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Kikuchi-Fujimoto Syndrome: A Rare Entity to Consider

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ABSTRACT

Introduction: Kikuchi-Fujimoto disease (KFD) is a rare, benign, necrotizing lymphadenitis of unknown aetiology with characterized by cervical lymphadenopathy, nocturnal diaphoresis and fever. Surgical excision of the adenopathy, his and immunophenotyping are crucial for diagnosis.

Patients and methods: This paper describes five patients with three different histological subtypes of KFD, including an a masquerading as pyelonephritis and two other cases where physicians mistakenly started chemotherapy. In one other c was identified as the responsible aetiological agent, while in the remaining patient, KFD evolved into an autoimmune c

Discussion: KFD, although rare, may mimic infectious, autoimmune and neoplastic diseases. It also poses a risk development of an autoimmune disorder.

LEARNING POINTS

- Kikuchi-Fujimoto disease (KFD), although rare, should be included in the differential diagnosis of patients with cervic and fever of unknown origin.
- Early recognition of KFD may minimize the use of unnecessary aggressive examinations and therapies.
- The course of KFD in most patients is self-limiting, but there is a risk of progression to an autoimmune syndrome.

KEYWORDS

Kikuchi-Fujimoto disease, lymphadenitis, fever of unknown origin, rare disease

INTRODUCTION

Kikuchi-Fujimoto disease (KFD)is a rare, benign, necrotizing lymphadenitis of unknown aetiology, with fever and pref of the cervical region[1]. This self-limiting condition may, due to its similarity, be confused with tuberculosis, systemic l and haematological malignancies[2, 3]. Thus, surgical excision of the affected adenopathy, histological study and immunop for establishing the correct diagnosis[3].

No aetiological factors have been identified, but it is thought that this syndrome results from an immune response by C and histiocytes to an infectious insult[1–3]. The main aetiological cause is assumed to be viral[3].

KFD, similarly to diseases such as systemic lupus erythematosus or other autoimmune conditions, is primarily fou However, there are reports of affected male patients, as well as individuals aged between 6 and 80 years[3].

The most frequent signs and symptoms are cervical and localized lymphadenopathies, fever, rash, arthritis, fatigue and h Other uncommon symptoms are night sweats, nausea, vomiting, diarrhoea, neck stiffness, weight loss and more ex involvement [3,4].

involvement [5, 6].

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The literature describes cases of KFD associated with pathologies such as Still's disease, B-cell lymphoma and cryptog risk of it developing into an autoimmune condition is higher in adults, and patients with late-onset systemic lupus eryth reported[3].

Surgical excisional biopsy is vital, as there are no radiological or ultrasound characteristics to establish the diagnosi three histological subtypes: proliferative, necrotizing and xanthomatous. The last subtype, unlike the previous two v progression, is a distinct histological variant of undetermined aetiology[2,3].

CASE DESCRIPTION

Case 1

A 72-year-old female patient with haematuria, dysuria and pollakiuria was admitted with pyelonephritis. A thor CT scan revealed accidental thoracic and abdominal adenopathy. Laparoscopic excision of the hepatic ganglia revea granulomatous lymphadenitis.

Case 2

A 19-year-old man presented with odynophagia, fever, rash, asthenia, anorexia and weight loss. Hepatomegaly, right c and neurological worsening were documented. The patient underwent a cycle of chemotherapy due to high suspicion of a syndrome with central nervous system involvement. Subsequently, excisional biopsy of the cervical adenomegaly cc lymphadenitis.

Case 3

A 58-year-old woman was admitted with to a 2-month history of myalgia, asthenia, weight loss, nocturnal diaphoresis productive cough. In light of clinical worsening, and the development of unilateral pleural effusion and bilateral axilla patient started a chemotherapy regimen due to suspicion of lymphoma. A biopsy of the axillary adenomegaly later hyperplasia.

Case 4

A 58-year-old man was admitted for painful adenomegaly measuring 3 cm in diameter in the right inguinal region, ass thoracic-abdominopelvic CT scan showed contrast uptake only in the enlarged gland proximal to the right external femc was positive for HBsAg, IgG CMV and IgM CMV. Excisional biopsy later confirmed necrotizing lymphadenitis.

Case 5

A 31-year-old female patient with KFD, which had been histologically confirmed 3 years previously, developed scal lesions on the face and scalp. A skin biopsy revealed discoid lupus erythematosus.

DISCUSSION

Despite the good prognosis of this clinical entity, patients with a fever of undetermined aetiology with associated should undergo surgical biopsy[3]. Two of the described patients mistakenly underwent chemotherapy due to high suspi KFD can be even more challenging to diagnose when there is ganglion involvement not localized to the cervical re abdomen resulting from this pathology have been described, and its presentation as pyelonephritis, as described above, To date, no therapeutic guidelines has been established for KFD. Patients with classic symptoms respond favourably t inflammatory drugs. However, patients with atypical and refractory symptoms may require corticosteroid therapy [3]. patients receiving intravenous immunoglobulin, hydroxychloroquine or combination therapy [3].

Most cases of KFD are self-limiting, but some patients may experience relapse. The recurrence rate is less than 7%, a patients with asthenia, non-localized ganglion involvement and persistent symptomatology are at higher risk of relaps There are no standardized methods for managing this condition. Therefore, patients with KFD should be followed u recurrence and also the high probability of the development of autoimmune conditions such as systemic lupus erythen

In general, KFS should be considered in the differential diagnosis of patients with cervical lymphadenopathy and fever
its early recognition can minimize the use of aggressive tests and therapies, and iatrogenesis [2–5].

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REFERENCES

1. Pandey V. Kikuchi-Fujimoto disease masquerading as acute appendicitis. *J Clin Diagn Res* 2017;**11**(6):ED26–ED28.
2. Vijayaraghavan R, Chandrashekar R, A S, Belagavi C. Kikuchi-Fujimoto's disease involving mesenteric nodes: a report and review of literature. *BM* bcr1020114945.
3. Richards MJ. Kikuchi disease. UpToDate [Internet]. Available from: https://www.uptodate.com/contents/kikuchi-disease
4. Song JY, Lee J, Park DW, Sohn JW, Suh SI, Kim IS, et al. Clinical outcome and predictive factors of recurrence among patients with Kikuchi's disease. *Int J* 326.
5. Parappil A, Rifaath AA, Doi SAR, Pathan E, Surrun SK. Pyrexia of unknown origin: Kikuchi-Fujimoto disease. *Clin Infect Dis* 2004;**39**(1):138–143.

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