



THERE IS MORE TO HLA B27 POSITIVITY THAN ONLY ARTHRITIS

Paediatrics

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ABSTRACT

Human Leukocyte Antigen (HLA) B27 associated arthritis is a distinct subtype of chronic arthritis characterized by genetic predisposition, arthritis with axial involvement, enthesitis and anterior uveitis. Extra-articular manifestations are not uncommon in this subgroup of patients with JIA. We report a 12-year-old boy who presented with bilateral knee and right hip joint arthritis for 3 months duration and HLA B27 positivity. On examination he had arthritis of bilateral knee joints and peripheral signs of aortic regurgitation. Two dimensional echocardiography revealed moderate aortic regurgitation. Although, cardiac abnormalities like valvular insufficiency, valvular incompetence and conduction defects are commonly reported in adults with prolonged disease course, but these are rarely seen in children and is exceptional at such a young age and so early in disease course. So baseline and regular CVS evaluation including echocardiography needs to be carried out in all young patients with HLA B27 associated arthritis.

KEYWORDS

Human Leukocyte Antigen B27; Arthritis; Aortic Regurgitation

INTRODUCTION

Juvenile Idiopathic arthritis (JIA) is a common chronic rheumatic disease in children (1). Human Leukocyte Antigen (HLA) B27 associated arthritis is a distinct subtype of chronic arthritis characterized by genetic predisposition, arthritis with axial involvement, enthesitis and anterior uveitis (2). The disease usually starts as an asymmetric lower limb arthritis in young children with involvement of knee, ankle and hip joints. Enthesitis is a prominent complaint but spondylitis usually does not occur until the late second decade (3). Extra-articular manifestations are not uncommon in this subgroup of patients with JIA (4). Although, cardiac abnormalities like valvular insufficiency, valvular incompetence and conduction defects are commonly reported in adults with prolonged disease course, but these are rarely seen in children (4,5). We report a child with HLA B27 associated arthritis who also had aortic regurgitation. This is distinctly unusual at such a young age.

CASE PRESENTATION

A 12-year-old boy presented with pain and swelling in both knee joints and pain in right hip for 3 months duration. Arthritis was additive without small joints involvement. There was no history of fever, rash and eye complaints. No family history of psoriasis or rheumatological disorder. On examination he had wide pulse pressure and dancing carotids. He had effusion with restricted range of movements in both knee joints and painful restriction of movement in right hip joint. He had hyperkinetic precordium with high pitched diastolic murmur in secondary aortic area and presence of pistol shot sounds. There was no stigmata or family history of psoriasis. On investigations, he had anaemia (haemoglobin 111 gm/L), elevated erythrocyte sedimentation rate (74 mm in 1st hour) and C-reactive protein (26 mg/L) and positive HLA B27 antigen. His antinuclear antibody (ANA) and rheumatoid factor was negative. Rest of the investigations were unremarkable. Two-dimensional echocardiography revealed isolated moderate aortic regurgitation with normal leaflets.

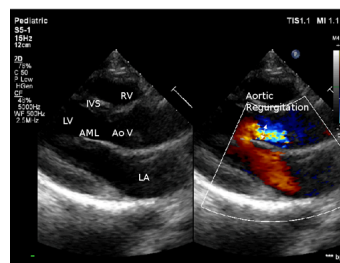


Figure 1: 2-D echocardiography with parasternal long axis view showing aortic regurgitation

He was initiated on bridge prednisolone therapy (1 mg/kg/day), oral naproxen (20 mg/kg/day) and oral sulfasalazine (30 mg/kg/day). He showed gradual improvement on follow-up.

DISCUSSION

Our patient is an adolescent boy with HLA B27 positivity, chronic asymmetric peripheral arthritis, aortic regurgitation and sero-negative for rheumatoid factor and ANA. A diagnosis of juvenile spondyloarthritis with cardiac involvement was proffered. While, cardiovascular system involvement is not uncommon in adults with longstanding disease, it is extremely uncommon in children (6–8). A large cohort study by Stamato et al documented prevalence of aortic regurgitation in 8% children with juvenile ankylosing spondylitis but with a mean age and mean disease duration of 14 and 4.3 years respectively (9). Aortic regurgitation in our case is exceptional at such a young age and so early in disease course. Aortic regurgitation in these patients occurs secondary to dilatation of the aortic root, fibrosis of the cusp bases and rolled edges of cusps (7).

CONCLUSION

Although, for long term outcome, our case is still to be followed up, but

control of the inflammation with oral non-steroidal anti-inflammatory drugs, disease modifying anti-rheumatic drugs like sulfasalazine and biologics like anti-tumor necrosis factor- α may slow down progression of the valvulitis (10). We conclude that baseline and regular CVS evaluation including echocardiography and color Doppler needs to be carried out in all young patients with HLA B27 associated arthritis.

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