Ewings Sarcoma Mimicking a Schwannoma: MRI Findings of a Rare Case

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Abstract

Ewing's sarcoma is among the most frequent yet highly aggressive neoplasms of the bones presenting in adolescents and children under the age of ten with a slight male predilection. It is now broadly categorized into a set of tumors recognized as Ewing's sarcoma family based on the same histology and genotype of these tumors. This group of tumors includes Ewing's sarcoma of bones, the 2nd most frequent bone malignancy, Askin tumors, PNET, and those rarely occurring extraosseous Ewing sarcoma (peripheral neuroepithelioma). Extra-osseous Ewing's is a rare presentation occurring in only 5% of patients. Here we present such a case of an extra-osseous Ewing's sarcoma in a 13-year-old female presenting to our hospital with a large mass in the sacral region. The interesting imaging findings and his histopathology are discussed.

Keywords: *Extra-osseous Ewing's sarcoma, MRI spine, lumbar spine.*

INTRODUCTION

Ewing's sarcoma, which bears James Ewing's name and is the second-most common malignant bone tumor overall and the first-most prevalent in adolescents and young children, was first reported in 1921 [1-3]. Males are more likely to experience it, with a ratio of 1.3 to 1.5:1, presenting in the first and second decade of life only a few cases presenting after the third decade [1-9]. Ewing's sarcoma belongs to a newly introduced family of tumors called the Ewing Sarcoma Family Tumors as these tumors have the same genetic and histological characteristics [1-4]. This group of malignancies comprises Ewing's tumor of the bone, PNET, extra-osseous ES (peripheral neuroepithelioma), and Askin tumours. On cytological and genetic grounds these tumors share the same translocation t (11; 22) (q24; q12) leading to the creation of a gene called EWSR1-FLI1 and also share similar oncogenes (c-myc, c-myb, c-ets-1, and N-myc). The EWSR1-FLI1 is found in approximately 85% of these tumors [2, 3]. Histologically these tumors show small round blue cells which are stained with hematoxylin and eosin. These may show areas of necrosis, rosettes, and pseudo-rosettes. Rosettes formation is related to the reactivity of these tumors to HBA-71, which is an antibody against MIC 2 involved in cell adhesion processes [2, 3].

The extra-osseous form of Ewing's is a very rare presentation with the spine being involved in only 3.5 to 7% of the case of EES. The first case of Ewing Sarcoma Family tumor was described by Stout *et al.* in 1918 of ulnar nerve origin with histopathological features suggestive of a neuroepithelioma [3]. Nesbitt, Vidone, and Martins *et al.* presented cases of PNET's of the sciatic nerve [3]. However, according to some literature, Tefft *et al.* were the first person to describe the extra-osseous form of Ewing's in 1969 renamed EES in 1975 by Angervall *et al.* [2]. Patients with EES most present with a fast-growing tumor and backache radicular pain, sensory disturbances, and urinary and fecal incontinence. These tumors present with distant metastasis in most cases. The most common sites include the chest wall, lung, pelvis, scalp, and neck including the larynx, paravertebral region, and limbs [1].

The imaging modalities that can be used include X-rays, CT scans, and MRI. The imaging findings of extra-osseous Ewing sarcoma can be non-specific however some cases have reported that these tumors usually have a pseudo-capsule and high vascularity with a dense post-contrast enhancement. Nevertheless, imaging can help define the extent of the disease and the extent of spinal involvement. The presence or absence of spinal stenosis is important information needed by clinicians [1-9].

These tumors are treated surgically, then with chemotherapy and radiotherapy containing a combination of vincristine, adriamycin, and cyclophosphamide called VAC and ICE *i.e.*, ifosfamide, cisplatin, and etoposide [1-9]. Histopathology plays an important role in supporting the differential diagnosis in this case. We have also presented here a case of an EES in a young female with no previous co-morbid.

CASE REPORT

A 13-year-old girl complained of lower back aches for the last 15-20 days associated with swelling at the back

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Histopathology showed Ewing's sarcoma

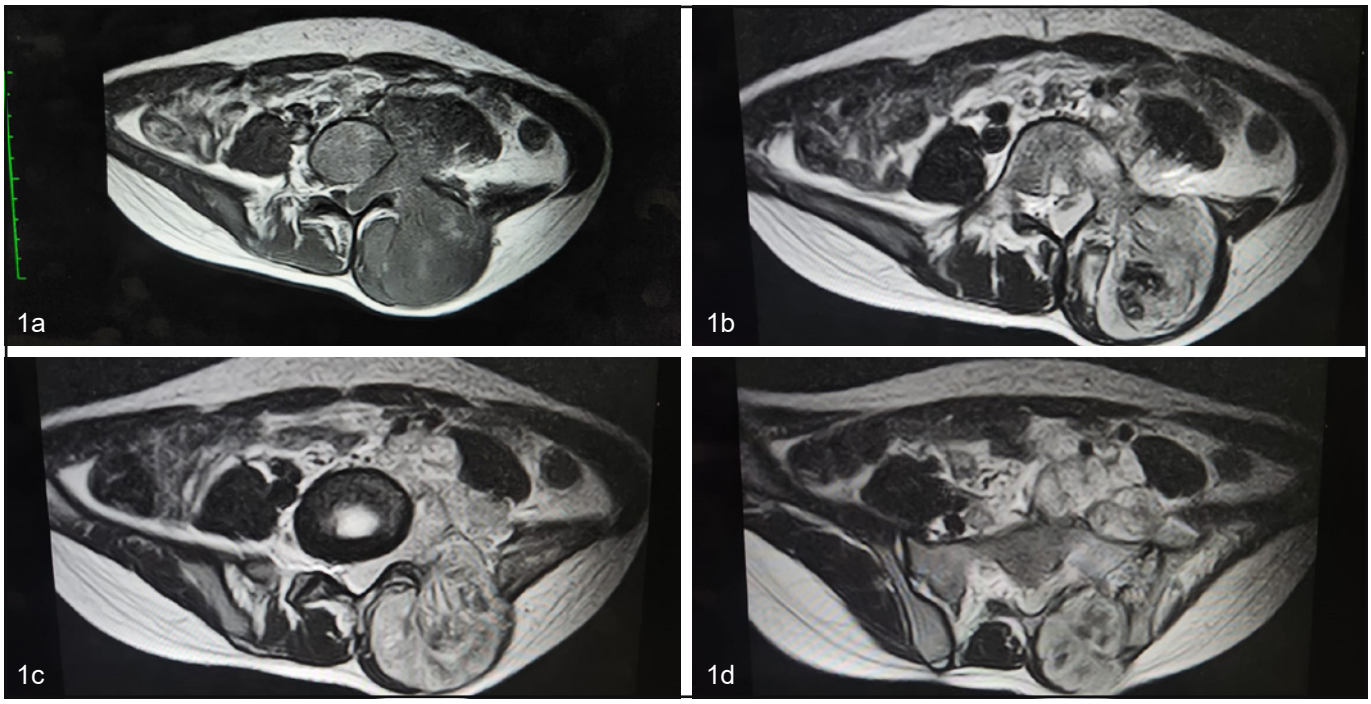

Fig. (1a-d): MRI T2 images axial cuts. An abnormal signal intensity mass showing areas of necrosis is seen in the left paraspinal region showing infiltration in the left paraspinal muscle at the level of L3/L4 vertebral bodies which was infiltrating into the left exit foramina and left psoas muscle and posterior abdominal and into the left exit foramina at the level of L4/5 and in the spinal canal, continuing up to the sacral region of about the level of S3.

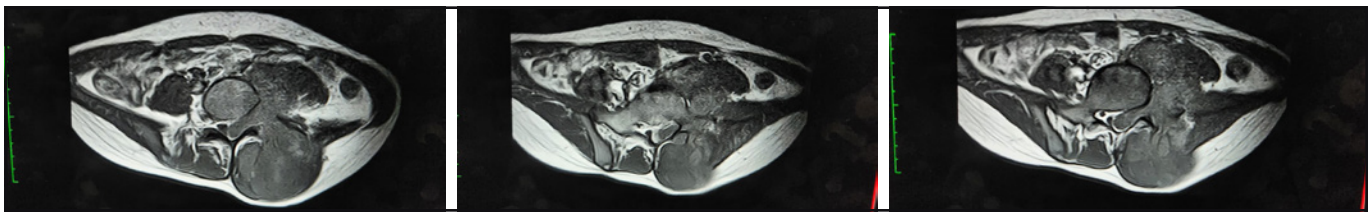

Fig. (2): MRI T1 images axial cuts showing a low signal mass with extensions as described above.

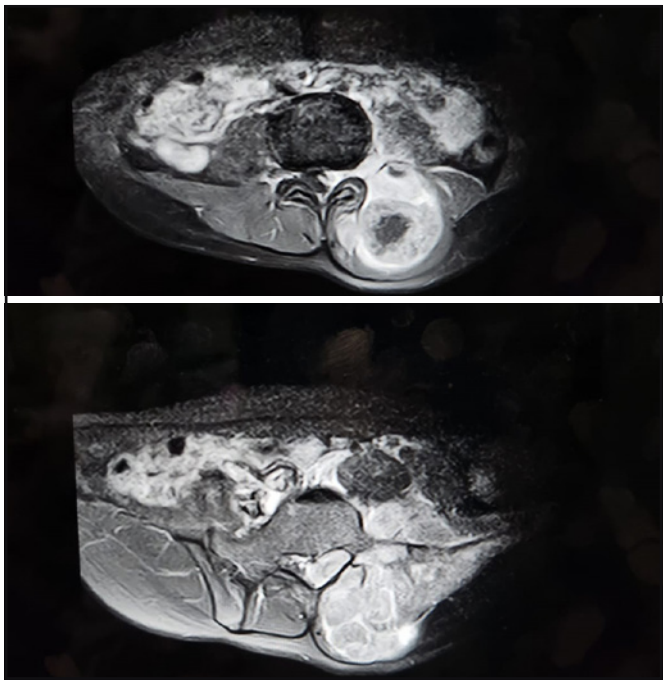

Fig. (3): MRI post contrast axial cuts show heterogenous post-contrast enhancement.

for around 15-20 days. There was no history of falls, trauma, heavy weight lifting, or fever. The bladder and bowel were intact. There was no complaint of urinary or bowel incontinence. She also had a complaint of left shoulder pain which was found to be a frozen shoulder which improved after physiotherapy. Back pain radiates in the left lower limb to the level of the toes, progressively increasing associated with numbness. She was unable to walk. She had severe dysesthesia. On examination, GCS was 15/15, cranial nerves intact, upper limb power was 5/5 bilaterally, right lower limb power was 5/5, left lower limb +4/5 proximally, left EHL was 0/5, dorsi flexion, plantar flexion was 2/5, sensations were intact, sensations impaired on crude touch and pinprick in the left L5, S1 dermatome. On local examination, a large soft tissue mass, firm in consistency in the paraspinal position at the level of L4 till S1 around 10 x 10cm fixed non-mobile. There were no associated skin changes noted. SLR was restricted on the left side to 30˚, right SLR was 70, FABER was bilaterally negative, reflexes of +2, and ankle reflex was absent on the left side. CT spine showed a neoplastic mass in the left posterior paravertebral region in the lower lumbar region, with

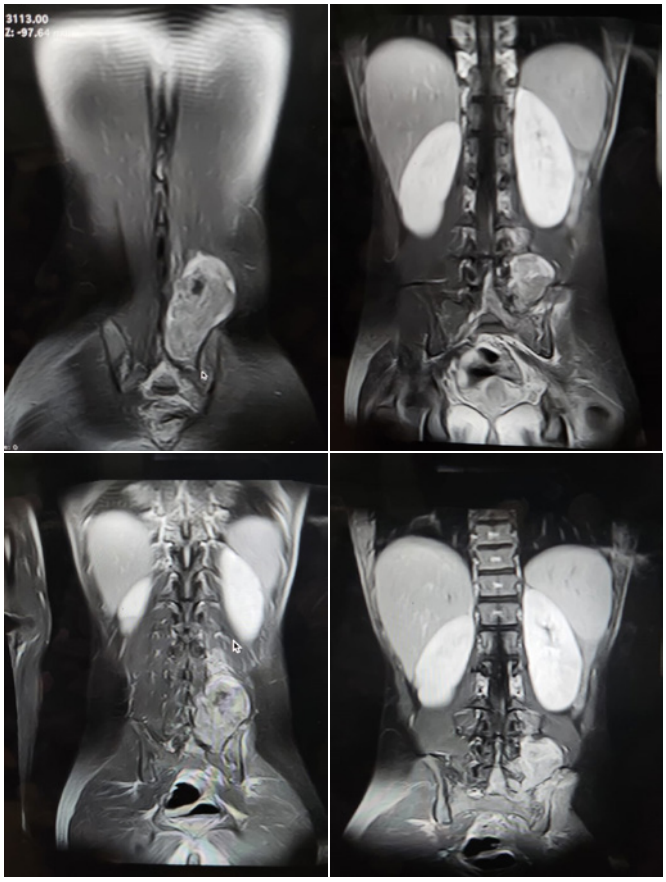

Fig. (4): Coronal MRI images T2 sequence showed infiltration in the left paraspinal muscle at the level of L3/L4 vertebral bodies which was infiltrating into the left exit foramina and left psoas muscle and posterior abdominal muscle forming a large mass with central area of low signal extending into the left exit foramina at the level of L4/5 and in the spinal canal, continuing up to the sacral region of about the level of S3.

subtle lucency seen in the center of the L3 vertebral body. MRI lumbosacral spine showed a large mass in the left paraspinal region measuring approximately 5.6 x 5.1 cm in diameter, due to the extension into the spinal canal there was a compromise of the spinal canal with pressure effect over the left descending and left exiting nerve root (**Figs. 1-4**). Her metabolic workup showed Vit 3 of 71.87 and Creatinine of 0.91.

After explaining the diagnosis, risk, and procedure, after all aseptic measures, the position was made. The patient was prepped and draped. The incision was given over L4 to the S2 region. A whitish grey-colored solid cum cystic tumor was identified in the left L4 to S2 paraspinal region extending from the spinal canal (L4 to S2) involving the left paraspinal region anterior till retroperitoneal region superior till L4 inferiorly till S1. Laminectomy was done from L4 till S2. The tumor was removed. The dural tear was identified and repaired with muscle flap and beriplast. Haemostasis secured. The wound closed in layers. Drain placed. An aseptic dressing was applied. Post-op, the patient was shifted to the ward and subsequently discharged to be followed up in the clinic.

DISCUSSION

A rare tumor of the bones called Ewing sarcoma typically strikes children and adolescents. Additionally, soft tissue like muscles, fat, and nerves are susceptible to it [1]. Schwannoma, on the other hand, is a benign tumor that arises from the Schwann cells, which form the protective covering around the nerves. While both of these tumors can occur in the nerves, they are distinct entities with different clinical and radiological features [1, 2].

Because of its quick growth, poor prognosis, and increased recurrence rate, ES is a particularly dangerous malignant tumor [4-6]. There are no specific symptoms the patients mostly present with pain or soft tissue swelling as the lesion presses down the relevant nerves and can cause neurological dysfunction [5]. In this instance, we describe a primary extra axial Ewing sarcoma of the nerve root case. The sufferer reported back pain and weakness in both lower limbs.

The case report discusses a highly aggressive malignant tumor called Ewing sarcoma (ES) that usually lacks apparent early warning signs and can be difficult to diagnose [5]. The patient in this case had primary L3 EES of the nerve root which brought about excruciating nerve root aches. Due to the lack of distinct imaging findings for ES and instead variable disease-site appearances, a high likelihood of misinterpretation occurs [4, 5]. Imaging however may provide the extent of disease and the diagnosis of Ewings should be borne in mind when we come across such cases. In our case a large mass measures approximately 5.6 x 5.1 cm in diameter with infiltration in the left Para spinal muscle at the level of L3/4 that is infiltrating the left exit foramina, left psoas muscle, and posterior abdominal muscle, forming a large mass with central area of low signal extending into the left exit foramina at the level of L4/5 and in the spinal canal, continuing up to the sacral region of about the level of S3. With extensions as mentioned above [3-5].

Pathology is used to make the diagnosis of ES because it displays a lot of primitive, tiny, spherical cells with irregular morphology. The recommended treatment for EES is chemotherapy, surgery, and radiotherapy, but the chance of recovery is still bleak, with a two-year survival rate of roughly 40% and a three-year survival rate of roughly 10%. This case report serves as a general guide for raising preliminary diagnosis rates, reducing misdiagnosis rates, encouraging prompt treatment and diagnosis, and enhancing patient prognosis [7, 8]. This case report discusses Ewing's sarcoma (EES) developing mainly from the spinal epidural space. The average patient is 22 years old at the time of diagnosis, and they frequently disregard general symptoms like back pain. Back pain and/or radiating pain, paresis of one or more limbs, sensory disruption, and bladder and bowel dysfunction are among the most prevalent symptoms [8]. These are a group of tumors that include ES, PNET, extraskeletal ES, atypical ES, and Askin tumors which had previously been assumed

to be unconnected. ESFT's hallmark translocation is between the EWS gene and the transcription factor FLI-1. A spinal epidural tumor has a wide range of possible diagnoses, including ES, and a crucial part of the curative approach for these tumors is surgical resection. Adjuvant treatments are of the utmost importance after surgical resection to manage microscopic involvement. Multi-agent chemotherapy is recommended because ES is a unique illness, and treatment strategies in older patients are similar to those in pediatric age [8, 9]. Diagnosis is frequently delayed, and effective therapy may also be delayed because it is misdiagnosed as an abscess or spinal tuberculosis. A laminectomy with maximal tumor excision is performed, accompanied by rigorous adjuvant radiation and chemotherapy but in speaking for children laminectomy is avoided whenever possible because it almost always results in main spinal deformity. Local management requires a form of radiation treatment, and a radiotherapy dose of 50 Gy along with combined chemotherapy is optimum. Yet due to the relatively small number of cases documented in the literature, commenting on an association between dose and response is challenging.

If there is any suspicion of Ewing sarcoma on MRI, a biopsy is typically recommended to confirm the diagnosis. Several factors, including the degree of tumor growth and site of the tumor, chemotherapy, surgery, and radiation therapy are frequently combined in the management of Ewing sarcoma. The goal of treatment is three; histology, maintaining stability, allowing a normal growth of the spine in children and adolescents, and obtaining the maximum resection possible.

CONCLUSION

Ewing's sarcoma although rare in the extra-osseous location can mimic extra-medullary tumors of the spine and should be considered as a differential in the tumors of this region. However, the definitive diagnosis undoubtedly depends on the histopathological findings.

CONSENT FOR PUBLICATION

Written informed consent was taken from the participants.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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Declared none.

AUTHOR'S CONTRIBUTION

All the authors contributed equally to the publication of this article.

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