Kikuchi–Fujimoto disease as a rare cause of fever of unknown origin in a septuagenarian

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Abstract
We report the case of a 72-year-old patient presented with fever of unknown origin. Initial clinical and radiological findings suggested a diagnosis of lymphoma. However, subsequent histology revealed Kikuchi-Fujimoto disease (KFD). KFD is predominantly a self-limiting disease of the young, but should be considered in the differential diagnosis of older patients presenting with fever of unknown origin or features suggestive of lymphoma.

Keywords: Kikuchi-Fujimoto disease, fever, necrotising lymphadenitis, elderly

Introduction
Kikuchi–Fujimoto disease (KFD) is a rare self-limiting idiopathic necrotising lymphadenitis. It was first described in Japan in 1972 [1, 2]. It is associated with systemic lupus erythematosus, non-infectious inflammatory conditions, viral infections and is more common in females (77%), especially those of Asian origin [3]. The disease frequently mimics high-grade lymphoma and may be mistaken for and treated as lymphoma [4, 5]. A recent review of 244 published cases since 1991 showed the mean age for presentation to be 25 (range 1–64), with 70% being younger than 30 years [3]. We describe a patient aged over 70 years with KFD who presented with fever of unknown origin.

Case presentation
A 72-year-old Caucasian female with a history of rheumatoid arthritis (quiescent on methotrexate) presented with a 2-month history of fever, malaise and weight loss. Clinical examination revealed a fever of 38.4°C, which persisted intermittently for 14 days, and a few small inguinal and cervical lymph nodes. There was no evidence of active rheumatoid disease and no hepatosplenomegaly was found on physical examination.

The white blood cell count displayed lymphopenia (lymphocytes 0.9) while her haemoglobin was 8.6 g/dl mean cell volume 85.4. The erythrocyte sedimentation rate (ESR) and C reactive protein (CRP) were elevated at 98 mm/h (max 130) and 138 (max 228), respectively. Serial blood cultures showed no growth. Chest X-ray was normal and trans-thoracic echocardiogram showed no evidence of vegetations. Methotrexate was stopped on admission. She was transfused 2 units of blood and treated with oral antibiotics and then with intravenous antibiotics, with no improvement in her fever or inflammatory markers.

Subsequent CT imaging of the abdomen and thorax showed widespread pathological lymphadenopathy and splenomegaly (Figure 1). This was reported to be highly suggestive of lymphoma.

While awaiting lymph node biopsy, the patient's fever abated and inflammatory markers improved (ESR 31, CRP 15).

An excision biopsy of an enlarged inguinal lymph node showed areas of amorphous necrosis, containing nuclear debris, and surrounded by a loose collection of histiocytes and lymphocytes in keeping with a diagnosis of KFD. The patient made an uneventful recovery and was discharged after 30 days. When seen at the clinic 2 months post-discharge, she was well with normal inflammatory markers and full blood count. A repeat CT scan at 3 months showed no pathological lymphadenopathy.

Discussion
KFD is a rare idiopathic condition predominantly affecting young patients of Asian origin. We describe a case in an
elderly Caucasian patient. It commonly presents with cervical lymphadenopathy and while mediastinal and peritoneal involvement is unusual, cases involving these nodes groups and/or extra nodal involvement of the liver and spleen are well documented [3, 6] (Table 1).

In addition to this, our patient had many of the more commonly associated findings in KFD—fever (35%), fatigue (7%), lymphadenomegaly (100%), hepatosplenomegaly (3%), leucopenia (43%), high ESR (40%) and anaemia (23%) [3]. The systemic presenting symptoms, as seen in our patient, are more common in cases with extranodal disease [7].

Imaging in KFD varies, but frequently demonstrates nodal enlargement with necrosis. As in our case, these findings encompass a wide differential, including lymphoma, metastasis and tuberculosis. Fine needle aspiration cytology is estimated to have a overall diagnostic accuracy of 56.3% in KFD [8] and as such, excision biopsy remains mandatory for diagnosis if clear cut cytological KFD findings are absent [9].

Accurate and early diagnosis is essential as KFD is generally a benign and self-limiting condition, with a minority of patients (16%) requiring corticosteroid treatment and an overall mortality of 2.1% [3]. The diagnosis should be considered even in elderly patients presenting with features typical of lymphoma.

The patient described in the case has given her consent for the case to be published.

Key points

- KFD could occur in a patient over the age of 70 years presenting with fever of unknown origin.
- Recognition of this condition is crucial, especially because KFD frequently mimics, and may be mistaken for lymphoma.
- CT findings vary but typically show lymphadenopathy with necrosis, and excision biopsy remains the gold standard for diagnosis.
- KFD typically is self-limited, and lasts from 1 to 4 months, treatment is generally supportive.

Conflicts of interest

None

References


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