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Case Report

Acute Transverse Myelitis and Primary Urticarial Vasculitis

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Abstract: We report the case of a 56-year-old man with severe normocomplementaemic primary urticarial vasculitis for 16 years. Nine and 11 years after the onset of the symptoms, he developed two severe neurological complications, seizure and transverse myelitis, that must be attributed to the vasculitis. Transverse myelitis has been reported in other systemic diseases, particularly lupus erythematosus, but this is the first case of transverse myelitis complicating urticarial vasculitis.

Keywords: Neurological disease; Transverse myelitis; Urticaria; Vasculitis

Introduction

Urticarial vasculitis (UV) is now considered to be a distinct entity characterised by urticaria-like lesions and systemic damage. We report the first case of transverse myelitis occurring during 16-year follow-up of normocomplementaemic urticarial vasculitis.

Case Report

The patient was a Caucasian man born in 1938. Since 1980, he had progressively suffered from unexplained attacks of fever lasting for less than 24 h, combined with arthralgias, myalgias and transient urticarial lesions. He was first admitted in 1982. An examination revealed no abnormalities, blood cultures were negative and laboratory tests revealed only a high erythrocyte sedimentation rate (89 mm/h) and an increased white cell count (12×10^9/l). Chest and bone X-rays were normal, as were immunological tests. A skin biopsy showed subacute dermatitis. He was initially diagnosed with rheumatoid arthritis or Still's disease and was treated with prednisolone (90 mg/day). Only a slight improvement was obtained and the patient decided to stop this therapy. From 1983 to 1989, he was treated with gold salts and non-steroidal anti-inflammatory drugs. The attacks became less frequent and the patient was able to lead almost a normal life. Laboratory tests always showed a mild leucocytosis (white cell count between 10 and 13×10^9/l).

In 1989, he presented with Jacksonian seizures. Computed tomography of the brain and cerebral arteriography were normal.

In August 1991, he was admitted again because of muscle weakness, bilateral lower extremity sensory loss and paraesthesias, loss of sphincter control and progressive paraplegia. An examination revealed an anaesthetic level of T12. Cutaneous lesions were present on the abdomen and trunk. The temperature was about 38°C. A lumbar puncture showed pleocytosis and an elevated albumin level (1g/l) in the cerebrospinal fluid. Cultures were negative for bacteria and viruses. Myelography showed no spinal compression or malformation. Magnetic resonance imaging (MRI) of the spinal cord (T2-weighted images) revealed abnormal high signal intensity within the cord, extended from T8 to T10, consistent with transverse myelitis. Antibodies to neurotropic viruses, Epstein-Barr virus, cytomegalovirus, mycoplasma, human immunodeficiency virus and human T-cell lymphotropic virus 1 were not detected. Serological tests for syphilis and Lyme disease were non-reactive. Visual, auditory and median somatosensory evoked potentials were normal. High-dose methylprednisolone (500 mg every 12 h) was administered. Only incomplete improvement of the neurological

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status was noted, and paresis and bladder dysfunction remained. Chrysotherapy had been stopped in April 1991.

In December 1991, the patient presented with a deep vein thrombosis of the left leg and a new seizure.

The patient was readmitted in September 1992, because of weight loss and recrudescence of fever, arthralgias and cutaneous lesions. On admission, the patient had a temperature between 38°C and 40°C. An examination revealed asthenia, paraparesis and urinary incontinence. Many urticarial lesions were noted, especially on the trunk, lower back and arms. When the fever was higher, these lesions became more prominent and arthralgias were present, but there was no synovitis or joint deformations. Livedo was present on the legs. Heart, chest, liver and spleen examinations were normal. Laboratory data showed an increased white cell count (12×10^9/l) and normal red cell count, platelet count and haemoglobin level. Routine chemistries, including creatine kinase and lactate dehydrogenase, were normal. Liver function tests and creatinine level were normal. No monoclonal gammopathy was found. Serological examination revealed no antinuclear antibodies or anti-DNA antibodies and a search for anti-RNP, anti-SSA, anti-SSB, anti-Sm, anti-Jo1 and ANCA was negative. Total haemolytic complement was increased. Serum C2, C3, C4 and C1 esterase inhibitor levels were normal. HLA typing was A2, A9, B5, B21. Serological tests for HIV, hepatitis B and C were negative. A search for antiphospholipid antibodies, including VDRL and anticardiolipin assay, was negative. A prolonged APTT (activated partial thromboplastin time) was noted only once but not confirmed. Circulating immune complex concentrations were normal and a search for cryoglobulins was negative. Bone scintigraphy and abdominal ultrasound did not show abnormalities. Brain MRI showed left temporal and frontal cortical atrophic areas, which suggested past ischaemic events. A biopsy of a typical urticarial lesion of the trunk was performed. It showed typical subacute leucocytoclastic vasculitis of the skin, with perivascular infiltrates of mononuclear cells, lymphocytes and polymorphonuclear leucocytes. Immunofluorescent studies on the skin were negative. So, the pathological diagnosis was urticarial vasculitis. The patient was then treated again with steroids (prednisone 120 mg/day). A decrease of urticarial flares, fever and arthralgias was obtained, but prednisone could not be tapered to less than 50 mg/day without clinical relapse. Slight improvement was noted when adding dapsone.

Discussion

Our case fulfills the criteria of UV, a syndrome characterised by recurrent episodes of persistent urticaria-like lesions, which differ histologically from urticaria because of vessel damage and leucocytoclastic vasculitis. Within the clinical spectrum of vasculitis [1] and considering the difficult classification of vasculitis which continues to be modified [2] and discussed [3], UV must be considered to be a non-infectious and hypersensitivity vasculitis, predominantly involving small blood vessels and with pathological features of leucocytoclasia. During UV, systemic involvement is not infrequent, including fever, arthralgias, myalgias, abdominal or chest pain, angiooedema, enlargement of the liver and spleen, pulmonary disease, renal disease, episcleritis, uveitis and central nervous system disease [4,5].

Our patient does not present criteria for other diseases that have been associated with UV, such as systemic lupus erythematosus (SLE), 'lupus-like' disease, mixed connective tissue disease (MCTD) and Sjögren's syndrome, or necrotising vasculitis such as polyarteritis nodosa or Wegener's granulomatosis [4]. He has neither hypocomplementaemia nor low C1q, C2, C3 or C4 levels. B51 antigen is not present [6]. There is no monoclonal gammopathy and no chronic viral infection, especially hepatitis B, C and Epstein-Barr virus. Classical criteria for primary antiphospholipid syndrome are not present [7].

Thus, we think that our patient must be considered as suffering from 'primary urticarial vasculitis', even though it should be recalled that, in some rare cases [8], this cutaneous vasculitis may constitute an 'ante' serological and clinical phase of SLE.

This patient now has a 16-year follow-up. The transverse myelitis occurred 11 years after the beginning of the disease. We feel that it must be attributed to the UV for several reasons. First, other aetiologies of transverse myelitis [9], including infection by virus, bacteria, parasite or fungus, post-vaccinal reaction and multiple sclerosis, have been ruled out. Secondly, this patient had presented with seizures, which are the main events frequently found in vasculitis syndromes. This complication has already been reported during UV, and the brain MRI scan of our patient showed abnormalities evocative of little foci of cerebral infarction, probably due to focal vasculitis. The effect of gold salts can be discussed, but to our knowledge no central nervous system disorder has been described with this drug.

Transverse myelitis is a classical complication of SLE, and it has also been described during MCTD, rheumatoid arthritis, anti-Jo1 syndrome and idiopathic hypereosinophilic syndrome. However, to our knowledge, this neurological event has never been described during primary UV. In SLE [10] and rheumatoid arthritis [11], transverse myelitis has been strongly associated with the presence of antiphospholipid antibodies. In our case, anticardiolipin antibodies were negative.

Normocomplementaemic UV is known to have a more benign clinical course than hypocomplementaemic UV, and there is no renal involvement [12]. Our case underlines that the clinical evolution of these patients over the years can be very disheartening, that severe neurological complications such as transverse myelitis can occur, and that treatment of this disease remains very problematical.

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