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PHOTO QUIZ

An unusual cause of cervical lymphadenopathies

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CASE REPORT

A 41-year-old female presented to our department with a three-week history of fever, and painful and tender swelling in the right side of her neck. After several cycles of antibiotherapy no improvement was observed.

On admission, her general condition was good, with an axillary temperature of 38°C, which usually increased at night. On physical examination, she had multiple and tender lymphadenopathies located in the right upper and middle jugular chain and posterior triangle of the neck; the largest was 2 x 2 cm in diameter. Physical examination revealed no other abnormalities.

Laboratory studies showed no abnormalities. A purified protein derivative (PPD) test was negative. Blood, urine, and throat cultures grew no organisms. Serology titres for Ebstein-Barr virus, cytomegalovirus, herpes simplex virus, *Toxoplasma*, *Rubella* and *Brucella* were negative. A cervical magnetic resonance imaging (MRI) scan demonstrated multiple lymphadenopathies which were 1.5 x 2 cm in diameter in the right upper and lower anterior cervical region and posterior cervical region (*figure 1*).

Due to the persistence of fever and swelling for three weeks, a fine-needle aspiration cytology was performed. Cytological features suggested a reactive lymphadenitis, but a proliferative disease could not be excluded. Thus, an excisional biopsy was performed, with removal of one of the enlarged cervical nodes. Histopathological examination of cervical lymph node biopsy disclosed necrotic areas, histiocytic accumulation, lymphocyte and immunoblast infiltration. Immunohistochemical study showed positive staining for CD68 on histyoctes (*figure 2*).

WHAT IS YOUR DIAGNOSIS?

See page 263 for the answer to this photo quiz.

Figure 1. *T1 sagital section of the MRI showing multiple lymphadenopathies in the right posterior cervical region*

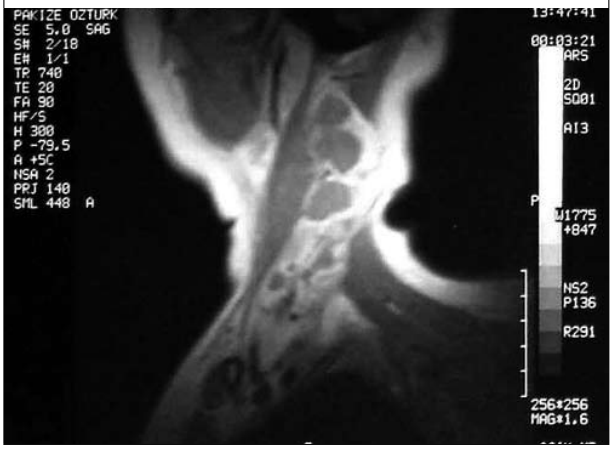
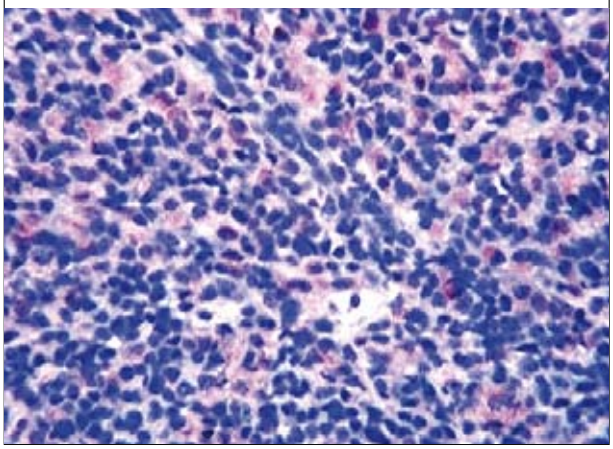


Figure 2. *Immunohistochemical study shows positive CD68 staining on histiocytes (DAB Chromogen 400 x)*



DIAGNOSIS

Based upon these histopathological findings, the diagnosis of Kikuchi-Fujimoto histiocytic necrotising lymphadenitis was made. There are no definite laboratory tests available for the diagnosis of Kikuchi-Fujimato disease (KFD). Definitive diagnosis of KFD can only be made on direct histopathological examination of a lymph node biopsy. The typical histopathological features of the KFD include lymph node necrosis with karyorrhexis surrounded by histiocytes, without granuloma, neutrophil or plasma cell infiltration.[1] Its recognition is crucial, especially because this disease can be mistaken for systemic lupus erythematosus and malignant lymphoma.[2]

There is no radiographic finding specific for KFD. Computed tomography and MRI do not yield features that distinguish KFD from other diseases which commonly involve lymph nodes such as lymphoma, tumour metastases, or tuberculosis.[3]

The characteristic clinical presentation is cervical lymphadenopathy, which is often painful or tender on palpation. Occasionally the sole symptom of the disease is a fever of unknown origin without other associated clinical features. Additional complaints include nausea, diarrhoea, headache, dermatological lesions and constitutional disturbances.[2]

There is no specific treatment for KFD, analgesics-antipyretics, nonsteroidal anti-inflammatory drugs and corticosteroids may be used to relieve distressing local and systemic complaints. Spontaneous recovery occurs in one to four months. However, a small percentage of cases may have a recurrence within a few weeks from the first event and relapses have been described many years after the initial episode.[4] Together with the high risk for development of an autoimmune disease or malignancy such as lymphoma, these characteristics mean that long-term follow-up is mandatory for these patients.

Cervical lymph node enlargement with a prolonged fever requires a careful differential diagnosis which should include the possibility of KFD.

REFERENCES

1. Dorfman RF, Berry GJ. Kikuchi's histiocytic necrotizing lymphadenitis: an analysis of 108 cases with emphasis on differential diagnosis. Semin Diagn Pathol 1988;5:329-45.
2. Yılmaz M, Camcı C, Sari I, et al. Histiocytic necrotizing lymphadenitis mimicking systemic lupus erythematosus: a review of two cases. Lupus 2006;15:384-7.
3. Na DG, Chung TS, Byun HS. Kikuchi disease: CT and MR findings. Am J Neuroradiol 1997;18:1729-32.
4. Mohanty SK, Arora R, Saha M. Kikuchi–Fujimoto disease: an overview. J Dermatol 2002;29:10-4.