Kikuchi–Fujimoto disease associated with polymyositis

Sir, I present the case of a 41-yr-old lady diagnosed with Kikuchi–Fujimoto disease (KFD) by clinical features and lymph node histology, who subsequently developed polymyositis with pulmonary involvement. There are no reports of this association in the literature.

A 41-yr-old female of Indian origin was admitted with a day’s history of periumbilical pain and 3 weeks of fever, general malaise, weight loss and painful lymphadenopathy. Her past history includes pulmonary tuberculosis. Physical examination revealed a fever of 38°C and tenderness in the right iliac fossa. Matted, tender, erythematous lymph nodes were present in the left axilla, and smaller lymph nodes in the right posterior triangle of the neck and right axilla. Laboratory findings revealed a lymphopenia of 0.9 × 10/l.

Further investigations showed normal plasma viscosity, chest X-ray and abdominal ultrasound. Blood cultures, sputum for TB culture and autoantibody screens were negative. Serology excluded active infection with
Epstein-Barr virus, cytomegalovirus and toxoplasma. Lymphocyte subsets showed reduced numbers of CD8 T cells and NK cells, but T-cell activation consistent with a reactive process. Swinging pyrexias with rigors continued, and a neutropenia of 1.61 × 10⁹/l and elevated plasma viscosity of 2.52 (normal range 1.5–1.72) developed. Lymph node biopsy revealed necrotizing lymphadenitis, as evidenced by reactive follicles and necrosis with extensive histiocytic infiltration, in the absence of giant cells and epithelioid granulomata. Culture for tuberculosis was negative. A diagnosis of KFD was made.

At the 3-month review, the fever has subsided and only a mild, normochromic, normocytic anaemia was present and plasma viscosity (PV) was 1.8. Annual follow-up was arranged because of the association of KFD with systemic lupus erythematosus (SLE).

Five months after initial presentation, she was readmitted with a 2-week history of polyarthralgia and a painful swollen left thigh. Other complaints were diffuse swelling of the limbs, anorexia and proximal muscle weakness. Physical examination revealed pyrexia (38°C), mild alopecia, proximal muscle weakness (Medical Research Council grade 4+/5–), fusiform swelling of the fingers and a large erythematous area over the anterior surface of the left thigh. Investigations showed creatine kinase (CK) 667 IU/l (normal range 25–200), lactate dehydrogenase 2063 IU/l (normal range 350–700), C-reactive protein 10.1 mg/dl (normal range 0–1), PV 1.77, albumin 33 g/l (normal range 35–55) and alkaline aminotransferase 110 IU/l (normal range 2–53). Full blood count, serum complement and postdate or coincide with the diagnosis of SLE. Patients immunoglobulins were normal, apart from a mildly elevated IgG concentration of 18.8 g/l (normal range 5–20). Anti-Ro and anti-Jo antibodies were present. Anti-cardiolipin antibody IgG titre was 26 IgG phospholipid units/ml (normal range 0–14).

Electromyography (EMG) showed polyphasic units and fibrillations of low amplitude and short duration. Muscle biopsy showed extensive inflammatory changes with necrosis, perifascicular atrophy plus perifibre and interstitial inflammatory infiltrate. Skin biopsy revealed superficial perivascular dermatitis with no evidence of vasculitis, and it was negative for immunofluorescence. Polymyositis was diagnosed, and the patient was commenced on oral prednisolone 60 mg once daily.

The myopathy did not improve and the patient developed interstitial lung disease, as confirmed by high resolution CT thorax and restrictive lung function tests. Treatment with i.v. cyclophosphamide and pulsed methylprednisolone was commenced, and was followed by gradual clinical improvement and reduction in CK to 2820 IU/l. Treatment with oral cyclophosphamide was complicated by severe immune thrombocytopenia, which responded to intravenous immunoglobulin. Alternative immunosuppression was commenced with cyclosporin and methotrexate. Unfortunately, the alveolitis worsened and she concurrently acquired pneumonia. Despite supportive care in our intensive care unit, multi-organ failure developed, resulting in her death.

KFD (or histiocytic necrotizing lymphadenitis) is a rare, self-limiting condition predominantly affecting young women and manifested by cervical lymphadenopathy and fever. It was first described by Kikuchi and Fujimoto in 1972 [1, 2] and is well known in the eastern hemisphere. More recently it has been documented in North America and Europe [3]. The cause is unknown, but it has been postulated that it is a self-limited autoimmune condition induced by a virus-transformed lymphocyte. Leukopenia, elevated hepatic transaminases and increased ESR are usually present in KFD [4].

‘Histiocytic necrotizing lymphadenitis’ describes the pathognomonic histological appearance of KFD, which is characterized by patchy necrosis (mainly paracortical) proliferation of histiocytes and immunoblasts, paucity of neutrophils and the absence of haematoxylin bodies [5]. Lymphomas and particularly lupus lymphadenitis can be mistaken for KFD histologically, but the absence of significant numbers of haematoyxin bodies, granulocytes and plasma cells favours the latter. Lymphadenopathy is a common clinical feature of both KFD and SLE. It is the presenting feature in 1.7% of cases of SLE and is present in >50% of patients with established disease [6].

In this case, the muscle biopsy, EMG findings and elevated CK strongly supported the diagnosis of myositis. There are no previous case reports of an association between polymyositis and KFD; however, several case reports have associated KFD and SLE and one an association with Still’s disease [7]. KFD can precede, postdate or coincide with the diagnosis of SLE. Patients diagnosed with it should be assessed for SLE and receive long term follow-up checking for the development of SLE; conversely, KFD should be ruled out if lymphadenopathy accompanies an SLE flare [8].

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Accepted 2 May 2000
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