

# Atypical Presentation of Primary Non-metastatic Ewings Sarcoma of the Lumbosacral Spine-“Cauda Equina Syndrome” Clinically and a “Tuberculosis mimicking challenge” on MRI-A Case Report

Mohit Kukreja<sup>1\*</sup>, V G Telang<sup>2</sup>, Chetan Anchan<sup>3</sup>

<sup>1</sup>Orthopedic Research Fellow, Desert Orthopedic Centre, Las Vegas, NV, USA

<sup>2</sup>Dhanavantari Hospital & Research Centre, Mumbai

<sup>3</sup>Bombay Hospital & Research Centre, Mumbai

\*Corresponding author: [drmohitkukreja@gmail.com](mailto:drmohitkukreja@gmail.com)

**Abstract Introduction:** The Ewing family of tumors comprises Ewing's sarcoma (EWS), extraskeletal EWS, primitive neuroectodermal tumor (PNET) of bone and soft tissue, and chest wall tumor (Askin tumor). The translocation t(11; 22) (q24; q12) is identified in more than 90% of cases. EWS is the second most common primary bone malignancy in childhood. In contrast to long bone involvement, delays in spinal EWS diagnosis may occur because symptoms may not be present until neurological deficits occur. To date, although there have been reported cases of EWS in the lumbosacral region, the reported cases are very less and the presentations might drastically differ. **Case Presentation:** A 14 year old boy comes with a 12 weeks h/o urinary incontinence and constipation. Over the next 4 months, the patient had new-onset intermittent abdominal pain with worsening urinary symptoms. MRI confirmed a diagnosis of Koch's spine. However, the histopathology reported it as an “Ewings sarcoma/Primitive Neuroectodermal Tumor”. **Discussion:** Ewing's tumor of sacrum is rare, but should be suspected in low backache in children. Cauda equina syndrome can be a valid presentation for EWS spine. MRI can identify cases early and enables early treatment though it is not specific. Histopathological diagnosis is a must before any definitive management. EWS spine can mimic Tuberculosis both clinico-radiologically, only to be confirmed with a tissue biopsy. **Conclusion:** Ewing's sarcoma of the lumbosacral spine can have an atypical presentation and there should be a high degree of suspicion to diagnose it early.

**Keywords:** Ewing's, cauda-equina, lumbosacral

**Cite This Article:** Mohit Kukreja, V G Telang, and Chetan Anchan, “Atypical Presentation of Primary Non-metastatic Ewings Sarcoma of the Lumbosacral Spine-“Cauda Equina Syndrome” Clinically and a “Tuberculosis mimicking challenge” on MRI-A Case Report.” *American Journal of Medical Case Reports*, vol. 5, no. 8 (2017): 221-225. doi: 10.12691/ajmcr-5-8-5.

## 1. Introduction

Ewing's sarcoma was first described by James Ewing in 1921. It is a poorly differentiated tumor of uncertain histogenesis and variable biologic behavior. It is more common in Caucasians and rarely arises in individuals of African and Asian races.

The Ewing family of tumors comprises Ewing's sarcoma (EWS), extraskeletal EWS, primitive neuroectodermal tumor (PNET) of bone and soft tissue, and chest wall tumor (Askin tumor) [1,2,3,4]. The translocation t(11; 22) (q24; q12) is identified in more than 90% of cases of EWS which help differentiate it from other small, round blue cell tumors [1,2].

EWS is the second most common primary bone malignancy in childhood and adolescence with peak age from 15 to 17 years and a slight male predominance [8,9].

The occurrence in adult age is quite rare [1,7]. The most common primary sites of involvement are the pelvis, femur, tibia, and fibula.

In the primary vertebral EWS, the division of the spine into nonsacral (cervical, dorsal, and lumbar) and sacral (sacral and coccygeal) is important and is dictated by the different behavior of EWS in these two regions. Sacral EWS being more aggressive and less responsive to therapy [1,4,5,6,7,10].

In contrast to long bone involvement, delays in spinal EWS diagnosis may occur because symptoms may not be present until neurological deficits occur. To date, although there have been reported cases of EWS in the lumbosacral region, the reported cases are very less and the presentations might drastically differ.

We, therefore, present this case report with a dual diagnostic atypicality of EWS clinico-radiologically and basic principles to follow.

## 2. Presentation of the Case

A 14 year old boy comes with chief complaints of gradual onset irregularities in bowel and bladder. Initially presenting elsewhere, he had symptoms of urinary incontinence, polyuria and straining at micturition for a period of 3 months. He also gradually developed bowel complaints like constipation and occasional dyspepsia. There was no h/o fever, abdominal pain or pyuria. There was no history of nocturnal enuresis or significant psychosocial anecdotes. Diagnosed as a case of urinary tract infection, patient was started on empirical antibiotics and Xray & USG KUB ordered. Both investigations were normal at this stage.



Figure 1. XRAY PELVIS WITH BOTH HIPs of this patient showing - NORMAL STUDY

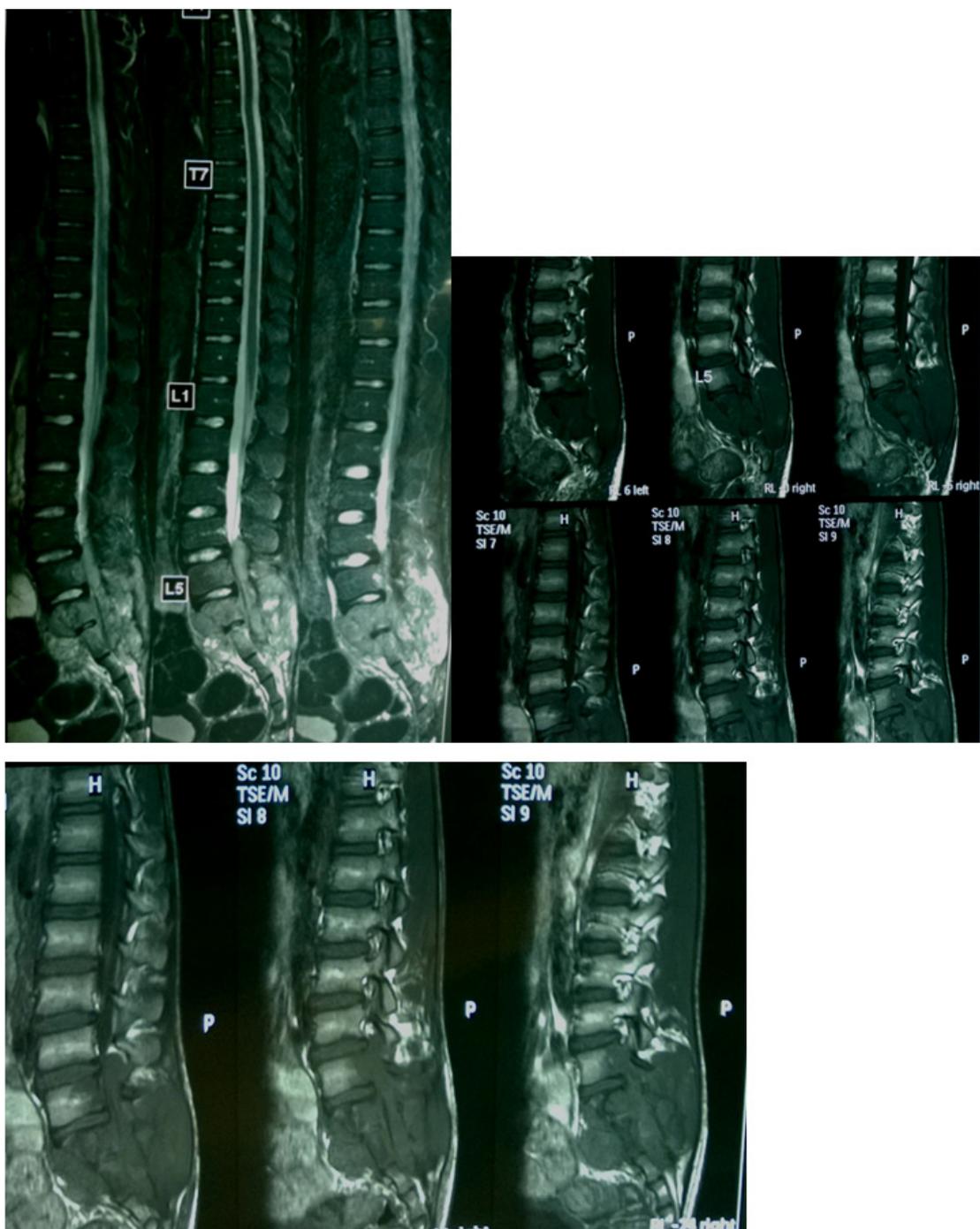
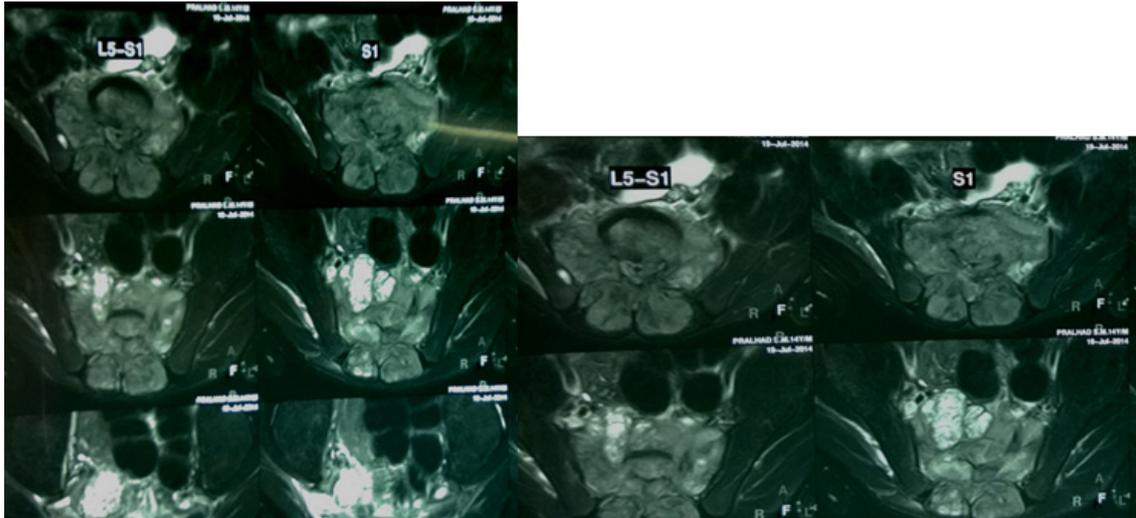


Figure-2. T1 AND T2 WEIGHTED SAGITTAL CUTS OF THE L-S SPINE showing a typical Tuberculosis-like picture with extensive soft tissue collection



**Figure 3. AXIAL CUTS OF MRI L-S SPINE** showing gross filling up of perisacral spaces

For the next 4 months, the patient also started complaining of intermittent abdominal pain along with worsening of the previous symptoms.

However, he also gradually started developing a swelling around the sacrococcygeal region with a spontaneous-onset skin sore over the swelling.

The parents, on a second opinion, were then advised an MRI-Lumbosacral spine (4 months after the first consultation). The following are the images on MRI.

The MRI suggested altered marrow signal intensity of the S1, S2 & S3 segments of both sides of the sacrum, coccyx and the vertebral body & posterior elements of L5, cortical erosions were seen involving these vertebrae, abnormal soft tissue mass seen in the presacral space invading bilateral neural foramina of L4-L5, L5-S1 & S1, 2, 3. There was epidural soft tissue component from L5-S2 levels and posterior paraspinal soft tissue mass from L3-S3 levels. These findings were suggestive of Infective etiology and Koch's (Extensive Tuberculosis) was emphatically given as the diagnosis. Following this, the patient was empirically started on Anti-Koch's 4-drug treatment regimen. Despite of a regular compliant drug administration, the symptoms of the boy worsened and the sacral swelling increased.

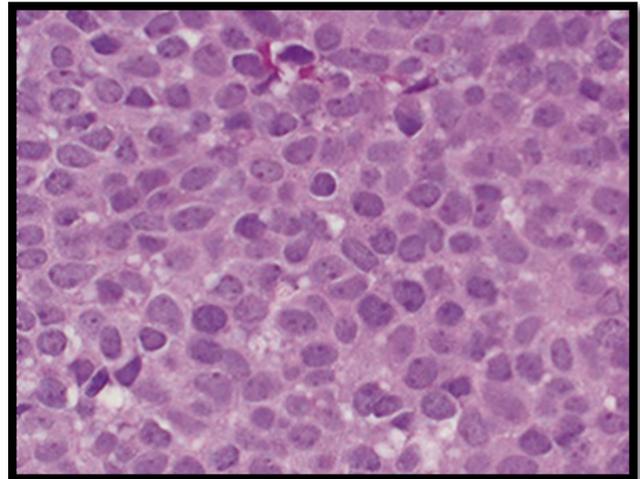
It was at this stage that the child presented to us (6 months after the first consultation and on 4-drug AKT since last 2 months). On a detailed clinical examination, there was a 4×4 cm soft, fluctuant boggy swelling in the sacrococcygeal region with a 1×1 cm superficial sore. There were no signs of acute inflammation. This was associated with sacral anaesthesia at the S2, 3, 4 levels with loss of the anal reflex. There was a motor weakness of Grade 4/5 according to the MRC Grading with no evidence of sensory loss in the lower limbs. Ankle reflex was poor(+). Bladder symptoms still persisted despite a 2 month compliant anti-tuberculous treatment. Blood investigations were nearly normal (Hb=11.4gm/dl, TLC=9400/cubic-mm).

A Chest Xray was clear. A review with 2 other Radiologists reconfirmed a typical picture of Koch's Spine. The clinical scenario of a cold-abscess like swelling in the sacrococcygeal junction supported the imaging findings. Thus, it seemed to be like an atypical presentation of Tuberculosis spine presenting as a Cauda-equina syndrome and required emergent decompression.

However, we decided to first confirm our diagnosis with a tissue biopsy as we were still not convincingly pursuing it as an atypical Koch's affection. Also, a review USG-KUB now gave a picture of Bilateral hydronephrosis with hydro-ureter with a distended, trabeculated and thickened bladder.

We decided to do a J-needle biopsy with a small 0.5cm incision which would help us acquire some sacral bone scrapings also. As soon as the skin was nicked, there was a constant gush of blood from the mass with little evidence of pus/cheesy material.

Histopathological diagnosis was "Ewings sarcoma/Primitive Neuroectodermal Tumor" with strongly positive CD-99 immunoreactivity and positive Mic-2, Vimentin & synaptophysin on Immunohistochemistry.



**Figure 4. HISTOPATHOLOGY** showed densely populated round cells arranged in sheets with a typical "blue tumor" appearance. The tumor islands are separated by fibrous stroma

The child was started with Chemotherapy on an urgent basis with further referral to a specialized Cancer Institute.

### 3. Discussion

Tumors belonging to the Ewing family are devastating malignancies that appear to be more common than it has

previously been reported [9]. The peak age for EWS is 15 to 17 years, being rarer in adults [1,7,8,9].

Primary Ewing's sarcoma of spine has male preponderance with posterior elements of the spine being commonly involved and are mostly extradural-extramedullary. The incidence in the sacrum is rare compared to other parts of the spine. Complete surgical resection is difficult most of the times. Distant metastasis is seen in 38 % cases, being commonest to the lungs.

EWS remains for several aspects an anomalous tumor with bizarre behavior. Hense et al. described an unexplained, intrinsic major aggressive behaviour of axial tumors [11].

Animals studies confirmed that axial and appendicular EWSs behave differently [1,2,11,12]. These data may collectively suggest biologic differences between axial and appendicular EWSs that ultimately result in prognostic differences.

There are certain strong basic interpretations that can clearly be drawn from this single case report.

1. Ewing's sarcoma can have a varied clinical presentation and should be suspected in all grossly destructive spine lesions in the pediatric age group. Cauda-equina syndrome, although a rare presentation, is a possible symptom complex in this disease. So far 15-20 cases of spinal PNET have been reported presenting with Cauda equine syndrome [13,14,15].

2. In Ewing's sarcoma of the sacrum, destructive lesions involving vertebrae with extra dural space involvement may be present and can be depicted on MRI scan. However MRI is non-specific [16].

Also, in fact MR imaging can be a prominent cause of misdiagnosis because EWS may very closely mimic Tuberculosis on its T1 and T2 W images. The affection, although bony primarily, may generate extensive soft tissue collection (akin a cold abscess) making it even more difficult to differentiate it from tuberculosis. We consulted 2 prominent radiologists for review of the films and they were confident about Koch's as a diagnosis and refrained from suggesting an apparent differential to it.

3. The starting of empirical AKT drugs just on the basis of imaging techniques is a very common practice adopted especially in a country like India where Tuberculosis as a disease is rampant. In this case, we suppose that a lot of crucial time was lost after AKT was started just on an imaging diagnosis and a sequential tissue biopsy should have immediately followed suit at that point for a Koch's spine diagnosis. Delayed confirmation of a malignancy like EWS as a diagnosis was grossly erroneous in the management timeline of this patient. Thus, this reinforces the significance of the traditional principle dictating a histopathological confirmation everytime Koch's is suspected and making no presumptions conjecturally.

4. EWS of the spine is a rare entity but a definite possibility especially if the disease seems grossly destructive. The cause of our inhibitions for not pursuing this as an extensive Koch's affection was the mass involvement of sacrum, presacral/posterior sacral spaces and coccygeal involvement.

5. Planning a staged mini-incision tissue biopsy/bone scrapings is always preferable over direct definitive management without a histopathological back-up. This

reinforces the significance of a pathological confirmation as the first step towards an unknown disease process.

## 4. Highlights & Teaching Points

1. Ewing's tumor of sacrum is rare, but should be suspected in low backache in children.
2. Cauda equina syndrome can be a valid presentation for EWS spine.
3. MRI can identify cases early and enables early treatment though it is not specific.
4. Histopathological diagnosis a must before any definitive management (conservative/surgical).
5. EWS spine can mimic Tuberculosis both clinico-radiologically to the extent that only a tissue biopsy can differentiate them.

## 5. Conclusion

Ewing's sarcoma of the lumbosacral spine can have an atypical presentation and there should be a high degree of suspicion to diagnose it early.

We are hereby declaring that this work had been prepared and edited in accordance with the CARE guidelines of making, preparing, writing and publishing a Case Report [17].

## Abbreviations

MRI- Magnetic Resonance Imaging  
 EWS-Ewings Sarcoma  
 USG-Ultrasonography  
 KUB-Kidney, Ureter, Bladder  
 L-S-Lumbosacral  
 AKT-AntiKoch's (Anti-tuberculous) treatment  
 MRC-Medical Research Council  
 PNET-Primitive Neuroectodermal Tumor.

## References

- [1] J. A. Ellis, R. J. Rothrock, G. Moise et al., "Primitive neuroectodermal tumors of the spine: a comprehensive review with illustrative clinical cases," *Neurosurgical Focus*, vol. 30, no. 1, pp. 1-13, 2011.
- [2] L. Venkateswaran, C. Rodriguez-Galindo, T. E. Merchant, C. A. Poquette, B. N. Rao, and A. S. Pappo, "Primary Ewing tumor of the vertebrae: clinical characteristics, prognostic factors, and outcome," *Medical and Pediatric Oncology*, vol. 37, no. 1, pp. 30-35, 2001.
- [3] C. Rodriguez-Galindo, S. L. Spunt, and A. S. Pappo, "Treatment of ewing sarcoma family of tumors: current status and outlook for the future," *Medical and Pediatric Oncology*, vol. 40, no. 5, pp. 276-287, 2003.
- [4] K. Fizazi, N. Dohollou, J. Y. Blay et al., "Ewing's family of tumors in adults: multivariate analysis of survival and outlook results of multimodality therapy in 182 patients," *Journal of Clinical Oncology*, vol. 16, no. 12, pp. 3736-3743, 1998.
- [5] S. J. Cotterill, S. Ahrens, M. Paulussen et al., "Prognostic factors in Ewing's tumor of bone: analysis of 975 patients from the European Intergroup Cooperative Ewing's Sarcoma StudyGroup," *Journal of Clinical Oncology*, vol. 18, no. 17, pp. 3108-3114, 2000.

- [6] G. Bacci, S. Ferrari, F. Bertoni et al., "Prognostic factors in nonmetastatic Ewing's sarcoma of bone treated with adjuvant chemotherapy: analysis of 359 patients at the Istituto Ortopedico Rizzoli," *Journal of Clinical Oncology*, vol. 18, no. 1, pp. 4-11, 2000.
- [7] H. Ilaslan, M. Sundaram, K. K. Unni, and M. B. Dekutoski, "Primary Ewing's sarcoma of the vertebral column," *Skeletal Radiology*, vol. 33, no. 9, pp. 506-513, 2004.
- [8] A. A. Gupta, A. Pappo, N. Saunders et al., "Clinical outcome of children and adults with localized ewing sarcoma: impact of chemotherapy dose and timing of local therapy," *Cancer*, vol. 116, no. 13, pp. 3189-3194, 2010.
- [9] G. Gatta, J. M. Van Der Zwan, P. G. Casali et al., "Rare cancers are not so rare: the rare cancer burden in Europe," *European Journal of Cancer*, vol. 47, no. 17, pp. 2493-2511, 2011.
- [10] K.R. Weiss, D.J. Biau, and R. Bhumbra, "Axial skeletal location predicts poor outcome in Ewing," *Sarcoma*, vol. 2011, Article ID 395180, 5 pages, 2011.
- [11] H. W. Hense, S. Ahrens, M. Paulussen, M. Lehnert, and H. Jürgens, "Factors associated with tumor volume and primary metastases in Ewing tumors: results from the (EI) CESS studies," *Annals of Oncology*, vol. 10, no. 9, pp. 1073-1077, 1999.
- [12] M. J. A. Sharafuddin, F. S. Haddad, P. W. Hitchon, S. F. Haddad, and G. Y. El-Khoury, "Treatment options in primary Ewing's sarcoma of the spine: report of seven cases and review of the literature," *Neurosurgery*, vol. 30, no. 4, pp. 610-619, 1992.
- [13] Dhatt S, Dhillon MS, Tripathy SK, Goyal T, Jagadeesh V. Peripheral primitive neuroectodermal tumor causing cauda equina syndrome with destruction of L5 vertebra. *Indian J Orthop*, 44, pp. 339-41, 2010.
- [14] Jithendra K S, rachna S ,karak AK ,Thavaraj V, Kabra SK. Primitive Neuroectodermal Tumor presenting as Cauda equine syndrome. *Indian J Med and Paediatr Oncol*, 28, pp. 34-37, 2007.
- [15] Krishnakumar R, Renjitkumar J. Acute cauda equina syndrome due to primary Ewing's sarcoma of the spine. *Neurol India*, 59, pp. 931-3, 2011.
- [16] Llauger J, Palmer J, Amores S, Bague S, Camins A. Primary Tumors of Sacrum-Diagnostic Imaging. *American Roentgen Ray Society*. 174, pp. 417-24, 2000.
- [17] Gagnier, Joel J., et al. "The CARE guidelines: consensus-based clinical case reporting guideline development." *Journal of medical case reports*, 7.1, pp. 23, 2013.