

CASE REPORT

HLA-B27 positive juvenile arthritis with cardiac involvement preceding sacroiliac joint changes

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Abstract

While cardiovascular disease develops in up to 50% of adult patients with ankylosing spondylitis, it is very uncommon in its juvenile counterpart. Regarding the early stage of the disease, before onset of sacroiliac joint changes, only two cases with aortic incompetence have been published while reports of mitral valve involvement are not available. A 13 year old boy is described with an HLA-B27 positive asymmetric oligoarthritis and enthesitis, without back pain or radiographic evidence of sacroiliitis. Echocardiography showed an echogenic structure measuring 8 × 11 × 20 mm at the fibrous continuity between the aortic and mitral valves extending through a false tendon into an echogenic thickened posterior papillary muscle, causing a grade II aortic and grade III mitral regurgitation. Short term corticosteroid and long term non-steroidal anti-inflammatory drug and disease modifying antirheumatic drug treatments efficiently controlled the symptoms. The cardiac findings remained unchanged during a follow up of 20 months. Careful cardiac evaluation appears to be mandatory even in these young patients.

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Keywords: juvenile ankylosing spondylitis; HLA-B27; aortic valve insufficiency; mitral valve insufficiency

Chronic inflammatory arthritis of the sacroiliac joints and less commonly the extrapelvic spine and peripheral joints, frequently associated with enthesitis, constitutes the disease entity ankylosing spondylitis.¹ Common features further include positive rheumatoid factor and antinuclear antibodies as well as a genetic basis with a high prevalence of the HLA-B27 marker and a family history of spondyloarthropathy.

The juvenile counterpart of ankylosing spondylitis differs from the adult form by its frequent involvement of the peripheral skeleton joints, usually as the first manifestation. At the onset of the disease, only a minority of children show symptoms of the lumbosacral spine or the sacroiliac joints.²⁻⁴ While cardiovascular disease is common in adults with ankylosing spondylitis, it is less common in the juvenile form and is

rarely observed in the early stage of the disease before the onset of sacroiliac joint changes.

Case report

We present the case of a 13 year old boy with a painful swelling of the left knee and pain of the left heel. He had a one year history of sustained pain and recurrent swelling of the left ankle and intermittent pain in the right hip. The upper extremities and the back were not involved and fever was never observed. There was no family history of arthritis or other connective tissue diseases.

PHYSICAL EXAMINATION

The left knee was swollen with tenderness and limitation of motion. The left heel was tender at the insertion of the Achilles tendon into the calcaneus. There was no tenderness, flattening, or rigidity of the lumbosacral spine. Chest expansion was normal and neurological symptoms or systemic signs such as fever, diarrhoea, urethritis, and iritis were absent.

Cardiac auscultation detected a grade 2/6 systolic murmur that was mesosystolic and with low frequency (vibrating) at the upper left sternal border, and holosystolic and with a high frequency (blowing) at the cardiac apex. Furthermore, there was a grade 1/6 holodiastolic decrescendo murmur at the lower left sternal border.

LABORATORY FINDINGS

Signs of active inflammation were an increased erythrocyte sedimentation rate of up to 114 mm/h and a maximal C reactive protein concentration of 32.5 mg/l but no leucocytosis. Mild microcytic anaemia (haemoglobin 1.70 mmol/l, haematocrit 0.326, mean corpuscular volume 77.9 fl) was associated with thrombocytosis (platelet count $571 \times 10^9/l$). The HLA-B27 marker tested positive, and the immunological markers rheumatoid factor and antinuclear antibodies were negative. As positive antistreptolysin O titres are very common in the Korean population, its occurrence in our patient was considered unspecific.

Signs of inflammation were also found in the synovial fluid (red cell count $864 \times 10^6/l$, white cell count $1.764 \times 10^9/l$, 60% polymorphonuclear neutrophil leucocytes, 35% lymphocytes) without evidence of a bacterial genesis (Gram stain and cultures were negative).

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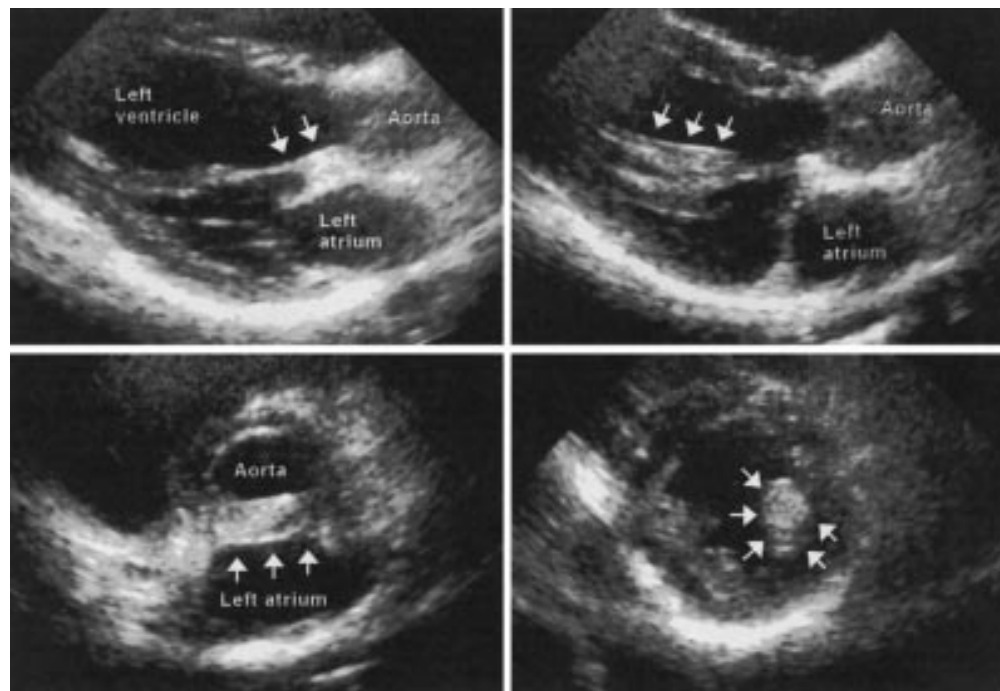


Figure 1 Echogenic $8 \times 11 \times 20$ mm structure between the aortic and mitral valves (arrows, parasternal long axis (upper left), parasternal short axis (lower left)) connected by a false tendon to the echogenic posterior papillary muscle (arrows, parasternal long axis (upper right), parasternal short axis (lower right)).

RADIOGRAPHIC EVALUATION

Radiographical (posteroanterior view) findings of both knees and the pelvis including the sacroiliac joints were normal. Active inflammation of the right hip joint was seen in a scintigraphic bone scan. Magnetic resonance imaging documented a pronounced joint effusion in the left knee with synovial enhancement and an abnormally enhanced bone marrow signal in the proximal tibia.

ECHOCARDIOGRAPHY

In the absence of pericardial or myocardial involvement, we found an echogenic structure measuring $8 \times 11 \times 20$ mm at the site of the fibrous continuity between the aortic and mitral valve (fig 1). This structure served as the origin of a left ventricular false tendon connecting to the posterior papillary muscle, which itself had a gross thickening with a diameter of 16 mm (fig 1). The false tendon explained the child's typical vibrating systolic murmur. Both the aortic and the mitral valve

were partially included in this echogenic structure, causing a grade II aortic and a grade I/II mitral regurgitation (fig 2).

TREATMENT

Short term low dose oral administration of a corticosteroid (prednisolone 0.5 mg/kg body weight, tapered over 12 weeks) led to quick relief of the symptoms. A parallel long term treatment with a combination of a disease modifying antirheumatic drug (sulfasalazine)^{5,6} and a non-steroidal anti-inflammatory drug (naproxen)⁷ was initiated. Single intra-articular injections of a corticosteroid (triamcinolone) effectively controlled recurrences of joint inflammation. Physical therapy included daily active bending and deep breathing exercises directed towards preventing loss of range and poor positioning in the spine and chest.

FOLLOW UP

The effectiveness of the treatment was shown by diminishing signs of inflammation in

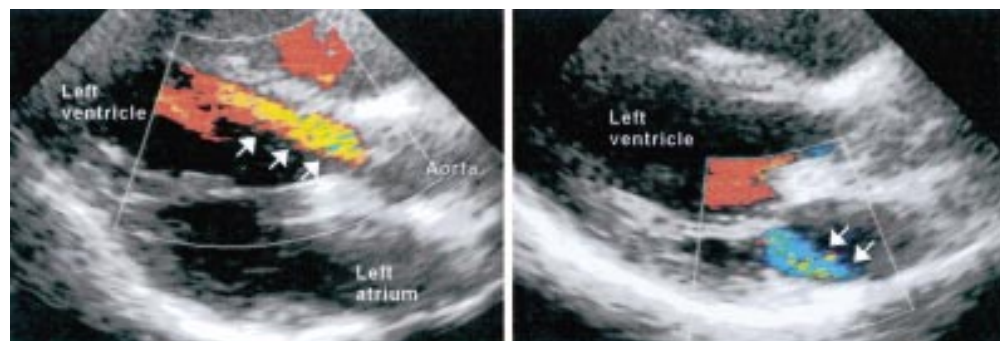


Figure 2 Colour Doppler echocardiography showing valve dysfunction: grade II aortic regurgitation (arrows, parasternal long axis (left)) and grade I/II mitral regurgitation (arrows, parasternal long axis (right)).

scintigraphy of the right pelvis. Two recurrences of painful swelling and inflammation of the left knee 7 and 15 months, respectively, after initiation of the oral treatment prompted acute local corticosteroid injections. The cardiac findings remained unchanged over the follow up of 20 months.

Discussion

The combination of pauciarticular arthritis of the lower extremities, typical enthesitis, positivity for HLA-B27, and negativity for rheumatoid factor and antinuclear antibodies in an adolescent boy is very suggestive of early juvenile ankylosing spondylitis, even without sacroiliitis.

Such patients, although most of them eventually develop sacroiliitis, do not meet the accepted diagnostic criteria for ankylosing spondylitis or juvenile ankylosing spondylitis at the onset of the disease. Therefore, descriptive terms such as “HLA-B27 positive juvenile arthritis” or “syndrome of seronegativity, enthesopathy, and arthropathy” (SEA syndrome)⁸ have been proposed and may be more appropriate.

Cardiovascular involvement is a common finding in adult patients with ankylosing spondylitis. However, it is considered rare in juvenile ankylosing spondylitis and only a small number of cases have been published with aortitis preceding the onset of sacroiliitis.^{9,10} Inflammation of the mitral valve is less common in adults, rare in juvenile ankylosing spondylitis, and not yet reported to precede the onset of sacroiliitis. The cardiac involvement in this case might also have been missed had a notable vibrating murmur of an aberrant left ventricular false tendon not alarmed the

involved clinician. In the present patient the false tendon might have promoted the development of inflammation and fibrous transformation of the involved structures by means of unusual shear and tension at both ends—the fibrous continuity between the aortic and mitral valves and the posterior papillary muscle.

Careful cardiac evaluation appears to be mandatory in all cases of HLA-B27 positive juvenile arthritis, even in young patients. Although the long term outcome is not yet clear in our case, control of the inflammation by systemic treatment with non-steroidal anti-inflammatory and disease modifying antirheumatic drugs seems to stop or at least slow down progression of the valvulitis.

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