Case Report

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Ewings Sarcoma Presenting as an Extra Dural Lumbosacral Lesion: A Rare Case Report

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Abstract

Primary malignant sarcomas of the spine are extremely rare. Because of biological heterogeneity, these tumors have variable sensitivity to radiation and chemotherapy. Presentation of severe radicular pain with MRI findings of an exradural lesion with extension through the neural foramen is rare to be an ewings sarcoma in any neurosurgeons carrier. We report about an eighteen years old female with primary Ewing´s sarcoma of the lumbosacral region which has been reported only a few times in literature.

Keywords: Ewings Sarcoma; Primitive Neuro Ectodermal Tumor; Spine.

Case Report

A eighteen year old married female presented in our opd with complains of severe low back pain with radiation to the posterior part of lower limb for six months and on examination was found to have weak extensor hallucis longus in left side and 4+/5 power in hip extension and knee flexion and dorsiflexion of ankle. There were no bowel or bladder complaints or sensory loss on examination. On MRI she was found to have an extra dural lesion towards the left side of spinal canal from L5 to s2, extending through s1, s2 neural foramina to left para vertebral region causing expansion of neural foramina. Patient was operated by L5, s1, s2 laminectomy with gross total removal of left sided l5 to s2 extra dural sol with neural formainal extension under general anesthesia.

Post operatively, her neurological findings were same as preoperatively and she was relatively relieved of symptoms for a short time. On histopathological examination findings of small round cell tumor with areas of necrosis favoring non Hodgkin's lymphoma or pnet was found and on immunohistochemical examination strong membrane positivity for CD 99 along with CD45 negativity for tumor cells was found. These features favoured Ewing's sarcoma or primitive neuronal ectodermal tumor.

The whole process of obtaining biopsy and immunohistochemistry report took around 3 months as we lost her on follow up. After the availability of reports, contrast enhanced ct scan of abdomen, pelvis and thorax was made and multiple secondary lesions in lungs were detected

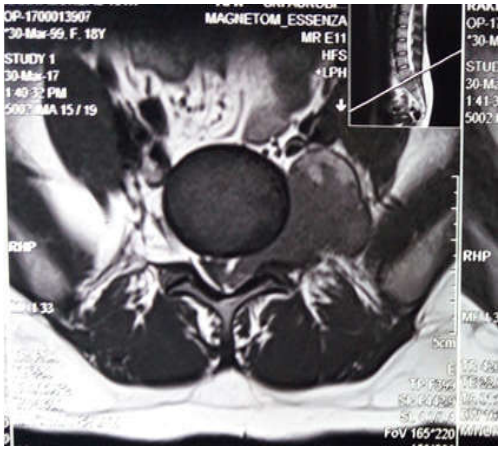

Fig. 1: Axial t 2 mri film showing a hypointense lesion in the left l5 s 1 region which traverses neural foramina and extends in to para vertebral region

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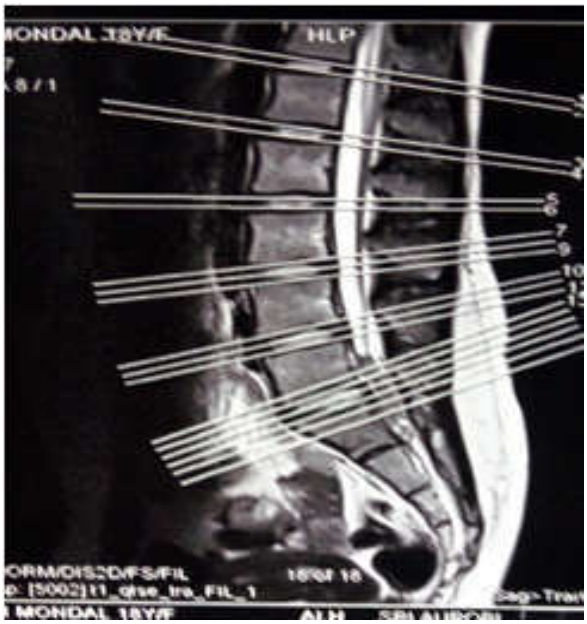

Fig. 2: A sagittal t2 mri film showing an hypointense space occupying lesion in the spinal canal at the region of l5 s1

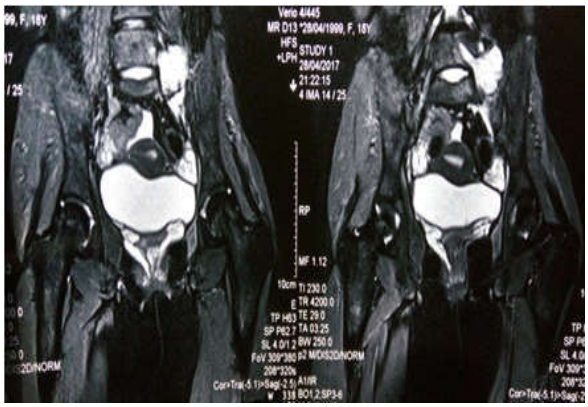

Fig. 3: Coronal section t2 flair mri showing the lesion which extends through left l5 neural foramina

along with fluid collection in abdomen. Patient is about to be started on chemotherapy regimen while writing this report.

Discussion

Undifferentiated round cell tumors include inhomogeneous group of malignant tumors which may arise in any organ because of their embryonic origin. As according to the 2007 CNS WHO classification a subgroup of these tumors named "PNET" may affect the CNS primarily [1,2]. The origin of PNET in CNS (CNS PNET) is found to be the matrix or germinal cells of the embryonic neural tube eg. medulloblastoma. The generic term 'embryonal tumours' has replaced CNS primitive neuroectodermal tumours (CNS-PNET) in the WHO 2016 classification. This separates the current terminology from historical concepts of PNET covering a diverse range of CNS tumours (medulloblastoma and supratentorial PNET) and from potential confusion from tumours of the same name arising outside the nervous system (e.g. Ewing's type sarcoma/peripheral PNET). Primary spinal primitive neuroectodermal tumors (PNET) and/or spinal Ewing's sarcoma family tumors are rare lesions appearing in the spinal extradural space [3,4].

Although CD99 shows crisp and strong membrane positivity in Ewings sarcoma Family Tumors, it can also be positive in other tumors viz. lymphoblastic lymphoma, rhabdomyosarcoma, synovial sarcoma, mesenchymal chondrosarcoma, blastemal component of Wilms tumor, and rarely in DSRCT [5]. Hence, a panel of immunohistochemical stains is employed to arrive at a definitive diagnosis. As stated above, CD99, FLI1, and NSE would be positive in ES/PNET. Non-Hodgkin lymphoma would express the lymphoid markers, i.e., CD45RB, CD3, CD20, and TdT; neuroblastoma would be positive for neuroendocrine markers (synaptophysin, chromogranin); rhabdomyosarcoma would be positive for skeletal muscle markers viz., desmin, myogenin, myo-D1, and myoglobin; and synovial sarcoma would also express pancytokeratins, EMA, BCL2, and calponin [6].

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, being sacral EWS more aggressive and less responsive to therapy [7,8-11]. Spinal involvement by EWS most commonly results from metastasis in advanced stages. Ewing sarcoma originating from the spine is rare, and extremely rare if the sacrum is excluded. Most studies on EWS involving the mobile spine are limited to case reports [7,10-12].

The most common symptom is muscle weakness. Other symptoms like sensory symptoms, local pain, and radiculopathy are also common common in primary intraspinal ewings sarcoma and most of them occurs in lumbar spine. Extra CNS metastasis occurs frequently, with lung being the most common site, followed by the skeletal system [13].

Definitive management of ES includes surgery, radiotherapy and chemotherapy. Given the low incidence of vertebral disease, there are no guidelines outlining optimal management. However a multitude

of therapeutic strategies have been employed with varying success. Initial chemotherapy is administered before local treatment with the aim to shrink bulky and unresectable tumors and eradicate micrometastases. Initial chemotherapy is also administered for acute relief of epidural compression. When there is epidural compression secondary to ES with rapidly progressing neurological symptoms or impending paralysis, prompt surgical intervention is the only primary alternative if an irreversible deficit is to be prevented [14].

Approximately 25% of patients present with metastatic disease at diagnosis. Patients with metastatic disease confined to the lung have a better prognosis than do patients with extrapulmonary metastatic sites [15,16]. The number of pulmonary lesions does not seem to correlate with outcome, but patients with unilateral lung involvement do better than patients with bilateral lung involvement [17].

Indications for surgery are (1) localized spine mass in patients with neurological symptoms; (2) primary instability or cases with extensive bony involvement where the instability is likely to occur after tumor necrosis resulting from treatment; (3) poor response to initial treatment with chemotherapy or radiotherapy; (4) residual disease; (5) sacral tumors where radical surgical resection is indicated by the aggressive biological behavior and poor prognosis of these lesions [18,19,20].

Chemotherapeutic agents used are ifosfamide, etoposide, vincristine, adriamycin, cyclophosphamide and actinomycin-D, and the regimen is the same as for ES at any other site as data do not strengthen the need for a specific protocol for unusual site ES [21,22]. Because of the anatomical constraints of the spine and neighbouring critical structures, and concerns about long term morbidity and functional loss in en bloc resection, spinal ES is often treated by intralesional excision or debulking. In a series of patients aggressively treated with surgery, Boriani et al. have reported that only en bloc resection with acceptable margins was associated with better local control and survival. Wherever tumor margins had to be compromised to preserve function, the outcome was not as good [23].

Radiation doses for spinal tumors should not exceed the limit of cord tolerance (45 Gy) to avoid the risk of radiation induced myelopathy and secondary sarcomas. The lower survival rates reported with radiation therapy compared with surgery for local control might be related to residual nests of viable tumor within the radiated site, as potential sources of local recurrence or distant relapse [21].

Data from the 30-year Surveillance, Epidemiology, and End Results Registry (SEER) has placed the 5-year and 10-year survival rates of spinal ES at 41% and 34%, respectively [10]. In an institutional study of non-metastatic ES, tumors primarily located in the sacrum had much poorer outcome than those located in the rest of the spine and concluded that regardless of the type of local treatment even when associated with neoadjuvant therapy, ESFT in the spine and sacrum has a poor outcome and prognosis is significantly worse than that of primary ESFT in other sites [21].

Fine needle aspiration cytology is a procedure highly underestimated and is an effective tool in vertebral masses and those with significant paraspinal extension like Potts spine and secondary spine. As mentioned by Aggarwal etal the procedure is quick and inexpensive and could be performed as an outdoor procedure. The procedure has almost no complications and over 75% positive results. In some of the cases the FNAC can give definite diagnosis but by no means it can replace the usefulness and accuracy of the open biopsy. The study is just an example of using the FNAC in diagnosing spinal lesions where diagnosis is in doubt prior to open biopsy [24].

In this case preoperative FNAC would have helped us with a neo adjuvant chemotherapy cycle and might have helped for total resection of tumor and thus better results.

This case also predicts the poor outcome in sacral origin ewings sarcoma and the need for early chemotherapy and local radiotherapy.

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