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Urticaria in Waldenström's macroglobulinaemia[1]

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Waldenström's macroglobulinaemia is a chronic disease of unknown aetiology characterized by a proliferation of B cells and a monoclonal increase in serum levels of IgM. The original report of Waldenström (1944) has been followed by a number of case reports and reviews which document the classical features of anaemia, fatigue and a variable incidence of lymphadenopathy, hepatosplenomegaly and hyperviscosity syndrome. Other less common complications include pulmonary infiltration, renal disease and dermatological manifestations. We report here a patient who presented with urticaria.

Case report

Mr TVS, aged 61, a retired works manager, had had, since 1971, joint pains, tiredness and a recurrent, transient, itchy rash. Examination revealed a widespread urticarial and erythematous eruption. There were no other abnormal physical signs.

Investigations

The ESR was 74 mm in 1st hour (Westergren), Hb 11 g/dl, WBC 15 400/mm^3 with 3% metamyclocytes. Serum protein electrophoresis revealed an M-component and on immunoelectrophoresis this proved to be an IgM-kappa. The immunoglobulin levels were IgM 33 g/l, IgA 1.1 g/l, IgG 11 g/l. Free kappa chains were present in the urine. The serum viscosity was 1.94 (normal <1.83). Analytical ultracentrifugation showed a macroglobulinaemic pattern with 27S aggregates. A skin biopsy showed conspicuous perivascular infiltration by lymphocytes, histiocytes, neutrophils and erythrocytes, but no immunoglobulin or complement deposits were seen. Normal results were obtained for DAT, latex, autoantibodies, cryoglobulin, CH_{50} and bone marrow.

He was treated with intermittent prednisolone and melphalan, and his arthralgia and tiredness improved, but his rash was unaffected.

Discussion

Dermatological manifestations in Waldenström's macroglobulinaemia are unusual, but may include purpura and cold injury or Raynaud's phenomenon – when the IgM paraprotein is a cryoglobulin – amyloid, xanthomatosis and cutaneous deposits. Urticaria is uncommon. In one series of 45 patients only one had recurrent urticaria (Krajny & Pruzanski 1976). A possible mechanism for the urticaria is the deposition of circulating immune complexes, and evidence that this may occur is afforded by the occasional development of vasculitis and glomerulonephritis in Waldenström's macroglobulinaemia (Lin *et al.* 1973, Martelo *et al.* 1975). In the case reported here there was no evidence for circulating complexes. However,

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the large 27S aggregates in the serum could have activated various inflammatory pathways, with consequent complement activation and release of local permeability factors.

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Diagnostic difficulties in Whipple's disease[1]

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Whipple's disease is a multisystem disorder which may present with features remote from the gastrointestinal tract. We describe a patient in whom arthropathy, personality change and neurological involvement were the principal features. The diagnosis of Whipple's disease, previously suspected, was delayed by a negative small-bowel biopsy but when the gastrointestinal symptoms became manifest, a positive biopsy with all the characteristic features of the disease was obtained. Several biopsies may need to be taken before Whipple's disease can be excluded.

A man of 58 presented in 1975 to hospital with a 12-year history of migratory joint pains affecting knees, ankles and shoulders. He complained of pain and stiffness in the metacarpophalangeal and proximal interphalangeal joints of both hands and had noticed swelling of the left wrist. For eighteen months he had felt depressed and had lost 6 kg in weight over this time. He was otherwise fit and worked full-time as a sheet metal worker. In 1953 and 1955 he had been treated with carbimazole for thyrotoxicosis. He smoked 30 cigarettes a day and had a productive cough.

He was a thin man who looked older than his years. He weighed 51.5 kg, height 1.78 m. The only abnormal physical signs were restricted lumbar spine movements and synovial swelling over the dorsum of the left wrist. He was seen regularly in outpatients as a case of seronegative polyarthritis. The only abnormal laboratory test was a persistently elevated ESR, usually 50–100 mm in 1st hour (Westergren). The arthralgia, joint stiffness and occasional joint swelling responded well to non-steroidal anti-inflammatory drugs and no erosive changes were seen on serial radiographs. There was no weight loss between 1975 and 1976 nor any abdominal symptoms or diarrhoea.

In February 1976 he complained of increasing lethargy and depression and of impotence for the first time. As no endocrine or neurological cause for the impotence could be found, and as there had been marital disharmony over many years, this was considered probably psychogenic.

In November 1976 he was admitted to hospital after collapsing at home. He had become giddy and disorientated whilst returning from work and had noticed increasing weakness of his left arm and leg over one hour. On examination he had a left hemiplegia and a mild left seventh cranial nerve palsy; the other cranial nerves were normal. There were no other relevant signs and he was normotensive. An isotope brain scan showed an area of increased uptake in the right inferoposterior parietal region consistent with a vascular lesion; a CAT brain scan was normal, as was the cerebrospinal fluid. The ESR was elevated at 52 mm in 1st

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