CASE REPORT

Ewing's sarcoma of the pelvis: an unusual, but not to be missed, cause of an irritable hip

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SUMMARY

A 15-year-old girl presented with a 2-month history of non-specific right hip pain associated with pain in the back, right flank and foot. Her symptoms deteriorated, interfering with weight-bearing. Following admission, she was found to be febrile with a flexion deformity of her right-hip, and tenderness over the iliac crest and posterior pelvis. A markedly elevated C reactive protein and erythrocyte sedimentation rate, with an ultrasound-proven scan effusion within the right hip were all suggestive of septic arthritis. However, full blood count demonstrated a significant anaemia, which together with the tenderness around the pelvis was not in keeping with this diagnosis. Surgical washout was therefore delayed to obtain a MRI scan of the pelvis. The scan revealed a 5×5×3 cm necrotic soft tissue mass within the gluteal muscles, arising from the right ilium, which biopsy confirmed to be a Ewing's sarcoma.

BACKGROUND

Irritable hip is a common presentation in children, in whom the clinical priority for the orthopaedic surgeon is the timely identification and management of septic arthritis. The most common cause for acute irritable hip is transient synovitis, and it can be difficult for the clinician to distinguish between this condition and septic arthritis of the hip. Both conditions present acutely with pain, difficulty in weight-bearing and fever. Transient synovitis is usually preceded by viral infection and is self-limiting. On the other hand, septic arthritis is a surgical emergency, and early operative drainage and IV antibiotics are necessary, in order to prevent loss of function and destruction of the bony joint.¹

Kocher *et al*² identified four clinical predictors to classify patients with septic arthritis with a 99.6% prediction rate: fever, pain on weight-bearing, white cell count (WCC) $\geq 12.0 \times 10^9$ cells/L and erythrocyte sedimentation rate (ESR) ≥ 40 mm/h. In 2006, Caird *et al*³ suggested that C reactive protein (CRP) is a strong independent predictor of septic arthritis and, in 2010, Sultan *et al*⁴ added CRP ≥ 20 mg/L as an additional predictor.

However, clinicians should be aware that while very rare, tumours can present as an irritable hip, with examination and investigation findings that can be very similar to septic arthritis of the hip, as this case demonstrates. Unless the clinician has an index of suspicion and detailed imaging investigation is performed, these cases could easily be missed and mismanaged.

CASE PRESENTATION

A 15-year-old girl presented with a 2-month history of non-specific pain affecting her right hip, with

some pain in her back, right flank and right foot. In the 2 weeks preceding her admission, the pain had worsened, making it very difficult for her to walk. There was no history of trauma or other significant medical history. She had consulted her family physician, who suspected that this may be juvenile idiopathic arthritis, and so prescribed diclofenac and naproxen, and referred her for an outpatient appointment with a paediatric rheumatologist. Blood tests at that point demonstrated an ESR of 77 mm/h, a WCC of 11.30×10^9 cells/L, raised platelet count of 538×10^9 cells/L and CRP of 30 mg/L.

Owing to the increasing pains, the patient attended the children's emergency department at the hospital, and was admitted for further investigation under the care of the paediatricians. Twenty-four hours after admission, she developed a fever of 38.3°C.

On examination, she looked generally unwell, but comfortable at rest. Her abdomen was soft but distended, with some tenderness on palpation in the right upper quadrant. There was no guarding, rebound tenderness or organomegaly.

She was tender along the right iliac crest, and the posterior pelvis in the region of the posterior superior iliac spine. She also had some tenderness in the lower lumbar spine. Thomas' test demonstrated that she had a fixed flexion deformity of her right hip. There were no changes in skin colour and no palpable masses in the right lower limb. There was no evidence of any fluctuant collections. Although there was limited active movement, the hip was passively flexed to 90° with some discomfort. Internal and external rotation was managed to 30°. There were no knee or ankle deformities and there were no external localising signs of an infection anywhere in the lower limb.

On admission, blood tests demonstrated that WCC had increased to 12×10^9 cells/L, and the haemoglobin level was noted to be 79 g/L. The CRP level had increased to 245 mg/L and ESR to 124 mm/h; this marked rise from the tests performed a week prior suggested some underlying severe acute process. With the fever and reduced range of movement at the hip, infection was suspected, either within the hip joint, or, given the tenderness around the pelvis, an osteomyelitis of the pelvic bones.

A MRI scan was requested, but due to logistical restraints, our radiology department performed an ultrasound scan of abdomen, pelvis and hip. No abnormalities were found within the abdomen or pelvis, but an effusion was found within the right hip, which was aspirated under ultrasound

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guidance. Turbid fluid of 2–3 mL was obtained from the hip; Gram stain and microscopy of this fluid demonstrated ++ white cells, but no organisms.

There was strong evidence to suggest that this patient had an acute septic arthritis of the hip joint, which would require emergent surgical washout. However, the tenderness around the ilium and sacrum were not in keeping with this diagnosis, and the anaemia was also unusual. The case was again discussed with the radiology department and the need for more detailed imaging of the pelvis reiterated. The scan was booked for the following day, and so surgical washout of the hip was deferred pending this result. The patient was, however, started on intravenous ceftriaxone.

The MRI scan was performed the next day, and showed a 5×5×3 cm necrotic soft tissue mass within the right gluteal muscles, arising from the posterior aspect of the right ilium. There was diffusely abnormal bone marrow signal throughout the sacrum and pelvis bilaterally, in keeping with marrow infiltration. The widespread bone marrow signal abnormality affected almost all the vertebral bodies, stretching to C2. There was necrosis within the right sacral ala. These findings could be visualised in a coronal view (figure 1) and a transverse view (figure 2). Biopsy later confirmed this to be Ewing's sarcoma.

INVESTIGATIONS

The main differentials in a case of irritable hip, as mentioned above, are transient synovitis or septic arthritis. Fever, pain on weight bearing and raised inflammatory markers have been shown by a number of authors to be useful in differentiating between these two conditions.^{2 3 5}

It has previously been suggested in the literature that in paediatric septic hip arthritis, ultrasound is sufficient for diagnosis and advanced imaging with MRI is not necessary.⁶ While ultrasound is a good way to look for fluid within the hip joint, it can fail to demonstrate pathology within bone, such as osteomyelitis.

Although the history, examination and ultrasound scan were all compatible with the diagnosis of septic hip, the pelvic tenderness and anaemia were not. We felt that MRI was the only imaging modality that could provide detailed enough imaging of the area to exclude pathology such as osteomyelitis or malignancy.

Figure 1 T2 coronal MRI of the pelvis with contrast.

Figure 2 T2 transverse MRI of the pelvis with contrast.

Supporting this view, White *et al*⁷ reported a prospective study of 50 children presenting with acute non-traumatic hip pain. They found that MRI correctly identified all patients who had a serious pathology, compared to ultrasonography, which only correctly identified this in 28%. The authors suggested that, where appropriate, MRI should be the imaging modality of choice in this presenting complaint.

DIFFERENTIAL DIAGNOSIS

This was essentially a case of an irritable hip, the most common cause of which is a transient synovitis, which is a diagnosis of exclusion. Transient synovitis was an unlikely diagnosis here as symptoms were ongoing for 2 months and, typically, this condition occurs in a younger age group (between 3 and 10 years). The patient was also pyrexial, with a high WCC, ESR and CRP, and had pain on weight-bearing, which made septic arthritis (infection of the joint) more likely.^{2 3 5} Less common, but another important cause of irritable hip, is infection within the bone (osteomyelitis). The chronicity of the symptoms and their insidious onset were not typical for septic arthritis. However, this could be accounted for by acute-on-chronic osteomyelitis. As the metaphysis of the proximal femur is intracapsular, it is even possible for an osteomyelitis to lead to a septic arthritis.⁸

With the abdominal tenderness, and the comparative restriction in active hip flexion compared to passive hip flexion, another differential diagnosis was psoas abscess (particularly given the lumbar tenderness). The abdominal and pelvic ultrasound excluded this diagnosis for us.

In hip pain, trauma is a differential that should always be asked about—it was negative in this patient's history. From the systematic viewpoint, a single joint effusion, if not accompanied by signs and symptoms of septic arthritis (high CRP, raised temperature, strong pain, etc), should also raise suspicion of Lyme disease.⁹ Although not present in this case, a history of preceding diarrhoea would incorporate reactive arthritis due to *Yersinia* into the differential diagnosis.¹⁰

Another possible differential diagnosis was juvenile idiopathic arthritis; however, this was much less likely with the monoarticular presentation of the hip joint at this age. Furthermore, one would not expect to find areas of bony tenderness.¹¹

In the face of chronic pain without obvious aetiology, malignancy must always be considered, especially in this case, where there was a history of night pain, and anaemia. It is important to note that Ewing's sarcoma, which itself is a particularly rare malignant cause, is also known to sometimes occur in the form of a bone and/or joint infection.¹²

With an ongoing history of 2 months, the MRI was pivotal in proving or disproving the presence of infection or neoplasia within or around the bone.

TREATMENT

The patient was transferred to a specialist oncology centre, where she was treated with the Euro-EWING 99 protocol (six cycles of vincristine, ifosfamide, doxorubicin and etoposide; and one cycle of vincristine, dactinomycin and ifosfamide).¹³ She was also given radiotherapy to the thoracic spine and iliac crest, and underwent an autologous transplantation of busulfan/melphalan conditioned peripheral blood stem cells.

OUTCOME AND FOLLOW-UP

Unfortunately, the disease progressed despite treatment, and the patient passed away about 1 year after diagnosis.

DISCUSSION

This case highlights the need for clinical acumen and timely access to detailed imaging investigations when dealing with cases of irritable hip.

Several authors have devised predictive algorithms for differentiating between transient synovitis and septic arthritis of the hip, based on clinical and laboratory features.²⁻³ They are commonly used in clinical practice, and the authors believe they can be very useful in the appropriate circumstances. However, it is clinical judgement that determines whether it is appropriate to use these algorithms, and they should be used with caution.⁴

In this case, Kocher *et al*'s predictive algorithm² suggests that there was an 83% chance of this case being a septic arthritis. Caird *et al*'s algorithm³ (which includes CRP) calculates the probability of this being 93%. This, together with the fluid seen within the hip joint on the ultrasound scan, would have been sufficient for many surgeons to have proceeded with surgery to wash out the hip joint. This could have led to significant adverse outcomes in this particular case, as the surgical approach would have been very difficult and could have seeded the tumour.

It was clinical acumen—allowing the recognition that some of the clinical features and the anaemia were not consistent with septic arthritis—that was important in preventing unnecessary surgery and making the correct diagnosis in a timely manner.

This case also highlights the importance of ease of access to MRI for investigating patients with this presentation. In the case presented, surgical washout was delayed by about 24 h in order to obtain the MRI; this was subsequently proven to be the right course of action, but if no other pathology had been found, this delay could have adversely affected the surgical outcome.¹⁴

There are only a few reports of malignancy presenting in children with an irritable hip, and no previous reports of Ewing's sarcoma presenting in such a manner. There have been two case reports and one case series of neuroblastomas.¹⁵⁻¹⁷ In the case series, it was noted that many cases were initially misdiagnosed as septic arthritis. All three reports highlight the importance of anaemia in differentiating malignancy over infection.

Tuten *et al*¹⁸ reported nine paediatric patients in whom leukaemia presented with a limp due to irritable hip or knee, associated with a raised temperature and anaemia.

Learning points

- Predictive algorithms for differentiating septic hip from transient synovitis exist, but clinical acumen is of paramount importance, particularly in deciding when it is appropriate to use these predictive tools.
- Malignancy is a rare cause for an irritable hip, but one that must be considered, especially in the presence of anaemia.
- MRI has an important role in the investigation of selected cases of irritable hip and, when required, it should be available promptly, since ultrasonography has its limitations.

Contributors APS was the consultant leading the case of the patient. PR is a junior doctor and JG is a medical student. All three authors contributed to the writing of this case report.

Competing interests None declared.

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