CASE REPORT

Juvenile spondyloarthropathy: an important clinical lesson to remember

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SUMMARY

Spondyloarthropathy (SpA) is a group of inflammatory conditions that include spondylitis, sacroiliitis, asymmetrical peripheral arthritis and enthesitis. This condition is known as juvenile SpA when the diagnosis is made in patients up to 16 years of age. Enthesitis is a highly specific feature that occurs more often in juvenile SpA than in the adult form. In contrast to adult onset SpA, the initial manifestation of juvenile SpA rarely presents as inflammatory back pain. Peripheral arthritis is the more common presenting feature. We report a case of a 12-year-old boy who presented with a 1-year history of progressive low back pain, gluteal pain and thigh pain. There were no clinical symptoms of arthropathy of the distal extremities. MRI of the whole spine was performed twice, which, unfortunately, was unyielding. Finally, MRI of the sacroiliac joints revealed asymmetric sacroiliitis as well as enthesitis of the hips and pelvis. Further laboratory data showed negative rheumatoid factor and positive human leucocyte antigen (HLA) B27. A diagnosis of juvenile SpA with sacroiliitis and enthesitis was made. The imaging characteristics of juvenile SpA are highlighted.

BACKGROUND

Spondyloarthropathy (SpA) is a group of inflammatory conditions that includes spondylitis, sacroiliitis, asymmetrical peripheral arthritis and enthesitis. This condition is known as juvenile SpA when the diagnosis is made in those up to 16 years of age.^{1 2} The subcategories of juvenile SpA encompass seronegative enthesopathy and arthropathy syndrome, juvenile onset ankylosing spondylitis (AS), ankylosing tarsitis, human leucocyte antigen B27 (HLA-B27)-associated juvenile onset psoriatic arthritis, inflammatory bowel disease-related arthropathy and reactive arthritis.¹ In contrast to adult onset SpA, juvenile SpA rarely manifests as inflammatory back pain. Peripheral arthritis of the lower extremities is the more common presentation and enthesitis is a highly specific feature for juvenile SpA.²

CASE PRESENTATION

A 12-year-old boy was referred to our centre, due to worsening low back pain, and bilateral gluteal and upper thigh pain of 1-year duration. He attributed the pain to an episode of falling on his buttocks and landing on a hard surface. The pain was not completely relieved by oral analgesics. Subsequently, he walked with an antalgic gait and was frequently absent from school due to the pain.

He visited two medical centres to seek help. Despite the visits, the pain was not alleviated. An MRI of the spine was performed, but no abnormality was detected. The pain progressively increased in intensity for a subsequent 3 months. Finally, he was referred to our centre for further management. Questioning revealed no history of fever, morning stiffness, eye symptoms, pain of the small joints of the hands and feet, and no constitutional symptoms. He gave no history of altered bowel habits or urinary problems. He had no family history of similar illness.

On examination, the patient was pink and afebrile. No muscle wasting of the lower limbs, no rashes and no skin or nail changes were noted, and no lymphadenopathy was observed. Tenderness was elicited at the lower back, glutei and bilateral upper thighs. He walked with a waddling gait. The power of bilateral lower limbs was reduced to three-fifth and sensation was reduced from L1 to L3 dermatome levels.

INVESTIGATIONS

Laboratory data revealed mild hypochromic microcytic anaemia (11.9 g/dL), normal white cell count, creatine kinase (136 U/L), lactate dehydrogenase (329 U/L) and C reactive protein (0.94 mg/dL). MRI of the whole spine and brain was performed to rule out tumour and spinal nerve root impingement. There was no abnormality detected in the spine and brain. However, there was an abnormal hyperintense signal noted on limited view of both sacroiliac joints.

As a result, a dedicated MRI of the sacroiliac joints ([figure 1](#)) was performed, revealing bilateral asymmetrical sacroiliitis. Asymmetrical erosion, subchondral marrow oedema and synovitis were observed on both sacroiliac joints, indicating acute inflammation. Moreover, there was evidence of previous inflammation involving the proximal two-thirds of the left sacroiliac joint; this appeared as periarticular fat deposition, joint space widening, articular erosion and subchondral sclerosis ([figure 2](#)). Enthesitis was noted at the right anterior superior iliac spine (ASIS), bilateral ischial tuberosities and left greater trochanter. The imaging features were in favour of SpA with bilateral sacroiliitis and enthesitis. There was no synovitis of the hip joints. Based on the imaging findings, further physical examination revealed marked tenderness at the right ASIS, bilateral sacroiliac joints and left greater tuberosity.

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Figure 1 MRI of the sacroiliac joints. (A–C) Short τ inversion recovery coronal view showing bilateral asymmetrical periarticular hyperintense signal of the bone marrow (black arrows) indicating active sacroiliitis. At the entheses (white arrows), there is subadjacent marrow and peri-enthesial oedema involving the right anterior superior iliac spine, bilateral ischial tuberosities and left greater trochanter. (D) There is synovitis of the left sacroiliac joint (white arrow) and marrow enhancement of the right sacroiliac joint (black arrowhead).

Subsequent laboratory investigations displayed negative rheumatoid factor, raised erythrocyte sedimentation rate (112 mm/h), raised C3 and C4 complements (204 and 56.8 mg/dL) as well as negative antinuclear antibody (ANA). Hepatitis B surface antigen and hepatitis C serology were not detected. HLA-B27 was found to be positive. A final diagnosis of juvenile SpA with sacroiliitis and enthesitis was made. The patient was put on indomethacin and followed up regularly in rheumatology clinic.

DIFFERENTIAL DIAGNOSIS

- Behçet's disease has rarely been associated with juvenile onset sacroiliitis. Peripheral arthritis affecting the medium

and large joints is more common. Furthermore, this entity is extremely rare in the South East Asian population.

- Familial Mediterranean fever has also been associated with juvenile onset sacroiliitis, although its clinical presentation is quite different from what was seen in the patient in this case report. This entity is also hardly ever encountered in South East Asia.

OUTCOME AND FOLLOW-UP

On follow-up, the patient had no more back pain. However, he began to report knee pain. A continuum to adult SpA is expected in this child.

Figure 2 MRI of the sacroiliac joint on coronal view in T2-weighted (A) and T1-weighted images (B). There is joint space widening, articular erosion and subchondral sclerosis of the proximal third of the left sacroiliac joint (black arrow). Periarticular fat deposition (asterisks) is noted at the middle third of the left sacroiliac joint. The changes indicate areas of previous inflammation. Subchondral oedema and articular erosion are observed at the right sacroiliac joint (B).

DISCUSSION

Juvenile SpA is a subtype of SpA that is diagnosed in patients under 16 years of age. Children usually present with undifferentiated SpA that gradually progresses into a differentiated form of SpA.¹ Juvenile SpA will extend into adulthood. The hallmark of juvenile SpA is enthesitis and arthritis affecting the peripheral joints, particularly of the lower extremities.² IgM rheumatoid factor (IgM RF) and ANA are characteristically absent in serum.³

Sacroiliitis as the initial manifestation of adult SpA is common, but it is rare in juvenile SpA. Sacroiliitis with pelvic and hip enthesitis as the initial manifestations in our patient were atypical presentations. His symptoms were initially interpreted as spinal pathology with nerve impingement. He was once thought to be malingering as he refused to go to school despite no abnormality detected on MRI. In our centre, while assessing the sacroiliitis on MRI, florid enthesitis of the pelvis and hip was discovered. Meanwhile, he had an undifferentiated form of juvenile SpA that did not allow further subcategorisation of his condition. His condition did not fit juvenile AS either, due to the absence of relevant family history and the asymmetric distribution of sacroiliitis. However, this patient may progress to the adult form of AS in the future. We excluded infectious sacroiliitis as the cause of his presentation due to bilateral joint involvement with multiple areas of enthesitis. Juvenile idiopathic arthritis was not considered in view of negative RF and florid enthesitis being the dominant findings in this case. He had no synovitis of hip joints and distal extremities.

MRI has made a major contribution in early diagnosis of juvenile SpA. The early changes of enthesitis or arthritis are not detectable on plain radiograph. Entheses are the bony attachment sites of the tendon, ligament, fascia and joint capsule.⁴ Inflammation of the entheses is known as enthesitis. The MRI features of enthesitis include thickening and increased signal intensity of the tendon or ligament, peri-enthesal soft tissue oedema, subadjacent marrow oedema or erosion, adjacent joint swelling and bursal effusion.⁵ The hip joint is a commonly affected site for SpA. Early hip joint involvement is regarded as a marker for severe disease.⁶ The entheses of the hip comprise the muscle attachments to the lesser and greater trochanters as well as the pubic and ischial bones.⁵

Radiographic changes of sacroiliitis such as bone erosions, joint space alterations, subchondral sclerosis and ankylosis reflect structural damage rather than active inflammation, and can only be appreciated years after the onset of symptoms.⁷ The superior contrast resolution of soft tissue on MRI allows for proper assessment of disease activity. The severity of subchondral osteitis, joint erosion and effusion are well demonstrated on MRI. Hyperintense signal of the bone marrow on short τ inversion recovery (STIR) and contrast-enhanced T1-weighted fat-saturated images indicate active inflammation or osteitis.⁸ The degree of enhancement reflects the severity of active inflammation. Definite subchondral osteitis is highly suggestive of active sacroiliitis, which is a mandatory criterion in diagnosing SpA. The presence of synovitis, capsulitis or enthesitis without

accompanying subchondral osteitis is indicative of sacroiliitis, but not sufficient for making a diagnosis of active sacroiliitis.⁹ Findings suggestive of chronic or previous inflammation of the sacroiliac joint are periarticular fat deposition, joint space widening, articular erosion and subchondral sclerosis.

In conclusion, a high index of suspicion is required to diagnose inflammatory sacroiliitis owing to the vague clinical presentation of this patient. Although uncommon, sacroiliitis needs to be considered in a child with significant history of low back and gluteal pain. Early disease recognition with the start of treatment prevents permanent functional disability. The MRI scanning needs to be extended to the sacroiliac joints if the MRI of the lumbosacral spine is normal. STIR sequence of the sacroiliac joints on coronal view is adequate to exclude pathology of the sacroiliac joint by assessing the bone marrow signal intensity.

Learning points

- ▶ Sacroiliitis needs to be considered in a child with significant history of low back and gluteal pain.
- ▶ A high index of suspicion is required to diagnose inflammatory sacroiliitis, as the clinical presentation can be non-specific.
- ▶ The MRI scanning needs to be extended to the sacroiliac joints if the lumbosacral spine image is normal.

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