Histiocytic Necrotizing Lymphadenitis (Kikuchi-Fujmoto's Disease) with Cutaneous Involvement

Sir,

Histiocytic necrotizing lymphadenitis (Kikuchi-Fujmoto's disease) was first described in 1972 as a benign lymphadenopathy with a self-limited clinical course. Later, this disease was frequently reported in Japan but rarely in Europe and in the United States. Its aetiology is unknown, possibly caused by a virus. The disease primarily affects cervical lymph nodes of young adult women, with fever and leukopenia. The histopathological features in the lymph nodes are peculiar: nodular areas with necrotizing foci in the cortex and paracortical areas of lymph node composed of foamy histiocytes and lymphoid cells, neutrophils being absent (1).

Cutaneous involvement is not frequent; when present, it is usually characterised by short and transient skin rashes (2). Some authors described other skin manifestations as papules (3, 4) or nodules (5) which showed similar histopathological features as the affected lesion in the lymph node.

An association with or a possible evolution of Kikuchi 's disease to systemic lupus erythematosus has been previously described (6). This kind of possible evolution in the skin of Kikuchi's disease should be considered in differential diagnosis with cutaneous lymphoid neoplasias, such as large cell lymphomas or Hodgkin's disease (2, 7).

A 21-year-old Italian woman came to our observation for a red-brown erythematous plaque measuring 2×1 cm, developed on the left cheek 1 year before with slow and progressive enlargement. In the past 6 years she had been admitted three times to the Medicine Department of our hospital for recurrent episodes of lymphadenopathy associated with systemic symptoms such as fever and leukopenia. Each episode lasted for 1 month; the previous two had been characterised by right cervical lymphadenopathy and interpreted as due to viral infection, according to serological positivity of Epstein-Barr virus antibodies. She went to the third admission because of left cervical lymphadenopathy, and a diagnosis of Kikuchi's disease was made, based on histopathological and immunohistochemical features of the lymph node biopsy.

Laboratory examinations, including serum titers for infective diseases, autoantibodies and bone marrow biopsy, showed leukopenia (white cell count of 2,510/cmm) but no other pathological alterations. The skin lesion which addressed the patient to our Department appeared shortly after the last episode of left cervical lymphadenopathy. No lymphadenomegaly and no systemic symptoms were present at the time of our observation, and haematological data were normal.

A total excision of the cutaneous lesion was performed, and the histological examination of the skin biopsy showed heavy lymphoid infiltration of the dermis and subcutaneous fat (Fig. 1). This lymphoid population did not show any epidermotropism and was mainly constituted by small round lymphocytes with scarce cytoplasm; scattered large mononuclear cells, with an irregularly shaped nucleus, could easily be seen. Among the lymphoid nodules, necrotic foci were present, surrounded by histiocytes with no granulocytes.

The immunohistochemical analysis of the lymphoid infiltrate failed to demonstrate any monoclonal populations. The mononuclear cells were positive for UCHL1/CD45 RO and MT1/CD43. In view of the morphological and immunophenotypical features, we consider this case as cutaneous involvement of Kikuchi's disease.

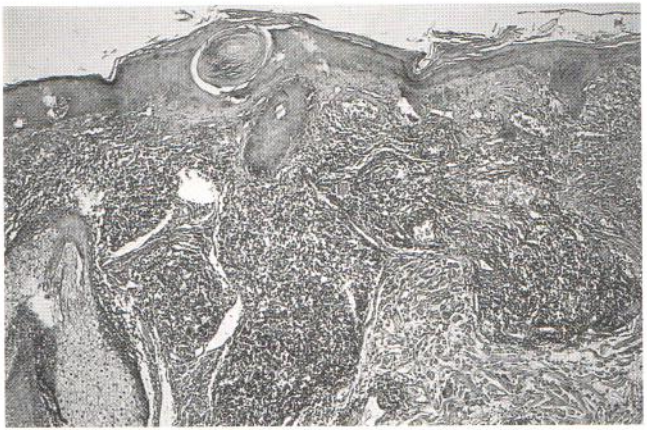

Fig. 1. Lymphoid infiltration with a nodular pattern of the dermis and subcutaneous fat. The lymphoid infiltrate did not show any epidermotropism. Among the lymphoid nodules, foci of steatonecrosis could be seen, with foamy histiocytes and no granulocytes. (E.E. 40 ×).

Skin rashes are frequently reported in patients with Kikuchi's disease, described as morbilliform, drug eruption-like, urticarial, maculopapular or disseminated erythema. The histogenesis and pathogenesis of these cutaneous manifestations are still unknown. The cutaneous involvement in a Caucasian patient with a skin tumour mass has previously been described (5). In that case no history of lymphadenopathy was reported, and, at present, it is difficult to make a diagnosis of Kikuchi's disease without nodal involvement.

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