

Ewing sarcoma: A rare report from the Indian pediatric population

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ABSTRACT

Ewing's sarcomas are rare, aggressive tumors with a tendency toward recurrence following resection, and early metastasis. Although patients of younger or older age account for almost 30% of instances, peak incidences occur between the ages of 10 and 20. We, hereby, report the case of a 10-year-old girl who presented with a 3-month history of pain in her right hip that was unable to be relieved by non-steroidal anti-inflammatory medicine and physical therapy. On examination, bone marrow aspiration revealed that the infiltration of malignant small round blue cells was managed by anticancer therapy successfully.

Key words: Bone tumor, Cancer, Children, Ewing sarcoma, Hip, Indian population, Round cell tumor

Ewing sarcoma (ES) is a round-cell tumor that typically arises in the bones and rarely in the soft tissues of children and adolescents. Ewing's sarcoma was originally termed diffuse endothelioma or endothelial myeloma. It was named after James Ewing, who identified it in 1921 [1]. At present, ES is a part of peripheral primitive neuroectodermal tumors (PNETs) as it shares a common cytogenetic translocation of chromosomes 11 and 22 [2]. The etiology of most primary tumors of the bone has not been determined. Radiation exposure does not appear to be causative for Ewing's sarcoma of the bone, and no association with familial cancer syndromes has been verified. Embryonic ES of the bone, the second most prevalent bone cancer in children and adolescents, affects an estimated 2.1 million children in the United States [3] and accounts for 4% of all childhood and adolescent malignancies [4]. ES accounts for 3.5% of cancers in children aged 10–14 years old in the United States, and 2.3% of cancers in teenagers aged 15 to 19. It is slightly more common in males than in females [2]. Most of it is found in individuals who are white or Hispanic and is extremely rare in children of Asian or African descent [3,5]. The only known established risk factor for ES is race and ethnicity [6,7].

Herein, we report the clinicopathological correlation in a 10-year-old girl with no known comorbidities who presented with pain and swelling over the right lower extremity. This case is being reported to enlighten the better clinical aspects of ES. Our case would be the first report of ES on the hip in the Indian pediatric population.

CASE REPORT

A 10-year-old female child presented with pain in the right hip and restriction of movement for 3 months. It was also associated with swelling in the right hip which was gradually progressive in nature. There was no history of trauma, rash, bleeding, or swelling in any other region.


On examination, the patient was conscious, alert, and afebrile, and his vitals were stable. There was tender swelling in the lateral aspect of the right hip joint. While walking, limping was noted. The pain score was recorded as 5/10 using the Wong-Baker FACES pain rating scale.

Her magnetic resonance imaging shows alteration in signal intensity of the right ilium, ischium, and pubis, including acetabulum, pubic rami, and right sacral wing, which is hypointense in T1-weighted images and hyper-intense in T2-weighted images with soft-tissue mass and thickening of the iliopsoas, gluteus medius and minimus, and obturator externus muscles on the right side. Positron emission tomography and computed tomography demonstrated an extensive right iliac or pubic tumor extending to the sacral spine (Fig. 1). It has metastasized to the femur and the ribs.

Aspiration of the bone marrow showed involvement by malignant small round blue cells. The neuropathic nature of pain is due to nerve infiltration by the malignant cells, so in view of adequate symptom management, she was started on opioids with adjuvants (pregabalin and tricyclic antidepressants). Palliative intent chemotherapy has been started with the drugs vincristine, doxorubicin, and cyclophosphamide alternating with ifosfamide

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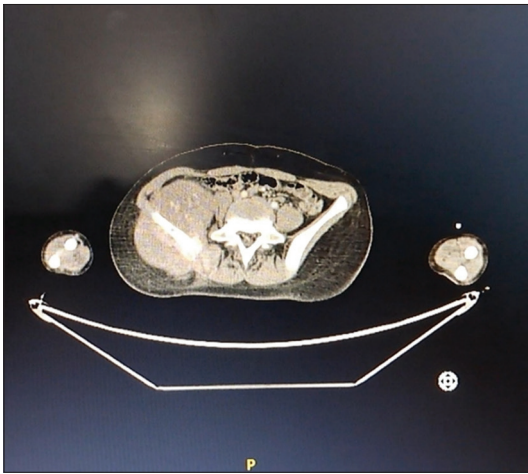


Figure 1: Pelvic computed tomography

and etoposide. The patient was clinically improved after four cycles of chemotherapy.

DISCUSSION

Dr. Arthur Purdy Stout first identified an ulnar nerve-derived undifferentiated tumor in 1918 [8]. Due to the rosettes, he gave it the name PNET. ES, which bears his name, was first identified as a malignancy affecting long bones in 1921 by renowned American pathologist James Ewing [9]. These two lesions were subsequently defined as distinct entities affecting diverse areas. The distinction between ES and PNET, however, started to wane by 1975, and it was discovered that these two tumors represented opposite ends of the same histological spectrum and were categorized as part of Ewing's family of cancers (EFTs) [7].

Clinically, it manifests as a mass that is palpable and associated with pain and swelling. Paraspinal or pelvic tumors can be related to back pain and paresthesia. Fever, exhaustion, weight loss, and anemia are typical non-specific systemic signs that can mimic an acute or ongoing infection. Depending on the precise location of the tumor, there may be other symptoms and indicators.

In 50% of patients, ES symptoms remained for longer than 6 months, according to the Grier HE research [10]. Instead of the initial tumor site, between a fifth and a quarter of people with ES [10,11] present with metastatic illness and discomfort from metastasis. In our case, the symptoms of pain and swelling in the right hip with restricted movements persisted for about 3 months before the diagnosis.

Patients should receive evaluations from experts from all fields before beginning treatment (e.g., radiologist, chemotherapist, pathologist, surgeon or orthopedic oncologist, and radiation oncologist). To ensure that the incision is made in the proper spot, the orthopedic oncologist or surgeon who will perform the definitive surgery should be involved before or during the biopsy. Systemic chemotherapy must be used in concert with either surgery, radiation therapy, or both modalities for local tumor control to successfully treat patients with EFT. The patients typically undergo pre-operative chemotherapy before beginning local control treatments. Vincristine, doxorubicin,

ifosfamide, and etoposide are always included in the multidrug chemotherapy regimen for EFT. Cyclophosphamide is also used in the majority of procedures. Primary chemotherapy lasts anywhere between 6 months and a year. Children who have unresectable tumors or who would have a loss of function are treated with radiation therapy alone, even though surgery is an effective and suitable option for those who can undergo complete resection with acceptable morbidity. Adjuvant radiation therapy may be helpful for patients who undergo large resections but still have a microscopic residual illness. This particular individual was treated with six cycles of vincristine 1.5 mg/m², doxorubicin 37.5 mg/m², cyclophosphamide 1200 mg/m² with mesna rescue alternating with ifosfamide 1800 mg/m² and etoposide 100 mg/m² (IE) as described by Grier *et al.* [5].

The prognosis is based on the severity of the disease, the size and location of the tumor, the existence or absence of metastasis, the response of the tumor to treatment, age, and the likelihood of relapse. At this time, 60–70% of centers report long-term survival. The occurrence of distant metastasis is the worst prognosis indicator. Patients with metastasis have a 20% likelihood of long-term survival, even with vigorous treatment [12]. The predictive value of histological grades is negligible. Fever, anemia, elevated white blood cells, erythrocyte sedimentation rate, and lactate dehydrogenase levels have all been linked to worse prognoses and more severe disease, as was already indicated [13].

CONCLUSION

We have described a unique case of ES, as it is the first pediatric report arising from the right hip in the Indian population. Ewing's sarcoma is an extremely rare entity, with a poor prognosis. Due to its high metastatic potential, it demands early intervention. The patient showed a favorable response to the treatment with anticancer drugs and pain relievers.

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