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KIKUCHI — FUJIMOTO DISEASE: CERVICAL LYMPHADENOPATHY SUGGESTIVE OF RELAPSING LYMPHOMA IN PATIENT WITH LYMPHOBLASTIC LYMPHOMA

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***Aim*: Kikuchi — Fujimoto disease (KFD) or histiocytic necrotizing lymphadenitis is a rare disorder and often confused with lymphoma. *Patient*: There is presented a case of 28-year-old patient with cervical lymphadenopathy, who had history of lymphoma. *Results*: On immunohistopathologic examination diagnosis of KFD was made and patient followed without any treatment. *Conclusion*: Patient's lymphadenopathy had almost resolved and he was completely asymptomatic after three months. In patient with cervical lymphadenopathy KFD should be considered in the differential diagnosis.**

***Key Words*: lymphoma, cervical lymphadenopathy, Kikuchi — Fujimoto disease.**

KFD or histiocytic necrotizing lymphadenitis is a rare disorder. Although it is primarily affecting young adults of Asian descent, KFD is being increasingly reported in other areas [1–3]. It is generally a self-limiting benign condition and is characterized by patients presenting with cervical lymphadenopathy, fevers and malaise. The clinical features can also be consistent with malign lymphomatous disorders [4]. The clinician being referred patients with cervical lumps should be aware of this condition as part of their clinical assessment. Histological examination is the only means of definitive diagnosis, so the provision of clear clinical information and adequate biopsy material to the pathologist is critical.

***Case report*.** A 28-year-old man recently presented with a 1-month history of palpable lump in the left posterior cervical region at October 2009. The lumps were increasing in size and were tender. He denied any fever, weight loss or night sweats. He has been following with diagnosis of lymphoblastic lymphoma which was in complete remission since April 1999.

On examination there were no abnormalities in the ears, oral cavity, oropharynx or larynx. In the left posterior cervical region lymph nodes could be palpated and the largest lymph node measured 2 cm in diameter. The rest of examination was normal. In laboratory examination of complete blood count and biochemistrical parameter there weren't any abnormality except slightly elevated lactate dehydrogenase. The erythrocyte sedimentation rate was 54 mm/h and C-reactive protein 15 mg/L. Neck ultrasonography (USG) revealed multiple nodal enlargements in left postrerior cervical region (the biggest one to be 20×11 mm). Neck, thorax, and abdominopelvic computerize tomographiic (CT) scan were done. Neck CT revealed multiple nodal enlargements in left posterior cervical region (the biggest one to be 20×10 mm). Thorax and abdominopelvic CT were normal.

A provisional diagnosis of relapsing lymphoma was suspected. Ultrasonographically guided needle biopsy was done.

On histopathologic examination, the lymph node architecture was effaced by paracortical expansion composed of necrotic areas with extensive apoptotic bodies, abundant karyorrhectic debris and numerous histiocytes (Fig. 1). Neutrophils and eosinophils were absent. Immunohistochemical examination

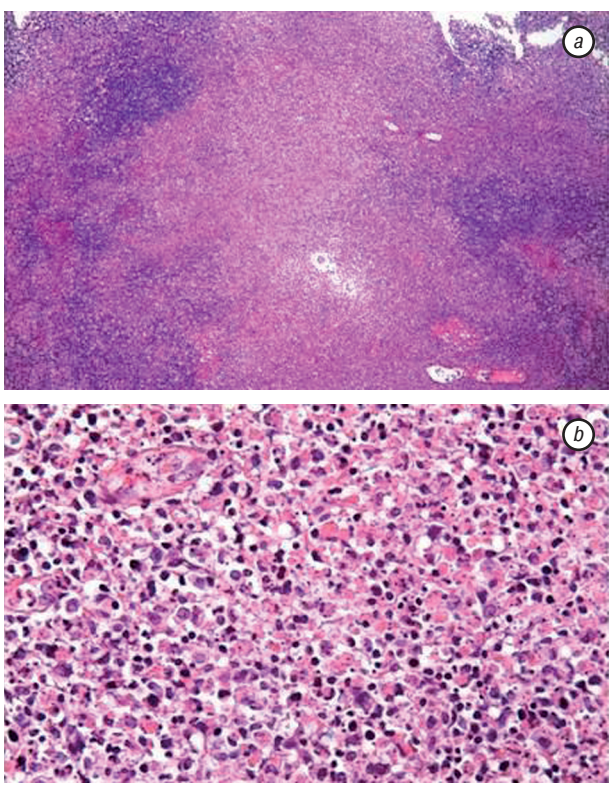

Fig. 1. Expansion of paracortical areas with necrosis (HE×4) (*a*). Necrotic areas with extensive apoptotic bodies, abundant karyorrhectic debris and numerous histiocytes (HE×40) (*b*)

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Abbreviation: KFD – Kikuchi-Fujimoto disease.

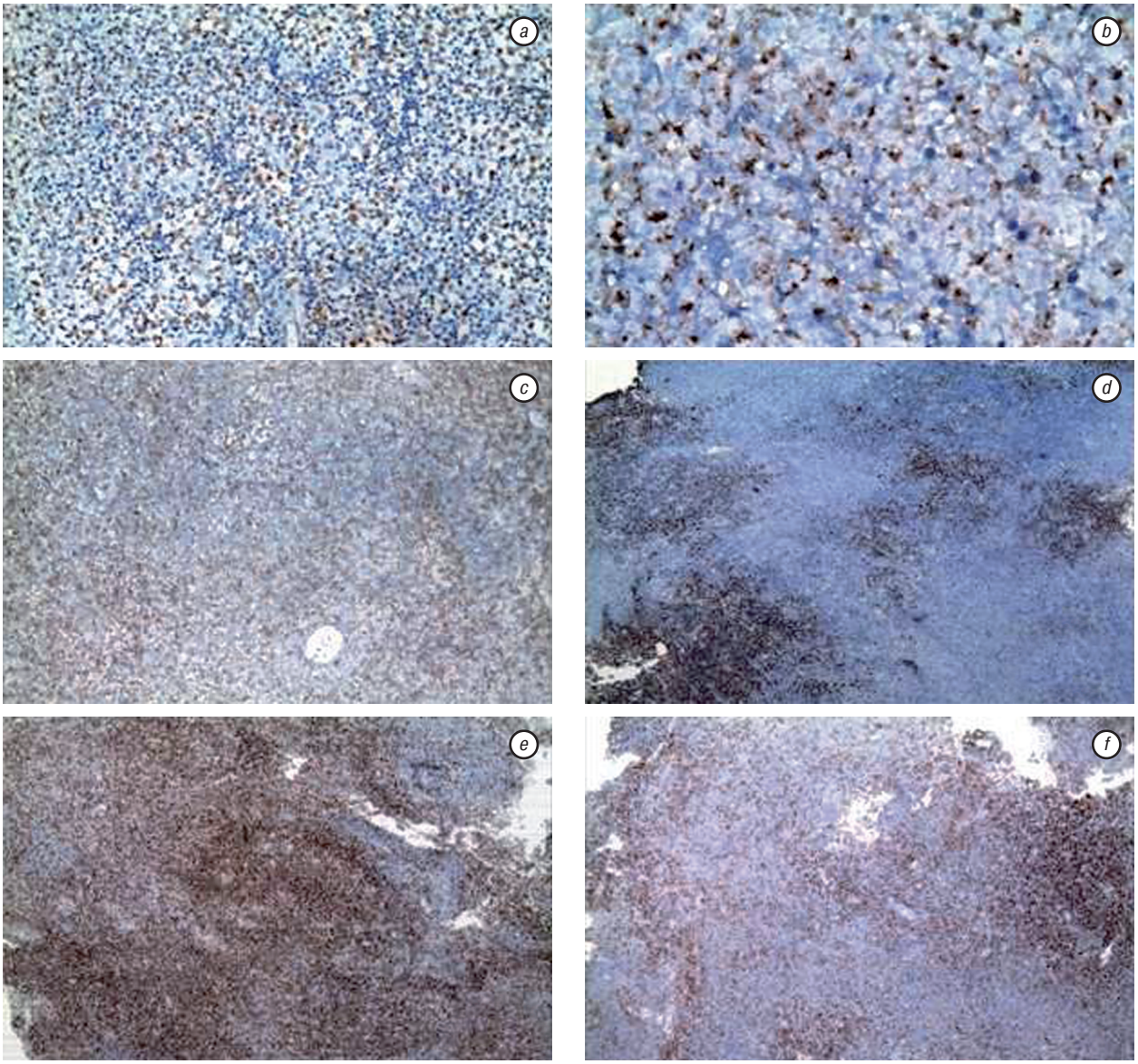

Fig. 2. CD68 and MPO positivity of histiocytic cells, respectively (*a*, *b*) (×20). CD3+ T cells predominated in the necrotic zones, in despite of very few CD20+ B cells (*c*, *d*) (×10, ×4). CD8 positivity of the majority of T cells and very few number of CD4+ T helper cells, scattered in the peripheral zones of necrotic areas (*e*, *f*) (×4)

revealed that histiocytic cells in the necrotic areas stained positively for antibodies against CD68 (Fig. 2, *a*) and myeloperoxidase (MPO) (Fig. 2, *b*). The lymphocytes in the involved zones were predominantly T cells (CD3+), whereas B cells (CD20+) were very few (Fig. 2, *c*, *d*). The majority of T cells were CD8+ cytotoxic cells and CD4+ T helper cells were scattered in the peripheral zones of necrotic areas (Fig. 2, *e*, *f*). Finally KFD (histiocytic necrotizing lymphadenitis) was diagnosed by the combination of histopathological and immunohistochemical findings.

After the diagnosis of KFD was made patient followed without any treatment. On review at 1 month, the patient's lymphadenopathy had almost resolved and at 3 months he was completely asymptomatic. All physical and laboratory examination were in normal limit.

Kikuchi's disease was first described independently in 1972 by Kikuchi and Fujimoto et al. The cause remains unclear although infections particularly with Epstein — Barr virus, paramyxovirus and Toxoplasma have been suggested as possible triggers. The differential diagnosis includes malignant lymphoma, systemic lupus erythematosus, metastatic carcinoma, AIDS and infectious mononucleosis [4].

The most common site of cervical lymphadenopathy is the posterior triangle and both sides of the neck can be involved. The involved lymph nodes are usually tender. Malaise, fever and leucopenia are common presenting symptoms. Less common symptoms include nausea, vomiting and diarrhea [1, 4].

An excisional lymph node biopsy and histological examination is the optimal method of ensuring an accurate diagnosis, as there are no specific laboratory tests available for KFD. This does, however, come with the proviso that there is close clinical liaison with the pathologist. Histological features that exclude malignancy include the absence of abnormal mitosis and the preservation of the sinusoidal pattern on intervening areas.

Most patients have spontaneous resolution of their symptoms within 6 months and treatment tends to be symptomatic. All patients require medical follow up as a small percentage can go to develop systemic lupus erythematosus, which can coincide or precede the Kikuchi's manifestations. As KFD is a generally benign self-limiting condition, it is important to establish the correct diagnosis quickly by biopsy and histopathology and it deserves more recognition among those specialists who are regularly referred patients with cervical lymphadenopathy.

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