ORIGINAL RESEARCH PAPER
Volume - 14 | Issue - 10 | October - 2025 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

CASE REPORT: KIKUCHI-FUJIMOTO DISEASE

General Surgery

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KEYWORDS

1. INTRODUCTION

Kikuchi-Fujimoto disease (KFD), also known as histiocytic necrotizing lymphadenitis, is a rare, benign, and self-limiting condition of unknown etiology. It is characterized by subacute necrotizing lymphadenopathy, most commonly involving the cervical lymph nodes. While KFD is a benign disease, its clinical presentation often mimics serious conditions such as lymphoma, tuberculosis, and systemic lupus erythematosus (SLE), making a definitive diagnosis challenging. This case report describes a patient with a typical presentation of KFD and highlights the diagnostic process and management of this rare disorder.

2. Case Presentation

A 36 year-old female presented to the clinic with a history of fever and multiple neck swellings for 15 days. The fever was low-grade and intermittent, and she also reported associated fatigue and myalgia. There was no history of weight loss, night sweats, or cough. The patient had been treated with a course of oral antibiotics , but her symptoms showed no improvement. Her past medical and family history were unremarkable.

On physical examination, she was afebrile with a stable heart rate and blood pressure. Palpation of the neck revealed multiple, firm, and tender lymph nodes in the right posterior cervical region with pain raditing towards right shoulder and back.The largest lymph node was approximately 2cm in diameter. There were no other palpable lymph nodes in the axillary or inguinal regions. The remainder of the physical examination, including a throat and skin examination, was within normal limits.

3. Investigations and Differential Diagnosis

Initial laboratory investigations, including a complete blood count with differential, liver function tests, and kidney function tests, were all within normal reference ranges, except for a mild leukopenia. The erythrocyte sedimentation rate (ESR) was slightly elevated. Given the persistent lymphadenopathy despite antibiotic treatment, and the presence of systemic symptoms, the differential diagnosis included:
Infectious causes: Tuberculous lymphadenitis, viral infections (e.g., Epstein-Barr virus, cytomegalovirus), and bacterial lymphadenitis.
Autoimmune diseases: Systemic lupus erythematosus (SLE).
Malignancy: Malignant lymphoma.
To rule out these conditions and obtain a definitive diagnosis, a lymph node biopsy was performed.

4. Histopathology and Diagnosis

An excisional biopsy of one of the cervical lymph nodes was performed. The histopathological examination revealed characteristic features of KFD. The lymph node architecture was partially effaced by multifocal, circumscribed areas of necrosis. These necrotic areas showed an abundance of karyorrhectic debris (nuclear breakdown) and a proliferation of histiocytes and lymphocytes. Importantly, there was a notable absence of neutrophils and eosinophils in the necrotic areas, which is a key distinguishing feature of KFD.

Immunohistochemistry staining confirmed the findings, showing a predominance of CD8+ T cells and histiocytes positive for CD68. Autoimmune markers, including antinuclear antibodies (ANA) and anti-double-stranded DNA (anti-dsDNA), were negative, which helped to differentiate the condition from SLE. Based on the characteristic histological findings, a definitive diagnosis of Kikuchi-Fujimoto disease was made.

5. Treatment and Follow-up

As KFD is a self-limiting disease, the patient was managed with symptomatic treatment. She was advised to take non-steroidal anti-inflammatory drugs (NSAIDs) for pain and fever control as needed. Given the benign nature of the disease, no corticosteroids or immunosuppressants were initiated.

The patient was advised to follow up regularly. Over the next two months, her fever subsided completely, and the enlarged lymph nodes gradually decreased in size. The symptoms completely resolved within four months. The patient continues to be monitored for any recurrence or the development of associated autoimmune conditions, such as SLE, which can occur in a small percentage of cases.

6. DISCUSSION

This case illustrates a typical presentation of Kikuchi-Fujimoto disease, a condition that, despite its benign course, poses a significant diagnostic challenge. The patient's age and gender (a young female) and the presence of painful cervical lymphadenopathy and fever are classic features. The lack of response to antibiotics is a crucial clue that points away from a common bacterial infection.

The definitive diagnosis of KFD relies on an excisional lymph node biopsy. This is essential to differentiate it from conditions like lymphoma and tuberculosis, which require more aggressive treatment. Histologically, the triad of paracortical necrosis, karyorrhexis, and the absence of neutrophils is pathognomonic.

The etiology of KFD remains unclear, but it is believed to be a T-cell mediated immune response, possibly triggered by a viral infection or an autoimmune process. While the disease is generally benign and self-limiting, a small percentage of patients may experience recurrences or go on to develop autoimmune diseases, particularly SLE. Therefore, long-term follow-up is recommended.

7. CONCLUSION

Kikuchi-Fujimoto disease should be considered in the differential diagnosis for any young adult presenting with subacute cervical lymphadenopathy and fever, especially when there is no response to conventional antibiotic therapy. A high index of suspicion, coupled with a thorough physical examination and an excisional lymph node biopsy, is critical for accurate diagnosis. Early recognition of this condition can prevent unnecessary and potentially harmful diagnostic procedures and treatments, such as prolonged courses of antibiotics or even chemotherapy, while allowing for appropriate symptomatic management.