CASE REPORT

Coexisting HLA-B27 positive spondyloarthritis and polyarteritis nodosa

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Abstract

A 38 year old woman presented with wide-spread polyarteritis nodosa a few years after the onset of HLA-B27 positive spondyloarthritis. The concomitant coexistence of these two disorders suggests a possible association in this genetically susceptible subject.

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Ankylosing spondylitis is a relatively common disease with an overall incidence of 1–2% and affects twice as many men as women (male/female ratio 2:1).¹ It may be atypical and milder in women and hence difficult to diagnose.² Approximately 88% of patients with ankylosing spondylitis are HLA-B27 positive.³ Disseminated vasculitis occurs rarely in most of the connective tissue diseases and is seldom the cause of death. Vascular lesions, other than those observed in the aorta, are rare in ankylosing spondylitis.⁴ It is, however, even more rare to find acute disseminated vasculitis in patients with ankylosing spondylitis.

There is only one previously published case report of the coexistence of disseminated vasculitis with ankylosing spondylitis.⁵ We report here the case of a young woman with ankylosing spondylitis who later developed fatal disseminated vasculitis, a clinical state consistent with polyarteritis nodosa.

Case report

A 38 year old Syrian woman was seen in April 1984 with low back and spinal pain which had started nine years previously. At that time she also had an episode of iritis and mouth ulcers and was diagnosed as having seronegative spondyloarthritis. She was subsequently found to be positive for the HLA-B27 antigen. During the seven years of follow up, her arthritis remained intermittently active and affected the whole range of spinal movements. She also developed progressive arthritis of her hips and knees. In 1987 she had an intermittent fever, at times reaching 38.5°C, frequent headaches, Raynaud's phenomenon, transient blurring of her vision and, at times, transient weakness of her legs, and also vascular lesions of a lace pattern, 'livedo reticularis', on her legs. In 1988 she developed a sudden transient bilateral loss of vision, but subsequently regained full vision in her left eye and partial vision in the right;

fundus examination showed only a slightly ischaemic retina. A possible diagnosis of vasculitis was considered and investigated, and the results were normal or negative except for a high erythrocyte sedimentation rate. She was treated with corticosteroids and her condition improved.

In October 1988 she was admitted with a sudden onset of a cold, blue, and painful right foot. On admission she was afebrile, pale, and was in severe pain. Examination showed a cold ischaemic foot, pulsations in the right dorsalis pedis and posterior tibial arteries were absent but all other pulsations were present. Signs of inflammation were present in her hip and knee joints, including the sacroiliac region and the spine. A provisional diagnosis of either embolism or acute vasculitis, or both, leading to vascular occlusion was considered likely. She was treated on admission with heparin and dextran 40 to promote her microcirculation, in addition to treatment with pulses of methyl prednisolone and cyclophosphamide. Over the next 48 hours the foot became progressively ischaemic in spite of these active therapeutic measures to promote circulation, later necessitating an amputation below the knee. Unfortunately, on the sixth day after the operation she suddenly became acutely dyspnoeic and hypotensive. Active resuscitation measures were undertaken but these were unsuccessful and the patient died.

The results of investigations carried out showed haemoglobin 105 g/l, white cell count $19.5 \times 10^9/l$, predominantly neutrophils (85%), and an increased erythrocyte sedimentation rate of 95 mm in the first hour (Westergren), a normal serum chemistry, and normal urine analysis. An immunology screen showed a positive result for C reactive protein, a negative test for rheumatoid factor, antinuclear antibodies, antibodies to double stranded DNA, and other autoantibodies. Normal serum complement C3 and a slightly low C4 concentration (0.16 (0.2–0.5) g/l) were found. A normal coagulation profile and negative tests for lupus anticoagulant and anticardiolipin antibodies and a positive test for hepatitis B surface antigen were found. Cryoglobulins were not detected. A pelvic radiograph showed evidence of bilateral sacroiliitis and those of the spine showed a calcified interspinous ligament (fig 1). A non-invasive vascular study showed an absence of blood flow in the right posterior tibial and dorsalis pedis arteries, and some flow abnormalities were also detected in the left leg. An arch aortogram showed an irregularity of the

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Figure 1 Radiograph of the pelvis and spine showing bilateral sacroiliitis and interspinous ligament calcification.

proximal portion of the descending aorta and occlusion of the right tibial artery after its origin without any reconstitution of blood supply below. HLA antigens, which were typed previously, showed the presence of HLA-A2, A9, B27 and BW35, CW4, DR3, DRW52, and DQ1 antigens. A temporal artery biopsy sample and those obtained at surgery showed evidence of widespread inflammation at various stages with intense cellular infiltration in all layers, predominantly lymphocytes; a few plasma cells, macrophages, and eosinophils were also noted. Fibrinoid changes and internal laminal proliferation were also striking features, histological

Figure 2 Histology of an artery sample after the operation showing a marked internal proliferation of cells with subtotal occlusion of the lumen, focal clusters of mononuclear cells, plasma cells, and macrophages affecting all the layers.

characteristics highly suggestive of diffuse arterial disease—that is, polyarteritis nodosa (fig 2). An electrocardiogram and an echocardiogram were normal.

Discussion

There is only one previously published case report of polyarteritis nodosa associated with ankylosing spondylitis.⁵ It is not yet known whether this is a chance finding. In this patient, the criteria for the diagnosis of the two disorders were met.^{6,7} It is often difficult to make a diagnosis of either ankylosing spondylitis and polyarteritis nodosa in women because of their rarity and, further, the features often are atypical, as shown in this case. It is likely, however, that the onset of these two rheumatic diseases in this patient was concurrent even though the arthritis had been diagnosed first, suggesting that an arthritis-arteritis association is not random. It is noteworthy that the clinical features and laboratory findings strongly support a diagnosis of polyarteritis nodosa and ankylosing spondylitis. This case also illustrates the rarity of these two disorders in women.^{2,3} It is not yet possible to identify a specific genetic marker common to these two disorders as the available data are limited and incomplete. This led to the speculation that there may be an unknown common factor responsible for causing either ankylosing spondylitis alone or polyarteritis nodosa alone, or an association of the two diseases in susceptible subjects with different immunogenetic pre-determinations. This is the second case in which the coexistence of polyarteritis nodosa and ankylosing spondylitis has been found in an HLA-B27 positive patient. It is hoped that this case report will alert clinicians to the possible concomitant occurrence of rheumatic diseases, particularly of genetically determined disorders, and that it will stimulate further investigations into a possible association between these disorders.

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