Case Report:

Paraspinal Extraosseous Ewing's Sarcoma With Disseminated Metastases Masquerading As Pott's Spine

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Abstract:
A 16 years old girl presented with low back pain, fever and weakness in both lower limbs for 3 months. Plain radiograph shows lytic lesion in first sacral vertebra and multiple lung nodules. In suspicion of tuberculosis, antitubercular drugs were advised. But magnetic resonance imaging revealed right paraspinal soft tissue mass with multiple lesions in several vertebrae causing spinal canal compression alongwith lesions in iliac bones, sacrum, lung parenchyma and scalp tissue. Fine needle aspiration cytology and biopsy revealed Ewing's sarcoma. Patient was referred to higher centre for chemotherapy/radiotherapy. Herein we report this rare case of extraosseous Ewing's sarcoma with disseminated metastases masquerading as Pott's spine.

Keywords: Ewing's sarcoma, extraosseous, metastasis, Spine, Magnetic resonance imaging

Introduction:

Ewing's sarcoma (ES) is a primary bone malignancy commonly involving long bones of the extremities and the pelvis. Primary malignant sarcomas of the spine are rare and accounts for only 3.5%–14.9% of all primary bone sarcomas and extraosseous Ewing's sarcoma are even rarer[1]. In 30% cases ES is multicentric in origin and approximately 25% of patients will have metastatic disease at the time of diagnosis with 10% lung, 10% bones/bone marrow, 5% combinations or others[2, 3, 4]. Neurological deficits due to spinal cord compression are often a delayed presentation. Extraosseous ewing's sarcoma can be seen in various locations like central nervous system, chest wall, retroperitoneum, kidney, pelvis, rectum, vagina, skin, small intestine, thumb, finger, arm, scalp, nasal fosse, lip, toe, para vertebral and perineum[5].

We report a rare case of paraspinal extraosseous Ewing's sarcoma with disseminated metastases in a 16 years old female masquerading as spinal tuberculosis (Pott's spine).

Case presentation:

A 16 years old female presented with a 3 months history of low back pain, low grade fever, weight loss and weakness in both lower limbs. She had developed retention of urine for one week and also mild intermittent headache with swelling over the vertex of head. There was no history of trauma. Blood investigations revealed elevated erythrocyte sedimentation rate (ESR). Montoux test was found to be negative. Physical examination revealed paraparesis in both lower limbs and right paraspinal tenderness.

Plain radiograph of lumbosacral spine shows expansile lytic lesion in the first sacral vertebra (S1) and chest radiograph (postero-anterior view) show multiple lung nodules. On the basis of clinical diagnosis of tuberculosis of spine, antitubercular drugs were started but no improvement was

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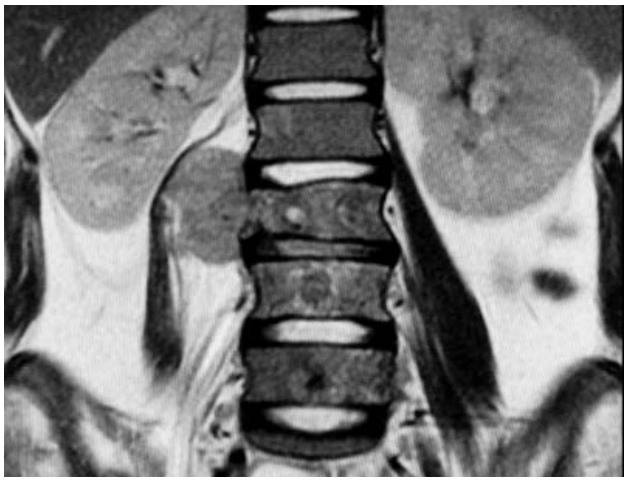
seen. To rule out any underlying malignancy, magnetic resonance imaging (MRI) of thoracolumbar spine and brain was done in axial, sagittal and coronal planes by taking T1-weighted, T2-weighted and STIR (short tau inversion recovery) and contrast enhanced T1-weighted sequences.



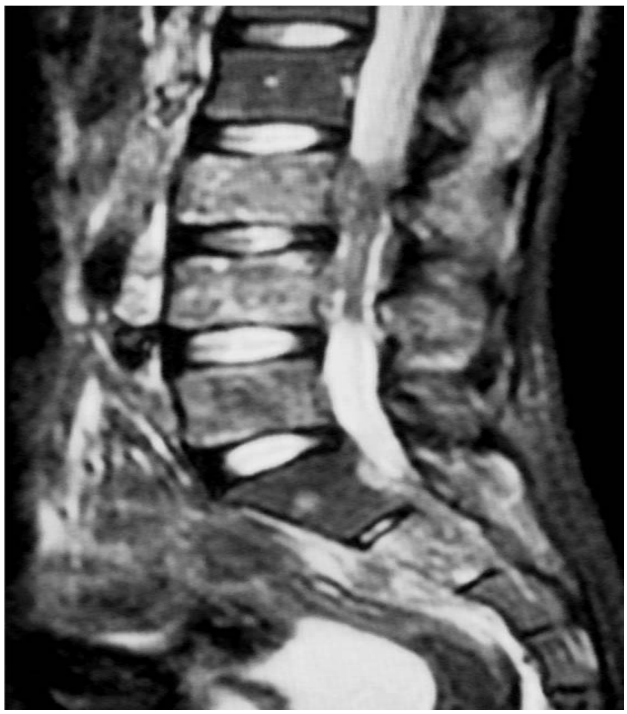


Figure 2a and 2b: *Contrast enhanced T1 weighted sagittal MR image of spine showing multiple patchy heterogeneously enhancing lesions in the L3-L5 (Figure 2a) and D6-D8 (Figure 2b) vertebral bodies with epidural soft tissue component. Heterogeneously enhancing expansile lesions also seen in S1 vertebral body (Figure 2a), manubrium and body of sternum (Figure 2b).*

MRI revealed a mild heterogeneously enhancing right paraspinal mass of size 3.5x3 cm, abutting and displacing the right psoas muscle (Figure 1).

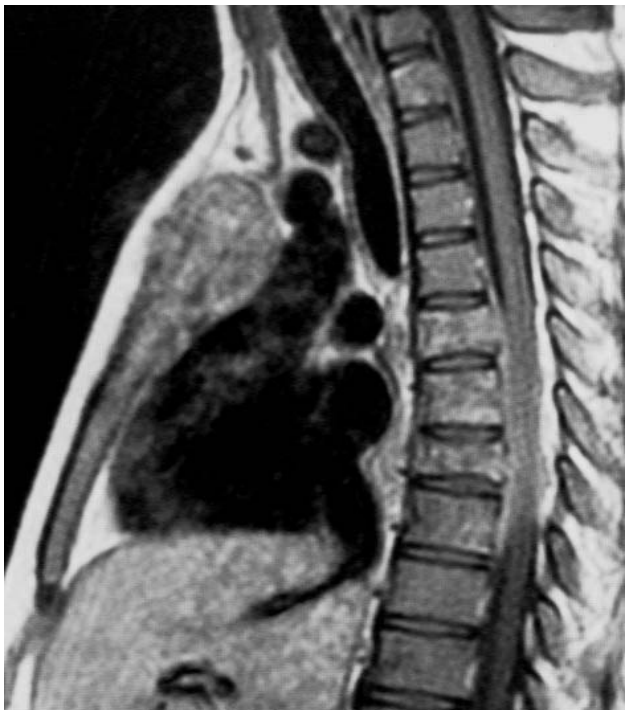


Figure II: *Contrast enhanced T1 weighted coronal MR image showing mildly enhancing mass in right paraspinal region.*

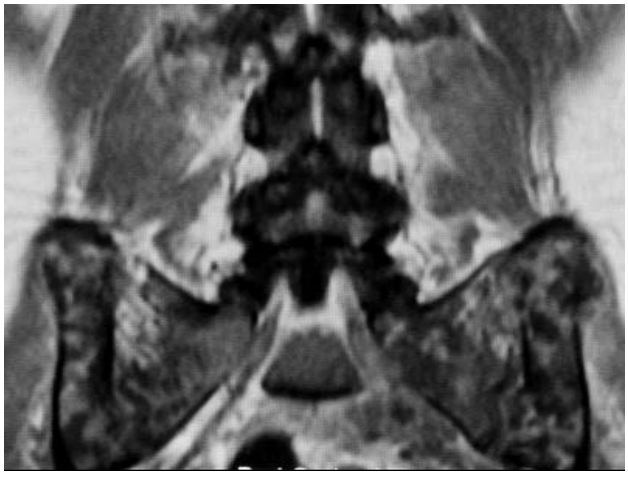


Figure III: *Contrast enhanced T1 weighted coronal MR image of pelvis showing multiple patchy heterogeneously enhancing lesions in bilateral iliac bones.*

Multiple patchy heterogeneously enhancing lesions seen in the D6, D7, D8, L3, L4 and L5 vertebral bodies with epidural soft tissue component causing spinal canal compression (Figure 2a and 2b). Similar signal intensity lesions also seen in bilateral iliac bones (Figure 3). Heterogeneously enhancing expansile lesions seen in S1 vertebral body (Figure 2a), manubrium and body of sternum (Figure 2b). Multiple small hyperintense lesions on contrast enhanced T1W images seen in bilateral lung parenchyma (Figure 4). A well defined

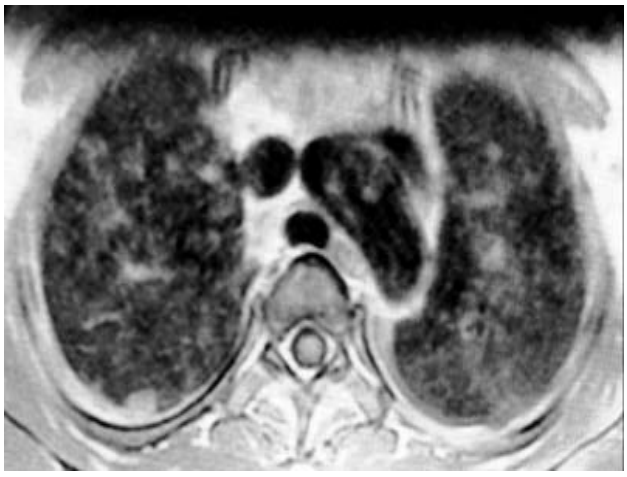


Figure IV: *Multiple hyperintense lesions in bilateral lung parenchyma on contrast enhanced T1W axial MR image of chest.*

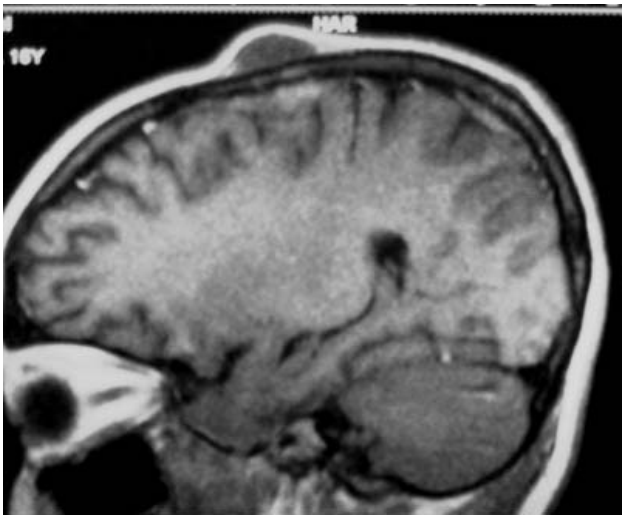


Figure 5: *Contrast enhanced T1W sagittal MR image of head showing a well defined peripherally enhancing lesion in the scalp tissue in temporo-parietal region.*

peripherally enhancing lesion also seen in the scalp tissue in temporo-parietal region (Figure 5); however brain parenchyma was normal.

Fine needle aspiration cytology (FNAC) from the right paraspinal soft tissue mass and biopsy from vertebra revealed malignant round cell tumor that was confirmed as Ewing's sarcoma by immunohistochemistry. She was referred to oncology centre for chemotherapy/ radiotherapy. On follow-up after 3 months, there was improvement in the symptoms.

Discussion:

Ewing's sarcoma (ES) accounts for nearly 4% of pediatric malignancies and is the second most common primary bone tumor in children[6]. It has the highest incidence in the children and young adults (median age is 15 years) and is more common in male than female[7]. The etiology of ES is not well understood, however some authors have described it origin from neural crest cells in bone marrow (small round cell tumor) and associated with t (11; 22) translocation.

Primary sites of ES in decreasing order are lower extremity, pelvis, chest wall, upper extremity, spine, scapula, ribs and skull. For extraosseous primary tumors, the most common primary sites are trunk, extremity, head and neck, retroperitoneum and other sites[8]. Based on the differences in the treatment responses and survival rates, primary vertebral ES is of two types i.e. sacral and nonsacral[8]. Clinically it presents with local pain, neurological deficit and palpable mass. ES disseminates by hematogenous route. Osteosarcoma, choriocarcinoma, melanoma and ES frequently metastasize to the lung, although the frequency of these primary tumors is relatively low[9]. On plain radiography, features of ES occur late and the most common finding is lytic lesions in vertebra[10]. The common variety is focal lytic lesion followed by complete flattening of the vertebral body i.e. vertebra plana[11]. Sclerotic changes of vertebra are also rarely seen. Computed tomographic (CT) scan has important role in evaluating the bony destruction (both vertebral bodies and posterior elements), soft tissue component and also in CT guided needle biopsy from vertebral or paravertebral lesions.

MRI is the best imaging modality for early detection of tumor, epidural compression, spread to the bone marrow and extension into adjacent soft tissues. It has the highest accuracy in determining the response to chemotherapy[12]. ES appears hyperintense to surround soft tissue on T2-weighted imaging and hypointense with heterogeneous enhancement on T1-weighted imaging. ES is also multifocal; hence skeletal scintigraphy is an important tool to exclude other foci and also used for follow-up treatment[13]. Chronic infective pathologies (pyogenic or tubercular) can be well differentiated from malignancy on MRI.

Histological features of ES are clusters of

monomorphic tumor cells with round vesicular nuclei and dissociated anucleated cells. The differential diagnosis of small round cell tumor includes ES, neuroblastoma, primitive neuroectodermal tumor (PNET), lymphoma and rhabdomyosarcoma. Light microscopic, ultrastructural and immunohistochemical features differentiate ES from other tumors[14]. It is difficult to differentiate ES from other tumors on histologically alone, but in our case the histologic criteria was sufficient to diagnose ES.

Multimodality treatment approach i.e. surgery, chemotherapy and radiotherapy is required for long-term outcome. Prognosis of patients with metastatic disease is poor. Predictors of poor outcome in metastatic disease are age more than 14 years, volume tumor more than 200 ml, more than one site of bony metastasis, bone marrow metastases and additional lung metastases[15-16].

Conclusion:

This case report concludes that extraosseous Ewing's sarcoma with disseminated metastases may masquerades as chronic infection. Patient in first two decades of life presenting with severe local pain and tenderness not relieved by bed rest, the clinicians should evaluate for the possibility of underlying malignancy, even if plain radiograph is normal. Multimodality treatment approach with strict follow-up is essential for successful management of Ewing's sarcoma but the prognosis still remains poor.

References:

1. Bacci G, Picci P, Gherlinzoni F, Capanna R, Calderoni P, Putti C, et al. Localized Ewing's sarcoma of bone: Ten years' experience at the Instituto Ortopedico Rizzoli in 124 cases treated with multimodal therapy. *Eur J Cancer Clin Oncol*. 1985; **21**:163-73. http://dx.doi.org/10.1016/0277-5379(85)90168-3

2. Miller RW. Contrasting epidemiology of childhood osteosarcoma, Ewing Sarcoma and Rhabdomyosarcoma. *National Cancer Institute Mono¬gram.* 1981; **56**: 9-14.

3. Esiashvili N, Goodman M, Marcus RB. Changes in incidence and survival of Ewing sarcoma patients over the past 3 decades: Surveillance Epidemiology and End Results data. *J Pediatr Hematol Oncol*; **30** (6): 425-30, 2008. http://dx.doi.org/10.1097/MPH.0b013e31816e22f3PMid:18525458

4. Bernstein ML, Devidas M, Lafreniere D et al. Intensive therapy with growth factor support for patients with Ewing tumor metastatic at diagnosis: Pediatric Oncology Group/Children's Cancer Group Phase II Study 9457-a report from the Children's Oncology Group. *J Clin Oncol* 2006; **24**:152-159. http://dx.doi.org/10.1200/JCO.2005.02.1717PMid:16382125

5. Miller ME, Emerson L, Clayton F, Bentz BG, Data RE, Salzman KL, et al. Extraosseous Ewing's sarcoma. *J Clin Oncol* 2007; **25**:4845-8.http://dx.doi.org/10.1200/ JCO.2007.13.0930 PMid:17947735

6. Ebb DH, Green DM, Shamberger RC, Tarbell NJ. Solid tumours of childhood. In: DeVita VT Jr, Hellman S, Rosenberg SA, editors. Cancer *Principles and Practice of Oncology*. *Philadelphia: Lippincott*; 2005:**19**19-23.

7. Morandi X, Riffaud L, Haegelen C, Lancien G, Kerbrat P, Guegan Y. Extraosseous Ewing's sarcoma of the spinal epidural space. *Neurochirurgie*. 2001; **47**(1):38-44. PMid:11283454

8. Raney RB, Asmar L, Newton WA, et al. Ewing's

sarcoma of soft tissues in childhood: a report from the Intergroup Rhabdomyosarcoma Study, 1972 to 1991. *J Clin Oncol*. 1997; **15** (2): 574-82.PMid:9053479

9. Paulussen M, Ahrens S, Craft AW, et al. Ewing's tumors with primary lung metastases: survival analysis of 114 (European Intergroup) Cooperative Ewing's Sarcoma Studies patients. *J Clin Oncol*. 1998; **16**(9):3044-52. PMid:9738574

10. Sharafuddin MJ, Haddad FS, Hitchon PW, et al. Treatment options in primary Ewing's sarcoma of the spine: report of seven cases and review of the literature. *Neurosurgery*. 1992; **30**:610-618. http://dx.doi.org/10.1227/00006123-199204000-00025PMid:1374853

11. Papagelopoulos PJ, Currier BL, Galanis E, et al. Vertebra plana caused by primary Ewing sarcoma: case report and review of the literature. *J Spinal Disord Techni*. 2002; **15**:252-257. http://dx.doi.org/10.1097/00024720-200206000-00015

12. Erlmann R, Sciuk J, Bosse A, Ritter J, Kusnierz-Glaz CR. Response of osteosarcoma and Ewing's sarcoma to preoperative chemotherapy: assessment with dynamic and static MR imaging and skeletal scintigraphy. *Radiology*. 1990; **175**:791-796.

13. Estes DN, Magill HL, Thompson EI, Hayes FA. Primary Ewing's sarcoma:follow-up with Ga-67 scintigraphy. R*adiology*. 1990; **177**:449-453. PMid:2217783

14. Triche TJ. Diagnosis of small round cell tumors of childhood. *Bull Cancer*. 1988; **75**:297-310. PMid:3285915

15. J Mohd Tahir, KN Gopalan, MB Marina, SHA Primuharsa Putra. A rare case of laryngeal Kaposi's sarcoma. *Bangladesh Journal of Medical Science*. 2010; **09** (2): 107-109. DOI: 10.3329/bjms.v9i2.5660

16. Ladenstein R, Pötschger U, Le Deley MC, et al.: Primary disseminated multifocal Ewing sarcoma: results of the Euro-EWING 99 trial. *J Clin Oncol*. 2010; **28**(20): 3284-91.http://dx.doi.org/10. 1200/JCO.2009.22.9864PMid:20547982