

Atypical Presentation of Kikuchi-Fujimoto Disease: Diagnostic Challenges in a Case of Persistent Cervical Lymphadenopathy with Acute Onset Quadriplegia

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Abstract: Kikuchi-Fujimoto disease (KFD), is rare, self-limiting disorder with necrotizing lymphadenitis as its hallmark, can be difficult to diagnose because it may have multiple nonspecific features. The study presented here describes the rare type of KFD that occurred in a young male patient suffering from quadriplegia and who was wrongly diagnosed as having tuberculous lymphadenitis, which is a common cause of lymphadenopathy in tuberculosis-endemic area. A 19-year-old man had presented with two week history of fever, quadriplegia, and cervical lymphadenopathy. On physical examination revealed cervical lymphadenopathy. A laceration procedure was performed to address the lymphadenopathy. The patient was treated with anti-tubercular medication for 11 days. Currently, his urinary and bowel functions are stable, and he is fully conscious, alert, and oriented to time, place, and person. Histopathology showed classical changes in histiocytic necrotizing lymphadenitis in the lymph nodes with no evidence of either tuberculosis or neoplasm. This KFD case is particularly glaring in terms of the obstacles it presented in making a diagnosis due to its endemicity of tuberculosis. The case actually had a complicated clinical picture with KFD's initial presentation of quadriplegia. Hence, the list of differential diagnosis should include KFD as one of the uncommon causes. Timely recognition and appropriate management of KFD can prevent unnecessary treatments and improve patient outcomes.

Keywords: Kikuchi-Fujimoto disease, lymphoma, tubercular lymphadenopathy

Introduction

Kikuchi disease, also referred to as Kikuchi-Fujimoto disease (KFD), is an uncommon yet benign illness that usually manifests as fever and cervical lymphadenopathy. Hepatosplenomegaly, dermatitis, arthritis, and exhaustion are possible further symptoms.¹ An early diagnosis can provide reassurance to patients and prevent unnecessary tests and treatments. KFD highlights the diagnostic challenges associated with the variable nature of lymphadenitis and the significant fluctuations in hematological parameters that correlate with disease activity.² KFD is linked to various autoimmune diseases, including systemic lupus erythematosus (SLE), polymyositis, lymphoma, and scleroderma. It is also associated with thyroiditis and Sjogren's syndrome, an immune system disorder.³ Kikuchi-Fujimoto disease (KFD), also referred to as benign histiocytic necrotizing lymphadenitis, is a rare condition. Initially discovered in Japan in 1972, it has been documented worldwide, with the majority of cases occurring in Asia. Kikuchi-Fujimoto disease (KFD) commonly affects jugulo carotid and cervical lymph nodes, measuring 0.5 to 6.0 cm, with tenderness or pain in 50% of cases. Key histological features include necrosis with karyorrhexis, histiocytic infiltration, and plasmacytoid dendritic cells without neutrophils. Recognizing KFD is crucial, as it can mimic infections, inflammatory conditions, autoimmune diseases (eg, SLE), and cancers like leukemia and lymphoma.⁴ Infectious agents including *Yersinia enterocolitica*, *Brucella*,

Bartonella henselae, *Entamoeba histolytica*, *Mycobacterium szulgai*, and *Toxoplasma gondii* were initially considered, but later studies did not support these associations. Various viruses—such as Epstein–Barr virus, herpesvirus, cytomegalovirus, parvovirus, paramyxovirus, parainfluenza virus, rubella virus, Hepatitis B, HIV, and Human T-lymphotropic virus type 1—were also proposed as potential causes of KFD, but none have been confirmed.⁵

Case Presentation

A 19-year-old male chief complaints quadriplegia and fever for the past 2 weeks. Physical examination notable lymphadenopathy in the cervical region. Vital Signs: BP: 127/89 mmHg, Respiratory Rate: 96/min, Pulse Rate: 97 bpm/SpO₂: 99%, Blood/Sugar: 109 mg/dL, Temperature: 37.6°C and Lungs: BAE+, no added sounds. Intervention incision procedure performed to address lymphadenopathy. Current treatment is anti-tubercular medication administered for 11 days. The neurologic examination revealed flaccid quadriplegia with significantly reduced muscle strength in all extremities (upper limbs: 1/5, lower limbs: 2/5). Deep tendon reflexes were diminished, and sensory examination showed hypoesthesia in a stocking-and-glove distribution. Cranial nerve function was intact, and no signs of bowel or bladder dysfunction were noted. Neurological Complications Secondary to KFD, the potential pathway we considered is Immune-Mediated Myelitis: The hyperinflammatory or autoimmune milieu in KFD might trigger an immune response leading to acute transverse myelitis or other spinal cord inflammation. The patient was started high-dose steroids after 2 weeks the neurologic complication disappeared gradually.

Laboratory Results: Hemoglobin was 11 g/dL; RBC count was 4.6 million/ μ L; WBC count was 3,190/ μ L, with neutrophils at 31.7%, lymphocytes at 40.8%, eosinophils at 18.9%, and monocytes at 8.5%. Platelet count was 388,000/ μ L. Hemogram showed sporadic reactive cells and moderate leukopenia. Erythrocyte sedimentation rate was elevated. Random plasma glucose was 101 mg/dL. Serum creatinine was normal at 0.6 mg/dL. Total bilirubin was 0.1 mg/dL, direct bilirubin was 0.09 mg/dL. Albumin was 3.8 g/dL, and serum urea was 21 mg/dL. PT with INR was 1.3 seconds. Lumbar Puncture Results: Protein levels were decreased; other parameters were normal. Decreased CSF protein levels may suggest conditions such as a spinal fluid leak or metabolic/nutritional disorders and should be correlated with clinical symptoms and other diagnostic results. The brain MRI examination is normal. Cervical MRI examination revealed several lymphadenopathies in the anterior and posterior cervical chains, with the largest measuring 2.5 cm on the right (Figure 1). EMG Findings show sagittal median, ulnar, and tibial nerve potentials were absent. The right peroneal nerve showed very low amplitude potentials. Sensory nerve

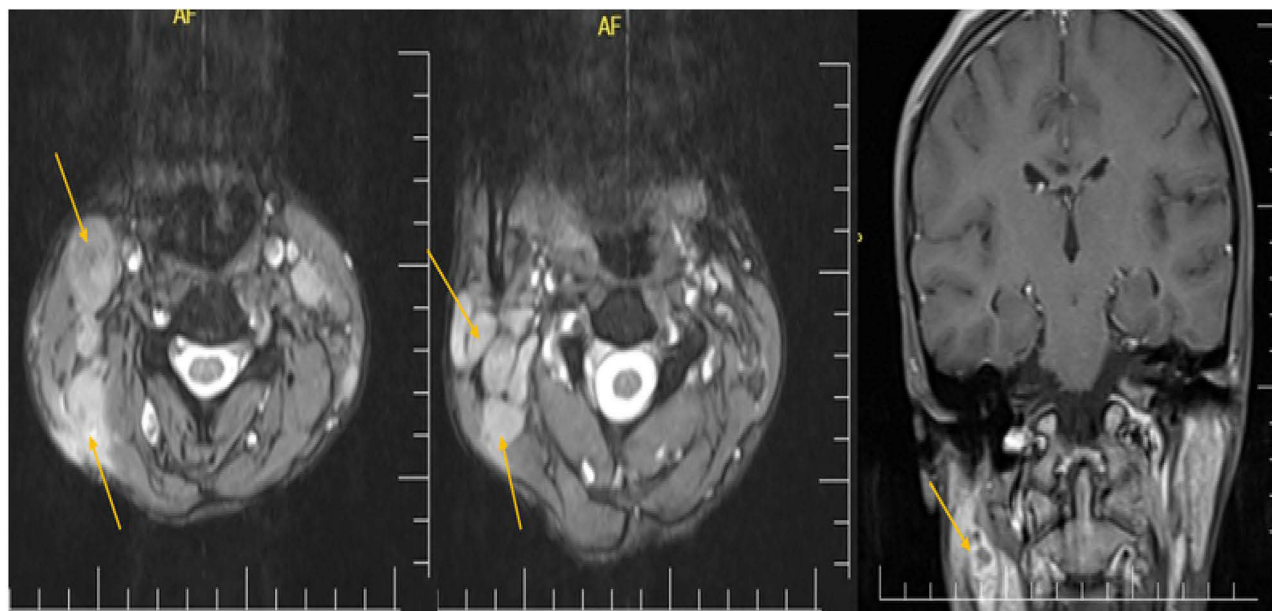


Figure 1 Cervical axial T2-weighted images and coronal T1-weighted post-gadolinium scans revealed multiple conglomerate cervical lymphadenopathies (Arrows).

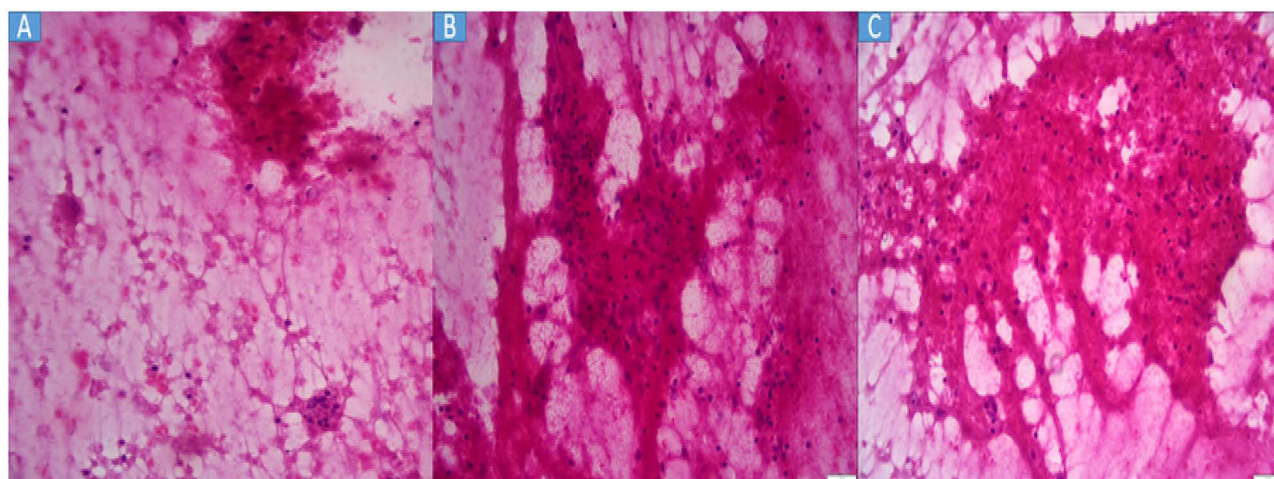


Figure 2 (A–C) FNAC smear showing crescentic histiocytes, plasmacytoid monocytes, phagocytic histiocytes with peripheral nuclei and extracellular (karyorrhectic) debris in a necrotic background (H and E, $\times 400$).

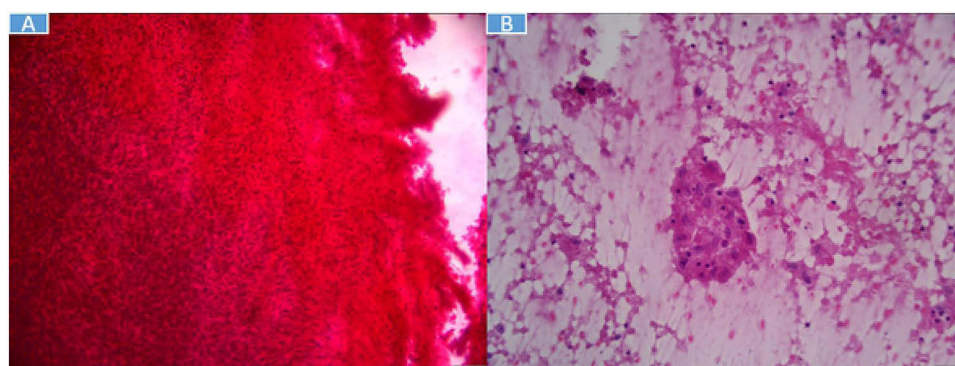


Figure 3 (A) Granulomatous inflammation in a background of necrosis with karyorrhectic debris. **(B)** Shows plasmacytoid monocytes in the background of necrosis surrounded by karyorrhectic debris.

conduction was normal. Needle EMG indicates no motor unit activity and signs of denervation and is consistent with acute onset quadriplegia. The patient is admitted to neurology service.

Pathological analysis of the lymph node aspiration; KFD is diagnosed based on cytopathologic findings obtained from fine-needle aspiration results. Cytopathological findings include varying degrees of coagulative necrosis with abundant karyorrhectic debris and an absence of eosinophils or neutrophils. Crescent-shaped histiocytes and plasmacytoid monocytes were seen surrounding the necrotic areas (see Figures 2 and 3). Such a pathological tool would be useful for both diagnostic and differential diagnosis.

Discussion

Kikuchi's disease, also known as Kikuchi-Fujimoto disease or histiocytic necrotizing lymphadenitis, was first described by a group of Japanese researchers in 1972. It is characterized by lymphadenitis with prominent nuclear debris, numerous histiocytes, and localized reticular cell proliferation.⁶ Lymphadenopathy and histopathology: Both cases exhibit cervical lymphadenopathy with histopathological findings of necrotizing lymphadenitis, characterized by nuclear debris and histiocytes, consistent with Kikuchi disease. Neurological Involvement: Our case includes acute motor axonal polyneuropathy leading to quadriplegia, which is not a typical feature of Kikuchi disease, whereas the other case focuses solely on lymphadenitis without neurological symptoms.

According to Bosch and et al, unilateral cervical lymphadenopathy in the posterior triangle lymph nodes was the most common clinical manifestation, occurring in 56–98% of cases. Lymphadenomegaly, typically ranging from 0.5 to 4 cm

and rarely exceeding 6 cm, was observed in 30–50% of patients, often accompanied by upper respiratory symptoms and low-grade fever. Our patient similarly presented with a low-grade fever and right cervical lymphadenomegaly measuring 1.3 cm. Skin involvement, seen in 40% of cases, can vary from a general rash to lupus-like symptoms.⁷ Skin Involvement: While skin involvement occurs in up to 40% of cases in Bosch's study, our patient did not exhibit any skin manifestations, such as rashes or lupus-like symptoms. Cervical Lymphadenopathy and Fever: Both cases present with unilateral cervical lymphadenopathy and low-grade fever, common clinical features in Kikuchi disease. Our patient's lymph node size (1.3 cm) aligns with the typical range noted in Bosch's study (0.5–4 cm). There are various restrictions on this case study. First, an invasive excisional biopsy is necessary for the diagnosis of KFD. Second, it is not possible to predict future relapse or progression to SLE during the brief follow-up period. Thus, it is essential to monitor KFD patients throughout the long term. Furthermore, more research is needed for KFD.⁸ Long-term Monitoring and Complications: While the cited case emphasizes the need for long-term monitoring of relapse or progression to SLE, our case does not mention ongoing concerns about recurrence or autoimmune complications at this stage. Additionally, no meningitis was observed in our patient. Diagnosis by Biopsy: Both cases required invasive excisional biopsy for a definitive diagnosis of Kikuchi disease, highlighting the importance of histopathological evaluation. Based on current understanding, isolated anti-DFS70 antibodies can be used as a biomarker for diagnosis to help rule out systemic autoimmune illness.⁹ Use of Anti-DFS70 Antibodies: The cited case discusses the role of anti-DFS70 antibodies as a biomarker to rule out autoimmune diseases, while our case does not mention the use of this specific antibody for diagnostic clarification. Autoimmune Considerations: Both cases raise the possibility of an underlying systemic autoimmune illness, as autoimmune diseases can mimic Kikuchi disease histologically. Microscopic observations revealed nodal tissue with a deformed architecture made up of lymphocytes and histiocytes intermingled with eosinophilic, granular material, and karyorrhectic detritus. There are still residual primary follicles, but either nonexistent or very rare are neutrophils and plasma cells. There were no abnormal cells or granuloma formations seen. These characteristics rule out lymphoma and tubercular lymphadenitis and support Kikuchi's illness diagnosis.¹⁰ Presence of Eosinophils: The cited case notes eosinophilic and granular material in the biopsy, while our case did not emphasize eosinophilic infiltration in the microscopic findings. Histopathological Features: Both cases display lymphocytes, histiocytes, and karyorrhectic debris in the lymph node biopsy, consistent with Kikuchi disease. Additionally, granulomas and abnormal cells were absent in both, ruling out lymphoma and tuberculosis.

Conclusion

The diagnosis of Kikuchi disease was confirmed after ruling out lymphoma and tuberculosis through biopsy. This case demonstrates an atypical presentation of Kikuchi-Fujimoto Disease (KFD) with persistent cervical lymphadenopathy, fever and acute onset quadriplegia, a rare but significant complication. While KFD is classically considered a self-limiting condition, this case highlights its potential for systemic and neurological involvement, underscoring the importance of recognizing such rare manifestations.

The diagnosis required a multidisciplinary approach, involving histopathological confirmation of KFD through lymph node biopsy and advanced investigations, including MRI of the spinal cord, and CSF analysis, to identify immune-mediated myelitis. Early recognition and timely initiation of immunosuppressive therapy were crucial in managing the neurological deficits and preventing further deterioration.

Data Sharing Statement

We declared that we had full access to all of the data in this case report, and we take complete responsibility for the integrity of the data. All original data are available at Mogadishu Somali Turkish Training and Research Hospital, Mogadishu, Somalia. Data used to support the findings of this study are available from the corresponding author upon request.

Ethics Approval

Based on the regulations of the review board of the Mogadishu Somali Turkish Training and Research Hospital, institutional review board approval is not required for case reports.

Consent for Publication

Written informed consent had obtained by the patient to have the case details and any accompanying images published.

Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, case presentation, or in all these areas; took part in drafting, revising or critically reviewing the case; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

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Disclosure

The authors declare no conflicts of interest in this work.

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