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EXTENSIVE CERVICAL LYMPHADENOPATHY MIMICKING MALIGNANCY: A CASE REPORT OF KIKUCHI-FUJIMOTO DISEASE IN AN ADOLESCENT FEMALE

Pathology

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ABSTRACT

Background: Kikuchi-Fujimoto Disease (KFD), or histiocytic necrotizing lymphadenitis, is a benign, self-limiting disorder that predominantly affects young females. Because its clinical and radiological presentation closely mimics lymphoma, systemic lupus erythematosus (SLE), and infectious diseases like tuberculosis, misdiagnosis is common.**Case Description:** A 16-year-old female presented with a painful right-sided neck swelling persisting for over a year. Clinical examination revealed a 4×5 cm infraparotid mass with palpable Level I and II lymph nodes. Computed tomography (CT) of the neck demonstrated widespread lymphadenopathy across Levels IB, II, III, IV, and V, with extension into the infraparotid space. An excisional biopsy of the Level IB lymph nodes was performed. Histopathological evaluation revealed effaced lymphoid architecture with focal map-like necroinflammatory zones, abundant nuclear dust (karyorrhexis), and an infiltration of plasmacytoid histiocytes. Crucially, there was a complete absence of neutrophils and well-formed granulomas, confirming a diagnosis of KFD. The patient was managed conservatively with close clinical follow-up.**Conclusion:** KFD should be considered in the differential diagnosis of young patients presenting with extensive cervical lymphadenopathy. Accurate histopathological identification prevents unnecessary, aggressive treatments like chemotherapy or antitubercular therapy.

KEYWORDS

Kikuchi-fujimoto Disease; Necrotizing Lymphadenitis; Cervical Lymphadenopathy; Case Report; Histopathology.

INTRODUCTION

Kikuchi-Fujimoto Disease (KFD), also known as histiocytic necrotizing lymphadenitis, is an uncommon, benign, self-limiting inflammatory condition first described independently by Kikuchi and Fujimoto in Japan in 1972 (1,2). While it has a higher prevalence among Asian populations, it is increasingly recognized globally. KFD most frequently manifests in young adults under the age of 30, showing a notable female preponderance (3).

The precise etiology of KFD remains enigmatic; however, current hypotheses point toward an irregular immune-mediated response triggered by viral entities (such as Epstein-Barr virus, cytomegalovirus, or Parvovirus B19) or underlying autoimmune conditions like systemic lupus erythematosus (SLE) (3,4). The primary clinical presentation is acute or subacute cervical lymphadenopathy, frequently accompanied by localized pain, low-grade fever, and fatigue. Because these features, combined with aggressive radiological appearances, closely masquerade as malignant lymphoma or tuberculous lymphadenitis, establishing an accurate histopathological diagnosis is paramount to prevent inappropriate clinical interventions (5).

Case Presentation

A 16-year-old female patient presented to the outpatient department with a chief complaint of a painful swelling on the right side of the neck. The swelling had been gradually progressing since its initial onset in April 2025. The patient's past medical history indicated prior clinical suspicion of Kikuchi's necrotizing lymphadenitis, though definitive tissue confirmation had not yet been established.

On localized physical examination, a prominent, tender 4×5 cm mass was palpable over the right infraparotid region. On deep palpation, distinct enlarged lymph nodes were noted at anatomical Levels I and II. The overlying skin showed no signs of acute suppurative changes, ulceration, or fistulization.

To evaluate the true anatomical extent of the mass, a Computed Tomography (CT) scan of the neck was performed. The imaging revealed an extensive, confluent nodal mass occupying the right infraparotid space, tracking closely along the parotid margin. Widespread lymph node enlargement was confirmed across multiple cervical segments, specifically involving **Levels IB, II, III, IV, and V**. Given the extensive, multi-level nature of the lymphadenopathy and apparent pericapsular involvement on imaging, a malignant lyric process such as lymphoma was considered a high-priority differential diagnosis.

The patient was subsequently scheduled for an open excisional biopsy of the right Level IB lymph nodes under surgical guidance to obtain material for histopathological examination.

Pathological Findings

The Department of Pathology received an excision specimen labeled as "Level IB lymph nodes."

Gross Examination

The specimen consisted of multiple (12) soft tissue fragments received altogether, measuring collectively 4.5×4.0×0.5 cm. The tissue pieces were soft to firm in consistency, displaying a patchy gray-white appearance on the cut surface without gross evidence of caseous necrosis or abscess formation. The tissue was fixed in standard formalin for microscopic processing.

Microscopic Examination

The tissue sections stained with hematoxylin and eosin (H&E) revealed a significantly effaced lymphoid architecture. The paracortical and cortical regions were interrupted by irregularly shaped, geographic, pale areas of **necroinflammatory changes**.

These necrotizing zones were heavily crowded with abundant **karyorrhectic debris** (apoptotic nuclear dust) and apoptotic cellular fragments. Interspersed within and surrounding these necrotic foci was a prominent proliferation of **plasmacytoid histiocytic cells** and immunoblasts. The histiocytes displayed characteristic eccentric, crescentic nuclei and abundant eosinophilic cytoplasm.

Examination of the structural boundaries showed that the

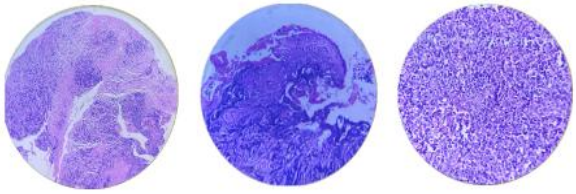
inflammatory process breached the lymphoid capsule, extending directly into the neighboring perinodal fat and skeletal muscle fibers.

Critical Diagnostic R exclusion Criteria:

- **No granulomas** were identified across any step-sections, effectively ruling out tuberculous lymphadenitis and sarcoidosis.
- **No neutrophilic infiltrate** or plasma cells were detected within the areas of necrosis, ruling out acute bacterial suppurative lymphadenitis.
- **No atypical monoclonal lymphoid sheets** or Reed-Sternberg cells were present, eliminating Hodgkin or non-Hodgkin lymphoma.

Histopathological Impression

Features are highly suggestive of Kikuchi Lymphadenitis (Kikuchi-Fujimoto Disease / Histiocytic Necrotizing Lymphadenitis).



DISCUSSION

This case illustrates the classic clinical and pathological journey of Kikuchi-Fujimoto disease, while underscoring the potential for severe radiological misinterpretation. In adolescent patients, the finding of multi-level cervical lymphadenopathy extending from Level IB down through Level V is highly alarming. When paired with CT imaging showing an infraparotid mass with extracapsular tracking into adjacent adipose and muscular tissue, a clinician's index of suspicion heavily leans toward lymphoma or an invasive chronic infectious disease like tuberculosis (5,6).

However, KFD is a critical "great masquerader" in pediatric and young adult medicine. While the imaging findings in our patient looked aggressive, the excisional biopsy provided definitive clarity. The microscopic findings in this case represent the pathognomonic footprint of the necrotizing phase of KFD: geographic necrotizing foci loaded with karyorrhectic fragments, a rich infiltrate of histiocytes with eccentric nuclei, and—most importantly—the **complete absence of neutrophils** (6,7).

The absence of neutrophils is the single most valuable histological differentiator. A standard necrotizing infectious process (such as a bacterial abscess or cat-scratch disease) triggers an influx of polymorphonuclear neutrophils. Similarly, tuberculosis would present with classic caseating epithelioid granulomas and Langhans giant cells, which were completely absent here (7,8).

Recognizing KFD is vital because it is fundamentally a benign, self-limiting disorder. The extensive node involvement across levels I-V seen on this patient's CT scan could easily have led to unnecessary staging or cytotoxic chemotherapy. Instead, because KFD was diagnosed correctly via histopathology, treatment can be safely restricted to supportive care. Management relies primarily on non-steroidal anti-inflammatory drugs (NSAIDs) to alleviate localized pain and tenderness, with spontaneous resolution expected within 1 to 4 months (8).

Nonetheless, long-term clinical monitoring is indicated for this 16-year-old patient. Literature indicates a clear pathogenetic overlap between KFD and Systemic Lupus Erythematosus (SLE). A small percentage of patients diagnosed with KFD either have synchronous SLE or develop it several years down the line, necessitating periodic autoimmune serology screening during follow-up (3,9).

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