

Case report

Aortic incompetence in HLA B27-positive juvenile arthritis

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SUMMARY The early onset of isolated aortic incompetence in a male child with HLA B27 and peripheral arthritis is reported. Acute anterior uveitis and lone aortic incompetence occurred at 1 and 9 months respectively after the development of the acute inflammatory arthritis. The uveitis resolved with local therapy and the arthritis remitted 10 months after the onset. There has been no recurrence of the arthritis after 10 years of close follow-up but the aortic incompetence has persisted, though it remains haemodynamically insignificant.

Isolated aortic incompetence as an extraarticular feature of ankylosing spondylitis in the adult is well recognised.¹ Adults with Reiter's syndrome may ultimately develop isolated aortic incompetence, and the association of spondylitis and Reiter's syndrome with a markedly increased frequency of occurrence of HLA B27 in these patients is well established.²⁻⁴ In children with peripheral arthritis who carry the B27 histocompatibility antigen isolated aortic incompetence is probably uncommon, though children with ankylosing spondylitis may occasionally develop lone aortic incompetence during long-term follow-up.⁵

Case history

A 15-year-old male presented in April 1969 with pain in the sole of his left forefoot of 6 months' duration. In May 1969 he developed a painful red eye 3 days after a bee sting to the back of his head. A diagnosis of acute anterior uveitis was made and he was admitted to hospital for 7 days. In September 1969 he developed synovitis of the left knee, pain in the metacarpophalangeal joint and proximal interphalangeal joint of his right index finger, and bilateral plantar fasciitis with soft-tissue swelling of both ankles.

When next seen in February 1970 he was asymptomatic, but a physical examination revealed a

decrecendo diastolic murmur at the lower left sternal edge. The electrocardiogram and chest x-ray were normal, but cardiac catheterisation and echocardiogram confirmed the presence of lone aortic incompetence in a tricuspid valve. Cardiac output was normal, and there was no evidence of cardiac failure. He has been asymptomatic for the past 10 years, though the murmur persists. His latest clinical examination and chest x-ray revealed minimal cardiomegaly with a normal electrocardiography. Musculoskeletal examination showed no abnormality, and x-rays of the sacroiliac and lumbar vertebrae were normal. Rheumatoid factor and antinuclear factor have been negative, but HLA typing was positive for the B27 locus.

Discussion

The association of HLA B27-positive arthritis, uveitis, and cardiac abnormalities is well recognised in the spondyloarthritides,^{2-4, 6, 7} though their frequency and temporal relationship differ.^{1, 2, 8} Our patient was seronegative for antinuclear factor and rheumatoid factor but was HLA B27-positive. Uveitis occurs with increased frequency in Reiter's disease, ankylosing spondylitis, and juvenile rheumatism. Approximately 1% of patients who carry the HLA B27 antigen will develop iridocyclitis,⁶ and approximately 50% of patients with acute painful uveitis will be HLA B27-positive.^{2, 7} The uveitis may precede the arthritis by several years, and approximately 30% of patients with ankylosing spondylitis or Reiter's disease will develop uveitis

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at some time in the disease course.^{2, 3} Two basic types of uveitis occur in juvenile rheumatism, an indolent destructive form which occurs predominantly in antinuclear-positive girls aged 5–10 years, and the painful type found in our patient, which occurs in boys aged 10–15 years, who are usually negative for antinuclear factor and rheumatoid factor.^{9, 10}

Disease of the aortic and mitral valves is associated with the seronegative spondyloarthritides; medial necrosis of the valve and dilatation of the valve ring are the predominant pathological feature.^{8, 11, 12} Symptoms of aortic root involvement occur in up to 4% of patients, but generally after prolonged disease activity, usually longer than 15 years. Necropsy reports, however, put the incidence of aortic involvement as much higher, about 25%.¹³ The majority of patients are thus asymptomatic, but a small group develop cardiomegaly with partial or complete heart block.⁸ The incidence of first-degree heart block may be as high as 1% in all patients with spondyloarthritis, the frequency of the conduction defects and the severity of the uveitis correlating with the severity of the peripheral joint involvement.^{8, 14}

Thus, despite several features consistent with the spondyloarthritides our patients' disease showed several inconsistencies. He never at any time had evidence clinically or on x-ray of sacroiliac disease; secondly, his joint symptoms remitted after 1 year; and, thirdly, he developed lone aortic incompetence within 1 year of diagnosis of his arthritis. Aortic incompetence is an extremely uncommon finding in children with juvenile rheumatism of the spondyloarthritis type and is likewise uncommon early in the course of the disease¹² (Ansell, personal communication). Unlike the patient reported by Stewart *et al.*¹² our patient is as yet asymptomatic from his cardiac disease, but he may go on to develop cardiac insufficiency as well as possibly active spondylitis.

Our case and the experience of Ansell and Stewart *et al.*¹² implies that a diligent search for valvular disease should be part of the routine assessment of children with juvenile rheumatism.

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