognition and histopathological confirmation can prevent misdiagnosis and unnecessary treatment.

Authors' contribution: MAR, RC planned the research. MAR drafted the manuscript. SAT revised the manuscript, BPD revised the slide. All authors read and approved the final manuscript for publication.

Consent: Informed written consent was taken from patients for publication of this case series along with accompanying images.

Conflicts of interest: Nothing to declare.

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