CASE REPORT

Bilateral hip pain in a young man? It may be worth considering juvenile-onset ankylosing spondylitis (JAS)

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SUMMARY

A 15-year-old boy with severe bilateral hip joint pain and restriction of mobility presented to the casualty ward. He had earlier been treated for tuberculosis of the hip, with no relief. Our work up revealed a case of severe juvenile-onset ankylosing spondylitis with predominant hip involvement and accompanying sacroiliitis.

BACKGROUND

Ankylosing spondylitis (AS) is an inflammatory disorder mainly affecting the axial joints and distinguished by a significant association with HLA-B27. Juvenile-onset AS (JAS), that is, the condition having onset of symptoms in patients under 16 years of age, forms the majority of juvenile spondyloarthropathy (JSpA) cases. Because the individual differences in the clinical manifestations are large and sacroiliac joints of children are in the developmental stage, imaging may not be of much help and diagnosis may be delayed. JAS has a different phenotype and prognosis than adult-onset AS (AAS).

Owing to the predominance of tuberculosis in countries such as India, and owing to lack of awareness, such cases are started on a long duration of antituberculous therapy. This leads to an unnecessary delay in proper diagnosis and treatment.

CASE PRESENTATION

A 15-year-old boy, unable to walk due to severe pain in his hip joints, presented to the emergency ward. He had experienced this pain for 4 months. He also reported fever on and off for 4 months. He had no history of lower back pain or fractures. There was no history of lower limb weakness and no changes in bowel/bladder function. He was started empirically on antitubercular treatment for his fever by his general practitioner in a local hospital; this continued for 3 months. However, he got no relief. He had a history notable for Pulmonary Koch's 8 years prior, for which he had received a full course of antitubercular treatment (no documentation for this was available).

On examination, the patient was haemodynamically stable. General examination showed pallor and systemic examination was normal. Power could not be evaluated in the lower limbs due to severe pain, but sensory system examination was normal. Upper limb examination was normal. Reflexes were

normal and plantars were flexor. Spine examination was normal. Local examination revealed severe tenderness on movement in bilateral hip joints (right>left). Movement of other joints was fully preserved. Flexion and extension of lumbar and sacral spine did not elicit any pain. Upper limb movements were also fully preserved and normal.

INVESTIGATIONS

Routine laboratory investigations showed haemoglobin of 6 g%. White cell counts and platelets were normal. Other biochemical investigations (renal and liver function tests) were normal. Further evaluation was carried out for the anaemia. A peripheral smear showed normocytic normochromic anaemia. Ferritin level was 350 ng/mL (normal range 28–397). Vitamin B₁₂ level was 376 pg/mL (normal range 240–900). Erythrocyte sedimentation rate (ESR) was 92 mm at the end of 1 h.

X-ray of the pelvis with bilateral hip joints showed maintained joint space along with margins showing sclerosis with no bone erosion or destruction. These findings were suggestive of an inflammatory pathology.

MRI of bilateral hip and sacroiliac joints revealed alteration in marrow signal intensity in bilateral hips (right>left) involving the head of the right femur, adjacent acetabulum and iliac bone, left acetabulum, adjacent iliac bone, and sacral and iliac articular surfaces of bilateral sacroiliac joints. These findings were suggestive of inflammatory arthritis (figure 1).

HLA B27 was positive in this patient. He was diagnosed as a case of JAS. Echocardiography was normal. Ophthalmic evaluation was carried out and no evidence of uveitis was noted.

TREATMENT

The patient was given general symptomatic and supportive care. He was maintained on the continuation phase of antitubercular therapy, as he had been taking it for the past 3 months, in order to prevent resistance to the drugs. In consultation with the rheumatologist, a single intramuscular Depomedrol injection was given for the severe pain. Salazopyrine 500 mg once daily was started, with doses increasing to 1 g two times a day. A short (7-day) course of Indomethacin was started for pain relief.

Physiotherapy was subsequently given.

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Figure 1 MRI of both hips showing alteration in marrow signal intensity in bilateral hips (right>left), suggestive of inflammatory arthritis.

OUTCOME AND FOLLOW-UP

The patient's pain gradually settled. He improved clinically and began standing without support, followed gradually by walking.

DISCUSSION

JAS causes inflammation of the spine and large joints, resulting in stiffness and pain. It tends to run in families and males are affected three times more than females.

Differences between juvenile and adult onset disease:

JAS strikes young people, typically between ages 17 and 35 years, but it also may affect adolescents. Reported ages of onset were 12.8 ± 2.7 and 25.0 ± 7.4 years for JAS and AAS, respectively.¹

Patients with JAS were more likely to present with peripheral joint symptoms (46.6% vs 33.2%; $p < 0.001$). A greater percentage of patients with AAS presented with axial symptoms, low back pain and stiffness (71.5% vs 66.0%; $p = 0.043$).^{1 2} There was no difference between the two groups in the frequency of enthesopathic manifestations, iritis, or neck pain at presentation. Patients with JAS had a significantly longer delay in diagnosis compared with patients with AAS (15.3 ± 0.79 vs 7.6 ± 0.2 years; $p < 0.001$).

Extraspinal arthritis was more common than that reported in adults with JAS.^{1 2}

Treatment of juvenile-onset spondyloarthropathy (JSpA)³ can be frustrating. Although therapeutic measures are aimed at alleviating symptoms of inflammation (ie, pain), maintaining or improving range of motion and muscle strength, preventing deformity, preserving function and preventing or managing disease complications, short-term and long-term results can be very unsatisfactory. Early and continuous physical and occupational therapy are critical to the patient's maintenance of independent functioning. Non-steroidal anti-inflammatory drugs (NSAIDs) are the initial preferred pharmacological treatment for JAS. Sulfasalazine may be useful in JSpA, but differences from placebo may be only slightly significant.^{3 4} Short-term oral corticosteroid treatment at low to moderate doses may be required to control severe episodes of arthritis; higher doses may be needed for severe enthesitis. Intra-articular corticosteroid injection (triamcinolone hexacetonide, 1 mg/kg per joint)

may resolve persistent arthritis, enabling therapeutic intervention, and prevents the progression of disease.

Recently, attention has been given to the inflammatory processes underlying rheumatic conditions as potential sites for therapeutic intervention. For spondyloarthropathies, high levels of the pro-inflammatory cytokine tumour necrosis factor (TNF) α have been documented in synovial tissue. The anti-TNF α agents etanercept and infliximab⁴ have both been used in JSpA.

Tubercular hip arthritis results from haematogenous spread from pulmonary or other visceral or lymph node focus.⁵ Irregular and hazy joint margins, destruction of bone on either side of the joint, erosions and reduced joint space, are classical plain X-ray findings on tuberculosis of the hip. Our patient had no joint space reduction or bony erosions on X-ray. MRI further aided in the diagnosis by showing the presence of sacroiliitis.

In a developing country such as India, where there is a predominance of tuberculosis, any fever with loss of weight and raised ESR tends to be treated with antitubercular treatment by general practitioners and quacks. Hence awareness of the reported disease and timely treatment by general practitioners can help patients get well at the earliest and also avoids unnecessary long-term therapy in the form of antituberculous treatment. In our case, the patient originally presented exclusively with hip pain and movement restriction associated with systemic symptoms such as fever, weight loss and fatigue. He also had anaemia. In this clinical setting in a rural area, he had been, without detailed work up, started on antituberculous therapy. However, eventually, careful work up revealed that he had JAS. The patient responded to treatment and regular physiotherapy was advised.

Learning points

- Juvenile-onset ankylosing spondylitis has a different phenotype and prognosis than adult -onset ankylosing spondylitis.
- Misdiagnosis as tuberculosis is very common in countries such as India, due to constitutional symptoms of long-term low-grade fever with raised erythrocyte sedimentation rate.
- Continued medical education and awareness through reporting of such cases can help in spreading knowledge about the disease to general practitioners.

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Competing interests None declared.

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