Ewing's Sarcoma Presenting as Hip Monoarthritis in a Spondyloarthritis Patient

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Abstract: Seronegative spondyloarthritis (SpA) and Ewing's sarcoma (ES) are common disorders affecting young males. A great challenge is to diagnose both diseases in the same patient. Here we describe a case of a young male patient having SpA, who developed ES that presented as monoarthritis of left hip which was misdiagnosed twice; first as peripheral manifestation of SpA and second as septic arthritis; before the correct diagnosis was reached.

A 17 year-old boy presented with inflammatory low back pain of gradual onset lasting for 4 months and knee arthralgias. Examination revealed the presence of localized tenderness over both SIJs and limited lumbar flexion. Plain x-ray showed bilateral grade 2 sacroiliitis and he was Rheumatoid factor negative. The diagnosis of seronegative SpA was made based on fulfillment of the Association of Spondyloarthritis International Society classification criteria for axial SpA. Nonsteroidal anti-inflammatory drugs (NSAIDs) and sulphasalazine were prescribed resulting in rapid improvement of back pain and knee arthralgia. Four months later, the patient had increased severity of his back pain with pain at the left hip region associated with painful limited internal hip rotation. The diagnosis of left hip arthritis associated with SpA was made without requesting imaging study at that time. The dose of NSAIDs was increased and a low dose steroid was added.

Few days later, the general condition of the patient deteriorated with the development of night fever, poor night sleep and poor appetite. On presentation, the patient looked very ill with fever. Hip examination revealed marked painful limited hip movement with flexion deformity. New investigations revealed an ESR of 110 mm/hr, and a CRP of 88 mg/L. There was normochromic normocytic anemia (HB: 9g/dl) and leucocytosis (WBC: 13.300/µL). The diagnosis initially considered was acute septic arthritis. Left hip arthrocentesis failed to aspirate any fluid and ultrasound revealed the absence of joint effusion. A multislice computerized tomography (CT) revealed the presence of a destructive osseous lesion targeting the left pubic bone and the left acetabulum with intra-articular extension into the left hip joint causing bony erosion of the medial aspect of the left femoral head. An MRI showed intra-pelvic extension with enlarged iliac lymph nodes. A biopsy was obtained from the mass. Histopathology revealed a malignant round-cell tumor consistent with ES. The patient was referred for chemotherapy and we knew that he was started on a combination of drugs for a short time before he died. To the best of our knowledge this is the first report in literature of ES in SpA patient and the second report of ES presenting as hip monoarthritis.

Keywords: Ewing's sarcoma, spondyloarthritis, arthritis.

INTRODUCTION

Seronegative spondyloarthritis (SpA) and Ewing's sarcoma (ES) are common disorders affecting young males. The hallmark of SpA is the presence of sacroiliitis causing inflammatory low back pain, and enthesitis causing localized tenderness over tendon insertions. The hip joint is the most commonly affected peripheral joint in SpA. ES involves bones of the pelvis and lower limb. It is the second malignant bone tumor in children and adolescents.

As both SpA and ES affect young males, occurrence of both in the same patient may lead to delayed diagnosis and treatment since manifestations resulting from ES of the pelvis can be mistaken as the musculoskeletal involvement caused by SpA. A great challenge is to diagnose both diseases in the same patient. Here we describe the first case of a young male patient having SpA, who developed ES that presented as monoarthritis of his left hip which was mistaken at first to be due to peripheral manifestations of SpA and then to be septic arthritis.

CASE PRESENTATION

A 17 year-old boy presented with mild low back pain of gradual onset lasting for 4 months. Pain was worse at night and with rest (typical inflammatory character). He had knee arthralgia for the last 2 weeks, but with no other joint complaints. He denied having any complaints related to the eyes, skin, gastrointestinal or

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urinary tracts. His sister was known to have ulcerative colitis and sacroiliitis (enteropathic SpA). Examination revealed the presence of localized tenderness over both SIJs and limited lumbar flexion (Schober test was 4cm). Meanwhile, chest expansion was normal. Peripheral joints were clinically free. Lab investigations revealed mild normocytic anemia (Hb:12.4g/dl, WBC:4.700/µL, platelets:260,000/µL), ESR:25mm/hr, CRP:5mg/L and Rheumatoid factor was negative. Plain x-ray showed bilateral grade 2 sacroiliitis.

The diagnosis of seronegative SpA was made based on fulfillment of the Association of Spondyloarthritis International Society (ASAS) classification criteria for axial SpA [1]. Nonsteroidal anti-inflammatory drugs (NSAIDs) and sulphasalazine (2gm/day) were prescribed which resulted in rapid improvement of back pain and knee arthralgia.

Four months later, through a follow-up phone call; the patient reported that his back pain started to increase in intensity with pain at the left hip region associated with limitation of the hip movements. A provisional diagnosis of left hip arthritis was made based on clinical situation without requesting a new pelvic imaging at that time. Accordingly, an increase in NSAID dose in addition to low dose steroids (10mg/day) were advised.

Few days later, no improvement was reported and the patient started to complain of deterioration of his general condition with the development of night fever, poor night sleep because of the severe pain. On presentation, the patient looked very ill, he had a muddy complexion and was in agony. His temperature was 39.5ºC, other vital signs were normal. There was flexion deformity of the left hip with inability to bear weight. Peripheral joint examination was normal except for the left hip, which showed marked painful limitation of all ranges of movement, both active and passive. New investigations were requested and revealed markedly elevated acute phase reactants; ESR of 110 mm/hr, CRP: 88 mg/L. Complete blood count showed normochromic normocytic anemia (HB: 9g/dl), leukocytosis (WBC: 13.300/µL) and normal platelets count (270,000/µL). The diagnosis initially considered was acute septic arthritis of the left hip. There was no history of trauma, or inherited coagulopathy or intake of anticoagulants. An immediate trial of blind left hip arthrocentesis was made but failed to aspirate any fluid. The patient was sent for ultrasound guided aspiration, but unexpectedly the scan revealed the absence of joint effusion.

Consequently, a multislice computerized tomography (CT) of the left hip was urgently requested. CT reported the presence of a destructive osseous lesion targeting the left pubic bone and the left acetabulum with intra-articular extension into the left hip joint causing bony erosion of the medial aspect of the left femoral head Figures **1-4**.

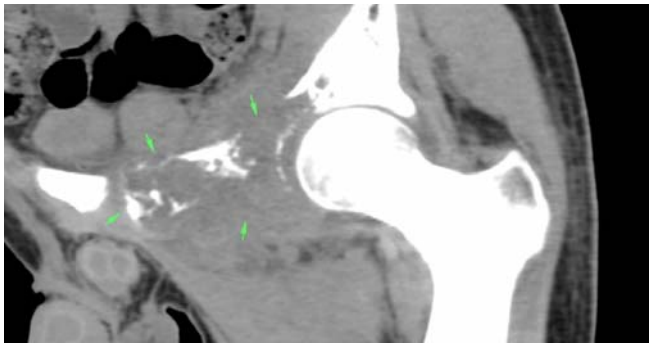

Figure 1: coronal CT Left hip (soft tissue window).

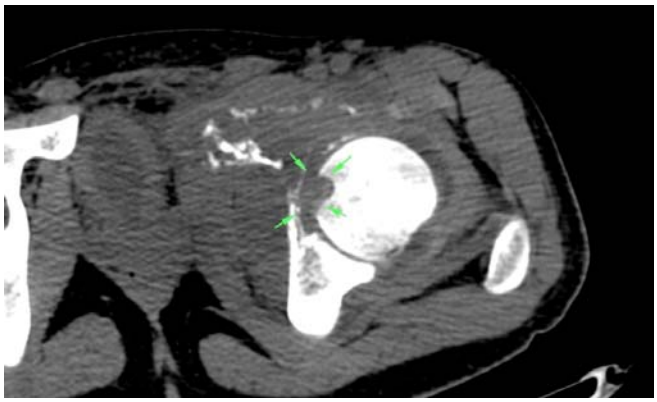

Figure 2: Axial CT cuts of the left hip joint (soft tissue window).

Figures **1** and **2** show intra-articular extension of a soft tissue mass lesion into the left hip joint causing bony erosion of the medial aspect of the left femoral head.

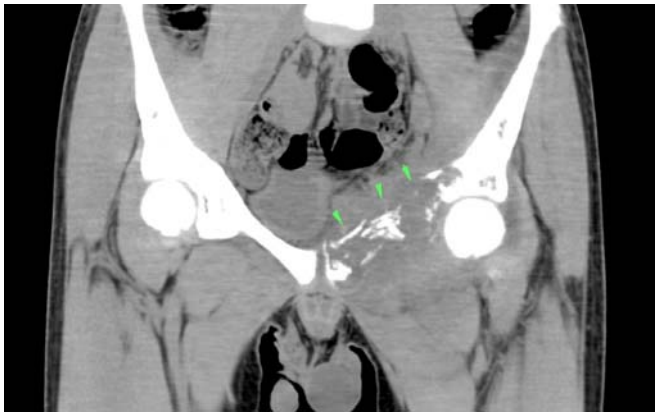

Figure 3: Coronal CT pelvis (soft tissue window) of both hips.

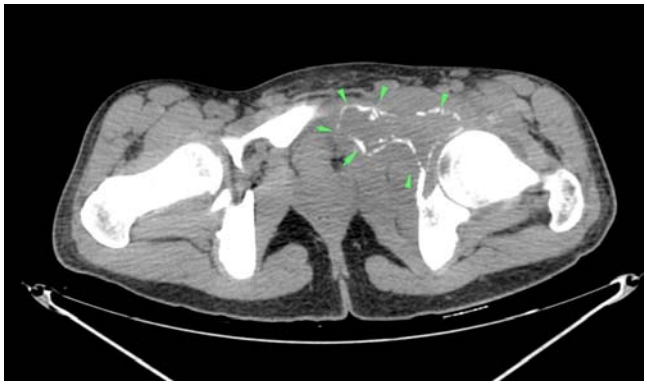

Figure 4: Axial CT pelvis (Soft tissue window).

Figures **3** and **4** show destructive osseous lesion targeting the left pubic bone and the left acetabulum with intra-articular extension into the left hip joint causing bony erosion of the medial aspect of the left femoral head. Non-remarkable appearance of the right hip joint.

Magnetic resonance imaging (MRI) of both hip joints was done to delineate the soft tissue component of the mass, which showed intra-pelvic extension with displacement of the related wall of the urinary bladder and enlarged iliac lymph nodes (Figure **5**).

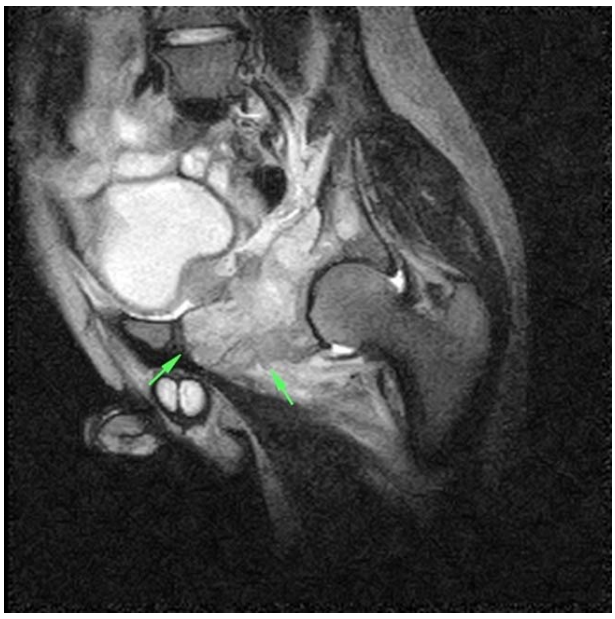

Figure 5: coronal MRI T2 weighted image showing infiltrative hyperintense lesion involving the pubic bone and acetabulum, and displacing the urinary bladder.

A biopsy was obtained from the mass. Histopathology of the lesion revealed a malignant round-cell tumor consistent with ES. The patient was referred for chemotherapy and we knew that he was started on a combination of drugs for a short time before he died.

To the best of our knowledge this is the first case report of ES in a SpA patient and the second report of ES presenting as hip monoarthritis.

DISCUSSION

ES is the second most common malignant tumour of bone in children and adolescents after osteogenic sarcoma [2] with highest frequency occuring between 10 and 15 years of age [3].

The earliest symptom in ES is pain, which depends on the site of the tumour. The most common site of ES is in the pelvis and bones of the lower limbs with 12.5% location in the iliac bone. In these cases pain may be felt in the buttock, hip, groin or low back [4]. Accordingly, cases can be easily mistaken for more common benign musculoskeletal disorders involving these regions, including sacroiliitis, lumbar disc prolapse or tendinitis [5, 6]. Sometimes a mass may be felt on palpation, if it is large enough.

ES has a bad prognosis, which is worse when it involves the pelvis and sacrum. Other bad prognostic factors include distant metastasis [7], thus early diagnosis can be a lifesaving factor which necessitates that this tumor should be included in the differential diagnosis of pain in the pelvic region [8]. A greater challenge is when a patient having a rheumatologic disease develops ES. This may cause further delay in the diagnosis since manifestations caused by ES can be attributed to the already present disease.

Seronegative SpA is a group of chronic inflammatory rheumatic diseases which affects mainly young males before their forties. Patients present with axial symptoms in the form of back pain, and peripheral asymmetric oligoarthritis involving mainly joints of the lower limbs especially the hips. In addition, the disease is characterized by the occurrence of enthesitis, which is pain and tenderness at sites of insertions of tendons, ligaments or capsules into the bones [9]. Although back pain is characterized by having an inflammatory character; i.e. increasing with rest and relieved by exercise, yet in many patients the pain is not classic [10]. Furthermore, systemic symptoms such as fever and weight loss, and increased sedimentation rate are often observed in ES [11] but also they may be part of constitutional manifestations in SpA.

The classical clinical and radiological presentation of ES may not be the rule, especially when the pelvis is involved as this is a difficult location for early

radiographic recognition. The plain radiographic abnormalities are not specific and could mimic benign disorders, this explains why cases of ES of iliac bone are frequently misdiagnosed as sacroiliitis or hip arthritis.

In a literature review on ES of the pelvis, there were three reported cases of ES presenting as unilateral sacroiliitis [12-14] and one as hip monoarthritis [15]. To the best of our knowledge this is the first report of ES in SpA patients and the second report of ES presenting as hip monoarthritis [15].

CONCLUSION

In SpA patients, ES should be included in the differential diagnosis of skeletal manifestations whenever there is a change in the character of an already present back pain, poor response to treatment following initial improvement, development of marked systemic manifestations (fever, weight loss, bad appetite) together with deterioration in laboratory investigations (marked elevation of ESR and CRP).

RECOMMENDATIONS

ES should be considered in the differential diagnosis of low back pain and hip region pain in young age group even if there is no direct pointer to the disease. In addition, in patients with a rheumatologic disease, especially SpA, the possibility of primary malignant bone tumors should be kept in mind when analyzing recent development of suspicious manifestations in order to avoid the fatal consequences of a delayed diagnosis.

REFERENCES

[1] Rudwaleit M, van der Heijde D, Landewé R, Listing J, Akkoc N, Brandt J, *et al.* The development of assessment of spondyloarthritis international society classification criteria for axial spondyloarthritis (part II): validation and final selection. Ann Rheum Dis 2009; 68: 777-83. http://dx.doi.org/10.1136/ard.2009.108233

[2] Grier HE. The Ewing family of tumors: Ewing's sarcoma and primitive neuroectodermal tumors. Pediatr Clin North Am 1997; 44: 991-1004. http://dx.doi.org/10.1016/S0031-3955(05)70541-1

[3] Vlasak R, Sim FH. Ewing's sarcoma. Orthop Clin North Am 1996; 27: 591-603.

[4] Widhe B, Widhe T. Initial symptoms and clinical features in osteosarcoma and Ewing Sarcoma. J Bone Joint Surg 2000; 82(5): 667-74.

[5] Thompson RC Jr, Berg TL. Primary bone tumors of the pelvis presenting as spinal disease. Orthopedics 1996; 19(12): 1011-6.

[6] Wilkes LL, Cannon CL, Ham OE. Malignant tumors of the pelvic girdle mimicking the herniated disk syndrome. Clin Orthop 1979; 138: 217-21.

[7] Kawai A, Healey JH, Boland PJ, Lin PP, Huvos AG, Meyers PA. Prognostic factors for patients with sarcomas of the pelvic bones. Cancer 1998; 82(5): 851-9. http://dx.doi.org/10.1002/(SICI)1097-0142(19980301)82:5<851::AID-CNCR8>3.0.CO;2-M

[8] Wurtz LD, Terrance DP, Michael AS. Delay in the diagnosis and treatment of primary bone sarcoma of the pelvis. J Bone Joint Surg 1999; 81(3): 317-25.

[9] Carron P, Van Praet L, Van den Bosch F. Peripheral manifestations in spondyloarthritis pelevance for diagnosis, classification and follow-up. Curr Opin Rheumatol 2012; 24(4): 370-4. http://dx.doi.org/10.1097/BOR.0b013e32835448de

[10] Underwood MR, Dawes P. Inflammatory back pain in primary care. Br J Rheumatol 1995; 34: 1074-7. http://dx.doi.org/10.1093/rheumatology/34.11.1074

[11] Arndt CAS, Crist WM. Common musculoskeletal tumors of childhood and adolescence. New Engl J Med 1999; 341: 342-52. http://dx.doi.org/10.1056/NEJM199907293410507

[12] Al-Adsani A, Mazy MN, Mohd M. Ewing's sarcoma of the ilium mimicking sacroiliitis. Rheumatology 1999; 38: 792-3. http://dx.doi.org/10.1093/rheumatology/38.8.792

[13] Pouchot J, Barge J, Marchand A, Carbonnier J, Vinceneux P. Ewing's sarcoma of the ilium mimicking an infectious sacroiliitis. J Rheumatol 1992; 19: 1318-20.

[14] Adelman HM, Wallach PM, Flannery MT. Ewing's sarcoma of the ilium presenting as unilateral sacroiliitis. J Rheumatol 1991; 18: 1109-11.

[15] Halwai MA, Mir BA, Wani MM, Bashir A, Hussain A. Ewing's sarcoma of the ilium mimicking inflammatory arthritis of the hip: a case report. Cases J 2009; 2: 6487. http://dx.doi.org/10.4076/1757-1626-2-6487

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