Original Article

AN INTERESTING CASE OF RIGHT ILIAC FOSSA PAIN

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Abstract

Background: The very aggressive bone tumour known as Ewing's sarcoma is more prevalent in youngsters between the ages of 10 and 20. Sometimes the conventional clinical and radiological presentation of Ewing's sarcoma may not apply, and the patient may appear in an abnormal way that causes diagnostic uncertainty. This is especially true in cases of iliac bone-related Ewing's sarcoma. We describe a patient who presented with right lower quadrant discomfort and nonspecific radiological abnormalities and who later developed Ewing's sarcoma affecting the right ilium.

Case report: A 16-year-old female presented with complaints of intermittent, deep, dull aching pain in the right groin region, fatigue, on and off fever, decreased appetite and loss of weight since 4 months. On examination temperature recorded was febrile (100^0F), pulse 110 bmp, BP - 110/70 mmhg. Pallor was present. On local examination a 2x1cm firm swelling with ill-defined margins was palpated in the right inguinal region, associated with tenderness.

Conclusion: It may not always manifest as the classic clinical and radiological presentation of Ewing's sarcoma of the ilium. Even if there is no overt sign of the illness, as there was in our instance, one should be extremely wary of it.

Keywords: Ewing's sarcoma, Fatigue, Ilium, Right Inguinal region

Introduction

The malignancy level of Ewing's sarcoma is well-known to be rather high. It was named after James Ewing,[1] and is the second most frequent primary malignant tumour of bone in children.[2] It has a specific propensity for an age group that falls between the ages of 10 and 20 years old. Even though the precise cause is unknown, researchers have discovered that 85 percent of people who have ES have a specific chromosomal rearrangement called t(11;22). This finding is despite the fact that the cause of ES is unknown. When ES is present, it can most frequently be seen in the pelvis and the lower extremities.[3]

In 12.5% of cases of ES, the genesis of the condition may be traced back to the iliac bone. In a clinical setting, the most prevalent presenting symptom is pain, which accounts for 90% of cases, followed by swelling, which accounts for 70%.[4] When a tumour is situated on the innominate bone, the patient may experience urinary and rectal symptoms as a consequence. Unless the condition has progressed relatively far, symptoms such as fever, weight loss, secondary anaemia, leucocytosis, and an elevation in the sedimentation rate are not commonly noticed in ES patients. It is possible that they will be misdiagnosed as osteomyelitis or lymphoma.

When it affects a long bone, it often causes a soft tissue mass and is characterised radiographically as a central, diaphyseal, lytic tumour. This type of tumour is frequently permeative and has a lamellated or "onion skin" periosteal response. Bone lesions are often lytic in nature, however they may also be sclerotic or mixed.[5]

The vast majority of the lesions are seen in the diaphyseal or metadiaphyseal layers. It is still difficult to diagnose Ewing's sarcoma of the ilium when it is in its early stages. This is due, in part, to the plethora of symptoms that are associated with it, as well as the extremely minor alterations that may be seen radiographically when it is in its earliest stages.[5,6]

We describe one such case that occurred in a patient who arrived with discomfort in the right lower quadrant of the abdomen and was found to have Ewing's sarcoma of the right ilium.

Case History

A 16-year-old female presented with complaints of intermittent, deep, dull aching pain in the right groin region, fatigue, on and off fever, decreased appetite and loss of weight since 4 months.

On examination temperature recorded was febrile (100^0F), pulse 110 bmp, BP - 110/70 mmHg. Pallor was present. On local examination a 2x1cm firm swelling with ill-defined margins was palpated in the right inguinal region, associated with tenderness.

Investigations

The laboratory investigations showed that the hemoglobin level of the patient was 9.8gm/dl, WBC count was 6690/ul, Platelet count was 4,06,000/cumm, ALP was 141u/lt, LDH was 503u/lt and ESR was 42mm/hr

USG Local Swelling (Right Groin Region) suggested of a well-defined solid intramuscular lesion appearing to be fibroid tumor in the upper head of adductor muscle.

X-Ray Pelvis with Hips

It showed ill-defined lytic lesion with permeative pattern involving the right superior pubic ramus, ischial tuberosity and anterior inferior iliac spine. Subtle cortical irregularity and discontinuity was noted along the ischial spine and superior aspect of right superior pubic ramus with periosteal elevation. (Figure 1)

Figure 1 MRI Hip Joint

This showed ill-defined lytic lesions in the inferior portion of the right Iliac bone, right acetabulum, superior aspect of right ischium, superior pubic ramus with abnormal soft tissue extending into the adjacent muscles such as right iliacus, deep gluteal muscles, adductor and pectinous muscles.

PET CT (whole body)

This demonstrated hyper metabolic lytic lesion in the same bony areas as mentioned above (Figure 2a) with associated soft tissue component in right inguinal region along right pelvic wall (Figure 2b). There was no evidence of any active disease elsewhere.

Figure 2a

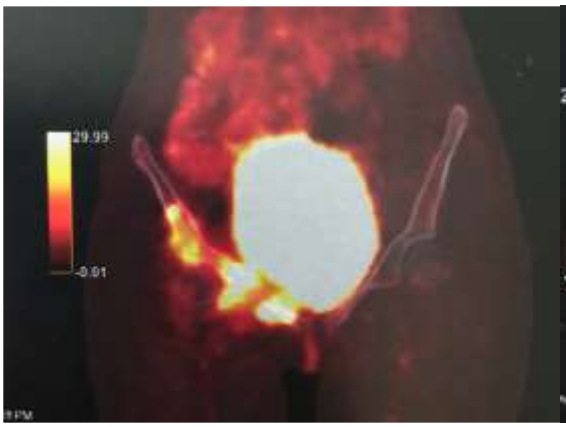

Figure 2b

Histopathology:

Biopsy from right iliac bone lytic lesion with adjacent lymph node) showed sheet like growth patter n of small uniform blue cells, separated by fibrous septa along with atypical lymphoid hyperplasia, few focal areas of discohesion and irregular nuclear contours giving an impression of small cell sarcoma favouring non-Hodgkin's lymphoma.

IHC of right iliac lesion biopsy stained positive for CD99, Ki67 (35-40%) and FLI1, and negative for CD20, cytokeratin, reticulin, desmin and other markers, suggestive of primitive neuroectodermal tumor (PNET).

Discussion

Early identification of Ewing's sarcoma of the ilium continues to be problematic, in part because the changes in the radiographs that indicate the disease are modest and difficult to spot, and also because patients often present with vague clinical symptoms. Inflammatory and tumorous lesions in the iliac bone might have a radiologic appearance that is defined by destructive changes and consolidations at the same time. This can contribute to the delay in diagnosis. This pattern does not fit any particular object. The nature of the iliac bone and the tissues that lie above it make it difficult to interpret the results of plain films taken of this region.[7]

There have been occasions in which writers have identified Ewing's sarcoma of the ilium as sacroilitis[8-10] and in some cases even septic arthritis,[11] probably due to transarticular dissemination in these individuals. Sacroiliitis is an inflammation of the sacrum and ilium. The authors of these instances relied mostly on clinical presentation and plain radiography during the earliest stages of the diagnostic process, which resulted in diagnostic perplexity. Additionally, there have been reports of instances of Ewing's sarcoma of the ilium presenting symptoms similar to those of juvenile rheumatoid arthritis[12] and discomfort in the hip.[13]

Again, the patient's presentation as having discomfort in the right lower quadrant led to diagnostic uncertainty, which in turn delayed treatment until a definitive diagnosis could be made. An ultrasound examination has been suggested in the past as a tool for evaluating the presence and extent of the extraosseus component as well as assessing the therapeutic response of tumours which originate in the skeleton in close proximity to the pelvis,[14] but with today's advanced diagnostic methods, its role is very limited.

The nature of the iliac bone and the tissues that lie above it make it difficult to interpret the results of plain films taken of this region. Because of this, a magnetic resonance imaging (MRI), computerised tomography (CT), and bone scan are all required. The pattern of bone deterioration and the accompanying soft-tissue mass may be better defined with the use of CT, which is helpful in determining the tumour of ES. When it comes to detecting involvement of soft tissues and metastases in the bone marrow, MRI is more sensitive than CT. An open biopsy and histological examination, together with immunochemistry and cytogenetics testing, are required to arrive at a definitive diagnosis.[7]

Patients diagnosed with Ewing's sarcoma require a treatment strategy that incorporates the expertise of several medical professionals, including oncologists, radiation oncologists, surgeons, and radiologists. Children who have tumours that cannot be resected or who would suffer loss of function are treated with chemotherapy and radiation therapy alone. Surgery is effective and appropriate for patients who can undergo complete resection with acceptable morbidity; however, surgery is not performed on children who have tumours that cannot be resected.

The prognosis is determined by the severity of the illness, the size and location of the tumour, whether or not the tumour has spread, how well the tumour responds to treatment, the patient's age, and whether or not the disease has recurred. The majority of facilities in the world have a long-term survival rate of between 60 and 70 percent nowadays. The existence of distant metastases is the element that is most detrimental to one's prognosis. Patients diagnosed with metastasis have just a 20% chance of surviving for an extended period of time, even with intensive therapy. There is no correlation between histological grades and patient outcomes.

Conclusion

We believe that Ewing's sarcoma should be kept as one of the rare differential diagnoses for lower quadrant pain. Ultrasonography should not be relied on much and X-rays may not add more to the knowledge. Lower threshold should be kept for investigations such as CT and MRI to detect the disease early and start the appropriate treatment.

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