



## Case Report

## “An unusual presentation of a sacral Ewing’s sarcoma in a young Syrian female”

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## ABSTRACT

A primary spinal Ewing sarcoma is extremely rare to be found. To the best of our knowledge, it is the first reported case for bilateral foot drop as a prodromal symptom of sacral Ewing sarcoma. The case, we are presenting, is of an 18-year-old athletic female, who was presented to the emergency department suffering from sudden severe low back pain radiated to the lower limbs, associated with bilateral foot drop and later urinary incontinence. Following the indicated surgery and subsequent histopathology study, we diagnosed this rare type of tumors. In the following article, we are describing the clinical presenting features of this tumor and discussing the clinical aspects.

## 1. Introduction

Primary spinal Ewing sarcoma is exceedingly rare. It usually occurs in children and young adults. It can be divided into sacral and non-sacral according to its response to treatment and survival rate [1,2]. Diagnosis of sacral Ewing’s sarcoma is quite challenging, so it oftentimes delays, as patients usually ignore their symptoms until they get worse. Consequently, this tumor usually increases in size and may reach the pelvic region, causing anatomical sabotages as well as neurological damages that may lead to paralysis of lower extremities [1,2]. Treatment depends on the combination of surgical resection and radio/chemo therapy to improve the prognosis [1,2].

We are reporting this case because of the rarity of this tumor and its possible vague manifestations, like the isolated back pain and the bilateral foot drop in our case; besides, clear recommended guidelines for treatment of spinal Ewing sarcomas are still lacking.

## 2. Case presentation

An 18-year-old athletic female was presented to the emergency

department suffering from sudden severe low back pain radiated to the lower limbs, associated with bilateral foot drop. She has already had a history of a post-traumatic low back bruise, associated with mild lumbar back pain for the last three weeks, which was revealed by analgesics. No clinically significant weight loss or loss of appetite was noticed. Medical and family history was unremarkable.

Physical examination revealed a complete left foot drop (0/5 on manual muscle testing- MMT-) with a partial foot drop (3/5 using MMT) on the contralateral limb. Both Achilles and patellar reflexes were absent on the left side, with a weak response on the other side. Babinski reflex was positive bilaterally. Saddle numbness and paresthesia were present. No urinary or fecal incontinence was reported. Admission work-up was within normal. Both lumbosacral computerized tomography (CT) and magnetic resonance imaging (MRI) revealed a large oval-shaped (3.5 × 5x5.5 cm) osteolytic mass extending from the upper border of the third lumbar vertebra to the whole part of the sacrum, and reaching to the para-spinal soft tissue and extradural space, as shown in Fig. 1.

Later, the patient experienced urinary incontinence. Consequently, a surgery was scheduled to relief compressive symptoms and confirm the diagnosis via biopsy. The patient was informed about the current

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situation and possible outcomes. Surgery was performed through a posterior midline incision extending from the level L3 to the sacrum; a necrotic bleeding mass was found infiltrating into the paraspinal soft tissues, and extending into the extradural space. Excisional biopsies along with decompressive laminectomy of L3-L4-L5 were performed. The surgery was operated by a consultant neurosurgeon.

Histo-pathological examination revealed characters of a malignant small round-cell tumor that was consistent with Ewing sarcoma.

Post-operatively, the patient was discharged three days later and attended a physical therapy course along with the proposed radio \chemo therapy protocol. The patient was followed for 6 months with regular monthly visits. The patient experienced a noticeable clinical improvement, as the back pain resolved significantly and the muscle strength improved (3/5 for left leg and 4/5 for right leg measured by MMT). Additionally, she gradually regained bladder control.

### 3. Discussion

Ewing's sarcoma (ES) was first mentioned in 1921 by James Ewing, it is considered the second most common primary bone sarcoma after osteosarcoma [1,3]. It comprises 3,5–5% of the entire bone sarcomas – 3 per million in children-; with a predominance among Caucasians. Nevertheless, it is still a rare tumor especially in the spine, this is what encouraged us to report this case. It can be divided into sacral and non-sacral according to its response to treatment and survival rates [1, 4]. ES affects individuals among the first two decades and this is consistent with our case. In contrast with our case, ES has a predilection for the male sex with a male-to-female ratio of 1.3–1.5:1 [1,3].

Symptoms and signs depend on the tumor site and severity of spinal cord compression. ES is usually associated with specific related symptoms such as night or rest back pain (about 100%) followed by

radiculopathy manifestations; i.e functional limb weakness (70%) and paresthesia (50%) [1–3]. Our patient suffered from backache for the last 3 weeks which responded to analgesics, without any radiculopathy manifestations. Nevertheless, out of the blue, she suffered from sudden severe backache associated with bilateral foot drop. This unusual presentation makes our case unique, as, to the best of our knowledge, it is considered the first reported case for bilateral foot drop as a prodromal symptom of sacral ES.

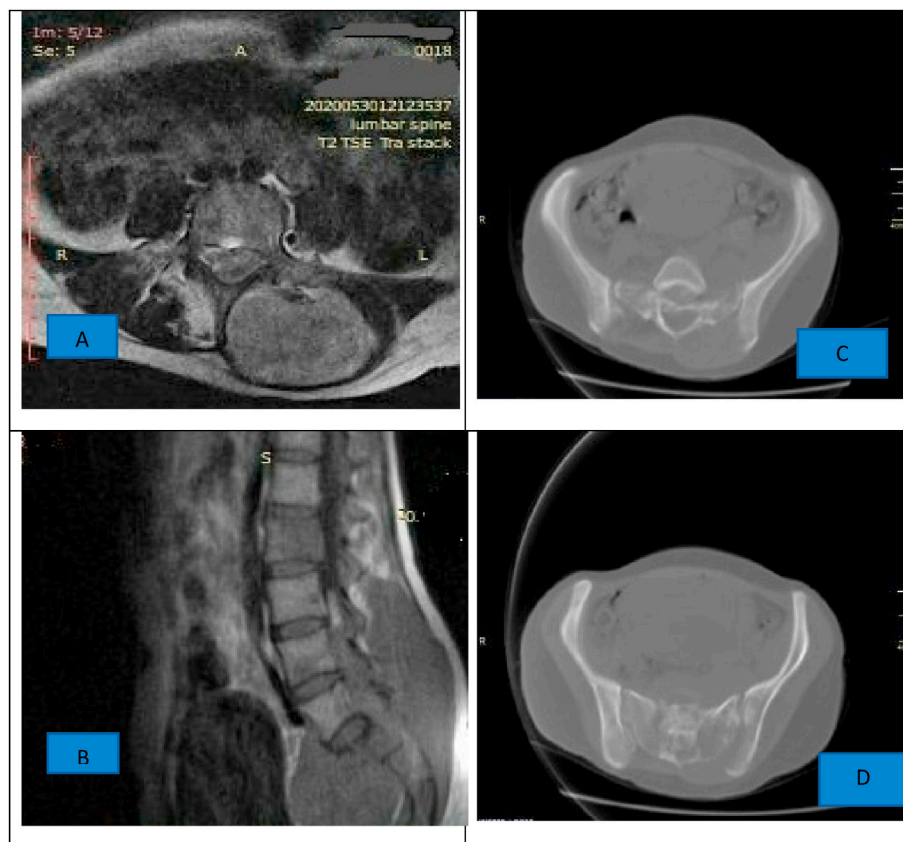
Laboratory work-up may reveal nonspecific findings of inflammation, such as increased sedimentation rate, moderate leukocytosis, and anemia; on the contrary, our patient's admission work-up was within normal even for the serum lactate dehydrogenase level [1–3]. Mostly, tumor size increases gradually until it could be palpable. Nevertheless, no palpable mass was found in our case.

In Ewing's Sarcoma, radiography usually shows lytic, sclerotic or mixed lesions involving para -spinal soft tissue with the extra dural space. However, MRI is considered the best diagnostic tool for such cases, it is non-specific. Radiological diagnosis of this tumor is considered a challenge, as the main role of radiography is excluding other differentials [1,2,5].

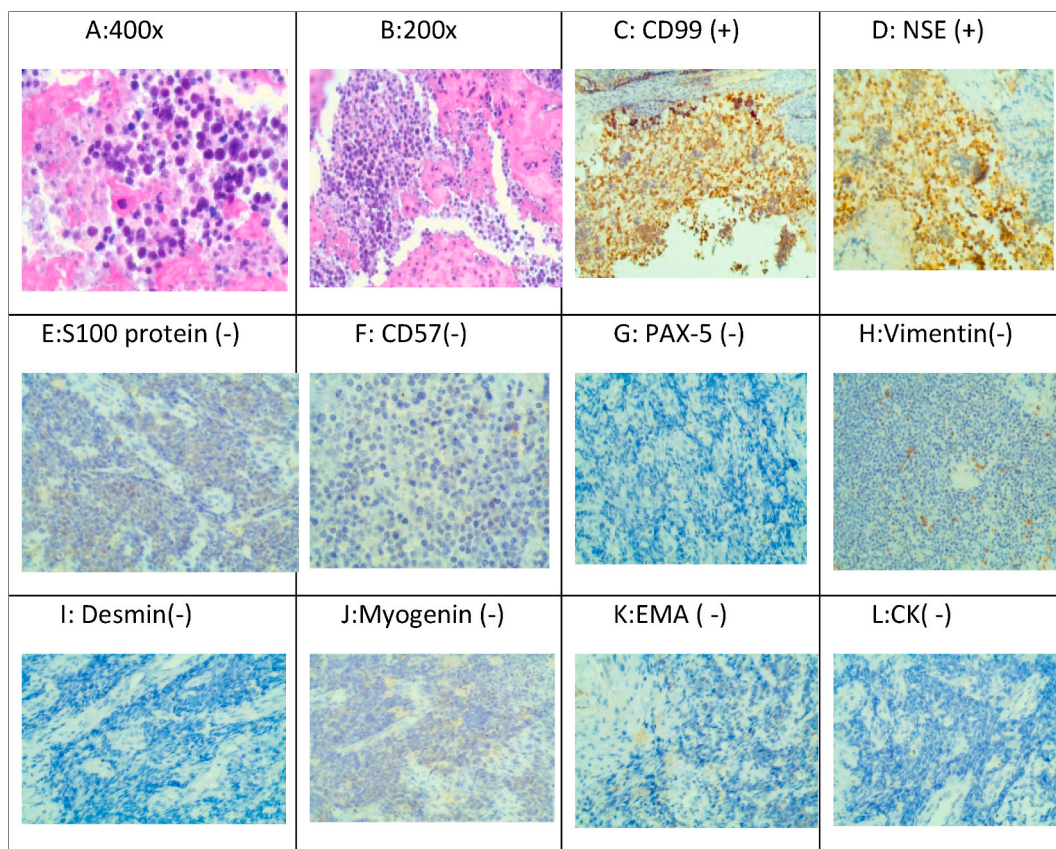
Differential diagnosis includes teratoma, chondroma, giant cell tumor, Ewing sarcoma, osteoblastoma, neuroblastoma, chondrosarcoma, meningeocele, aneurysmal bone cyst meningioma, neurofibroma, dermoid cyst, abscess, embryonal rhabdomyosarcoma, schwannoma, synovial sarcoma, osteogenic sarcoma, hemangioma, nerve sheath tumor, lymphoma, and leukemia [1–3].

The definitive diagnosis depends on the histopathology, which reveals a homogeneous population of small round cells with high nuclear tocytoplasmic ratios that are arrayed in sheets, nests and pseudo acinar sub-groups [1–3]. Apoptosis and mitosis are found, as shown in Fig. 2.

The Immune stains show positively for CD99 and NSE, and



**Fig. 1.** Pictures, A&B: The MRI shows a large oval-shaped (3.5 × 5x5.5 cm), which appears isointense on T1 WI and mildly hyperintense on T2 WI. Pictures, C&D: The CT demonstrates an osteolytic lesion of the fifth lumbar vertebra and sacrum.



**Fig. 2.** Histopathology reveals a homogeneous population of small round cells with high nuclear to cytoplasmic ratios that are arrayed in sheets, nests and pseudo acinar sub-groups. Apoptosis and mitosis are found (pictures A&B). The immune-histochemical stains show positively for CD99 and NSE, and negatively for Vimentin, cytokeratin (CK), Desmin, EMA, Myogenin, S100 protein, Pax5 and CD 56.

negatively for Vimentin, FL-1, LCA, TDT, Desmin, Synaptophysin, EMA, Myogenin, S100 protein, Pax5 and CD 56 [1–3], as shown in Fig. 2.

Once the diagnosis of Ewing’s sarcoma is confirmed, neo adjuvant chemotherapy should be started aiming to shrink the tumor, thereby increasing the chances of total excision, besides taking care of micrometastases, resulting in presenting an idea about responsiveness of the tumour to adjuvant therapy(chemo/radio therapy) to control the possible micrometastases [1–3].

Concerning the prognosis, Lahl M. et al. determined accurately the negative and positive prognostic factors as shown in Table 1 [4]. In our case, we can notice that the site of the tumor plus the age and the osteolytic lesions are considered negative factors. Owing to the rarity of sacral tumors, there are no standard survival rates, however some, like Marco et al. have estimated the survival rate of patients treated with chemotherapy and radiotherapy for spinal Ewing’s sarcoma, as about 50% at five years and 35% at 10 years [3,4]. This work has been reported in line with the SCARE 2020 criteria [6].

**4. Conclusion**

Sacral primary Ewing’s sarcoma is an extremely rare tumor, which

needs higher suspicion, even when the patient presents with isolated back pain. Therefore, to keep in mind, isolated back pain should be investigated widely. The mainstream of treatment depends on the combination therapy of chemotherapy, radiotherapy and surgery to improve the survival rate.

**Ethical approval**

No ethical approval necessary.

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**Author contribution**

All authors contributed in all the phases of preparing the manuscript.

**Consent**

Written informed consent was obtained from the patient for

**Table 1**  
Prognostic Factors of Ewing’s Sarcoma Tumors based on data adapted from Lahl M. et al., 2008.

Negative prognostic factors	Positive prognostic factors
1-Presence of metastases	1-Good radiologic/pathologic response to induction chemotherapy
2-Primary tumor site in the pelvis/central axis/proximal bone	2-Primary tumor site in distal bones or ribs
3-Large tumor volume	
4-Older than 17 years of age	
5-High lactate dehydrogenase levels (>200 IU/L have an increased probability of disease recurrence)	

publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Registration of research studies

Not applicable. Our manuscript is a case report.

#### Guarantor

Amjad Soltany, MD.

#### Declaration of competing interest

All authors confirm that there are no conflicts of interest to declare.

#### Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.amsu.2021.102407>.

#### References

- [1] A. Angelini, D.G. Letson, P. Ruggieri, Ewing's sarcoma of the sacrum, in: P. Ruggieri, A. Angelini, D. Vanel, P. Picci (Eds.), *Tumors of the Sacrum*, Springer, Cham, 2017, [https://doi.org/10.1007/978-3-319-51202-0\\_17](https://doi.org/10.1007/978-3-319-51202-0_17).
- [2] M.R. Farrokhi, F. Ranjbar, M.A. Mashari, M. Ardeshiri, Primary Ewing sarcoma of the sacrum, *Jentashapir J Health Res* 5 (1) (2014) 509–512.
- [3] C. Report, K.K. Kannan, R.J. Sundarapandian, V.J. Surulivel, Unusual Presentation of a Primary Ewing ' S Sarcoma of the Spine with Paraplegia : A Case Report, 2015, <https://doi.org/10.7860/JCDR/2015/11647.5663>, 1–3.
- [4] M. Lahl, V.L. Fisher, K. Laschinger, Ewing's sarcoma family of tumors: an overview from diagnosis to survivorship, *Clin. J. Oncol. Nurs.* 12 (1) (2008 Feb) 89–97, <https://doi.org/10.1188/08.CJON.89-97>. PMID: 18258578.
- [5] S. Srinivasalu, A.D. Souza, Sacral Ewing ' s sarcoma and challenges in it ' s diagnosis on MRI 3 (1) (2009) 23–26, <https://doi.org/10.3941/jrcr.v3i1.79>.
- [6] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, For the SCARE Group. The SCARE 2020 guideline: updating consensus surgical CAse REport (SCARE) guidelines, *Int. J. Surg.* 84 (2020) 226–230.