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Original Research Paper

Extradural lumber Ewing's sarcoma in an adult female: A rare case report

Neurosurgery

KEYWORDS: Extraskeletal Ewing's Sarcoma, lumbar Spine, Radicular pain, Extradural tumour, Immunohistochemistry.

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ABSTRACT

Extraskeletal Ewing's Sarcoma constitutes about 6% of all primary malignant tumours (1). However, extradural presentation involving the lumbar spine is a very rare finding. In this report, we discuss a primary lumbar spine extraskeletal Ewing's sarcoma in a 39 years old woman who presented with chronic radicular pain and proximal muscle weakness of right leg which is a rare finding of extradural lesion affecting L2-L5 vertebra and spreading paraspinally along its nerve roots through the intervertebral foramina and involving the psoas muscle. Imaging studies revealed the possibility of a neurofibroma, a benign tumour. After surgical decompression and postoperative histopathological study the diagnosis of Extradural Ewing's sarcoma was confirmed, an uncommon presentation.

INTRODUCTION

Ewing's Sarcoma was first described by Sir James Ewing in 1921 as a series of bone tumours he called as diffuse endothelioma (2,3). It is a malignant primitive neuroectodermal tumour (PNET) that primarily presents during the first two decades of life (88%) (1). Approximately 85 % of these cases present primarily in the skeleton, less commonly in the muscle. With an annual incidence of 1-3 per million, it is generally a rare diagnosis (4). The spinal canal remains a rare site involvement for Ewing's Sarcoma; even more uncommon is the lumbar spine involvement which often mimics a benign condition before spreading extensively (5,6). Primary involvement of the non-sacral (lumbar) spine represents approximately 0.9% of all cases (6). We report a rare case of extradural lumbar spine Ewing's sarcoma that presented with a typical radicular pain ad proximal muscle weakness over a period of 8 months which improved with radical tumour decompression & histopathogical report confirmed its diagnosis. Ewing's sarcoma is one of the most aggressive bone tumours with high proliferative and invasive potential, presenting a confusing variety of imaging manifestations that can mimic other diseases, especially in adult patients who already suffer from other spine pathologies.

CASE REPORT

A 39 year old adult female came to the emergency department with complains of severe progressive, cramping, nocturnal pain in right thigh & leg over a period of 8 months duration which initially started from lower spine to right buttocks and extending to mid thigh and right leg calf muscles. The pain was initially pin pricking in nature and later aggravated during sleep, making her awake & restless whole night, giving rise to a classical "night cry" followed by periodic tingling & burning sensation of the same leg which gradually developed over last 2 months. She gave history of repeated falls following normal walking since 1 month which exacerbated her condition further by producing a mild limping gait more on left side. No abnormalities were found on neurological examination, beside a diffuse tenderness in the lower back and a restriction of the active flexion-extension motion and motor power of 3/5 of right lower limb with significant wasting of right thigh muscles. The passive straight leg-raising test was positive at more than 30°. Lhermitte's sign was positive indicating the progression of the condition. Clinically no lump was palpated in the lower spine. Her blood parameters were not that unusual apart from mild vitamin D deficiency (19.67ng/ml) and mild anemia (10.2gm/dl). Her preoperative cervical smear report (The Bethesda System) showed mild inflammatory changes without any dysplasia/malignancy. Lumbosacral MRI revealed a solid heterogenous patchy enchancing mass lesion of 48* 58*70 mm size in the right paraspinal region involving the psoas muscle extending from L2- L5 level with extension to the extradural space. The thecal sac was mildly compressed with minimal B/L foraminal encroachment along with spinal canal stenosis (fig1,2).

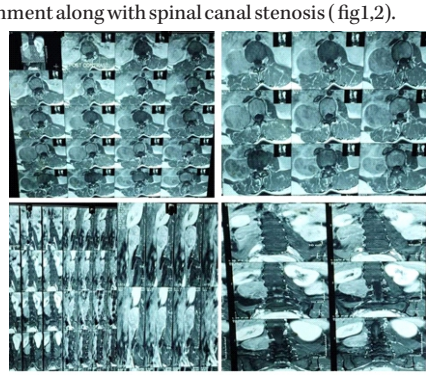

Figure 2: Post contrast T1 fat saturated images following intravenous administration of Gadolinium shows mild patchy post contrast enhancement of the mass with the STIR hyper intense areas showing non-enhancement.

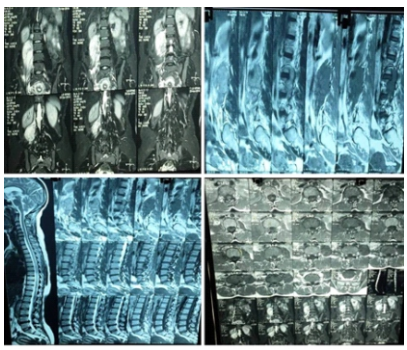

Figure 3: MRI of the lumbosacral spine shows a well defined right paraspinal soft tissue mass lesion L2 – L4 vertebrae bodies. The mass

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lesion B hypointense on T1W Sequence, hyperintense on T2W sequence and brightly hyperintense on STIR sequences. On STIR images there B variegated internal architecture with few small cystic area within the mass lesion. There appear to be an incomplete T2 hyper intense thin capsule surrounding the lesion.

However, the ultrasonography picture of the whole abdomen was more in favour of benign condition such as Neurofibroma/ Schwannoma, with the right paraspinal large fusiform lesion of size 14* 7.3cm infiltrating the psoas muscle. The patient made an uneventful postoperative recovery and was referred for radiation for further management.

OPERATIVE FINDINGS

Following laminectomy from L2-L5 the tumour was found to be extradural to the L2 vertebra. L2 nerve root was comparatively thickened. The lesion was connected to the large extra vertebral mass, lying anterolateral to the vertebral body. It was densely adherent to the psoas muscle and mostly encapsulated. Intra operatively, the lesion was moderately vascular, brownish, friable, intracapsular and with a greyish to white friable material inside it. The lesion was removed into, except adherent capsule. Resection was up to apparent normal bone, well beyond the limits of the involved vertebral body as disclosed by MRI. The resected fragment was sent for histopathological studies.

POSTOPERATIVE FINDINGS

Postoperative period remained uneventful where the patient improved gradually and significant improvement seen in the reduction of radicular pain, but the limping gait still remained predominant. Stitches were opened on the 8th POD with no signs of infections or inflammation.

HISTOPATHOLOGICAL REPORT

Postoperatively, Squash cytosmears revealed small round cell tumour more in favour of non Hodgkin's lymphoma/ PNET. Received sample of given bits of tissue together measuring 3*2.5*1 cm. The histopathological study revealed multiple nerve bundles, fibrocollagenous tissue, blood vessels, skeletal muscle bundles intervened by tumour tissue comprised of round cells having high nuclear- cytoplasmic ratio, hyperchromatic nuclei, and inconspicuous nucleoli and scant to moderate clear cytoplasm(5). The cells were arranged diffusely as well as in rosettoid pattern. It was immunonegative for leucocyte common antigen (LCA negative), but diffusely and strongly immunopositive for CD99 (7). Thus the diagnosis of PNET/ extraskeletal Ewing's sarcoma was given (fig3).

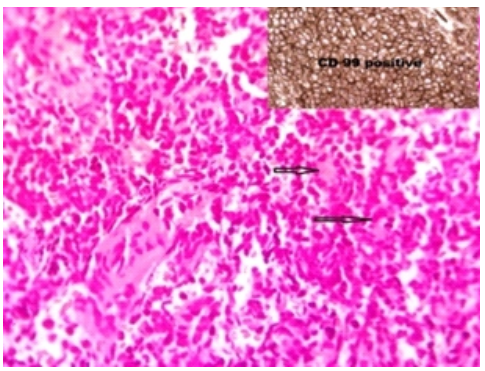

Figure 3: Showing microsection shows a tumour comprised of small round cells having hypochromatic nuclei with indentation in few, inconspicuous nucleolus, and scant to moderate clear cytoplasm. The cells are arranged diffusely as well as in rossettoid pattern (H &E * 400). The inset shows CD 99 immunopositivity.

DISCUSSION

Ewing's Sarcoma or Blue cell tumour (7,2) is a type of round cell tumour which remains for several aspects an anomalous tumour with bizarre behaviour. Hense et al described an unexplained, intrinsic major aggressive behaviour of axial tumour (4). Ewing's Sarcoma/ PNET family of tumours are more devastating malignancies that have slight male preponderance (M/F: 2:1), occurring more often in children & young adults (peak age group between 15-20 years). In certain cases, prognosis of Ewing's Sarcoma turns worse with advancing age, which is otherwise more difficult in the overall survival rate. The most common areas of involvement are pelvis (25%) (8,2), ribs, femurs, head and neck, buttocks, lower extremities, chest wall (Askin Tumour)(2,4) and retroperitoneal space. The spinal canal remains a rare site for Ewing's Sarcoma to arise, more involvement of sacrum (50%) (4), rarer being the lumbar spine involvement. In our report, an adult female with 8 month h/o of right sided radicular pain and proximal muscle weakness underwent disc laminectomy with tumour decompression. The tumour was spreading along the nerve roots encroaching the psoas muscle. Post operative the patient had rapid relief of symptoms including relief of monoparesis (right leg) and radicular pain. There was no sign of post laminectomy kyphosis which is common complication (40%) post resection of tumour. The patient was discharged a week following with oral corticosteroids & healthy wound site scar. She was referred for radiotherapy at a local tertiary centre. After 6 months follow up, the patient still remained asymptomatic.

The rarity of the case depended mostly on the extradural approach of the tumour, uncommonly found in an adult female. Optimal treatment of Ewing's sarcoma after diagnosis is provided by a combination of surgical excision along with radiotherapy with or without chemotherapy. Local radiation therapy to the spine is usually given at a dose of 50-55 Gy (mentioned full form) with inclusion of an adequate margin as deemed appropriate by the CT or MRI (1). Higher doses of radiation can lead to postradiation myelopathy.

In a recent review of the data collected in the cooperative Ewing's Sarcoma Study and the European Intergroup Cooperative Ewing's Sarcoma Study (IESS), local recurrence occurs in 22% of patients receiving radiation alone and 18.7% of those receiving surgery & radiation(1).

The ideal treatment for Ewing's sarcoma is often institution specific, and driven by trial based protocols both for paediatric & adult age groups, which still remains undefined (4). Surgical decompression of tumour gives immediate relief from the acute symptoms however radiation is needed to prevent recurrence and overall survival of the patient.

The accepted chemotherapy protocol developed by the "IESS" group consists of cyclophosphamide, vincristine, dactinomycin and doxorubicin when administered with radiotherapy has proven superior to a three drug regimen with local irradiation or to four drug regimen with bilateral pulmonary radiation therapy (1).

CONCLUSION

To date, the number of reported cases of EES with primary localization on mobile spine in adult patients remains small and they are all sporadic reports. This is the reason why there are still several concerns about the optimal treatment, especially in adults (2). The optimal treatment however remains as surgical resection followed by neoadjuvant chemotherapy combined with radiotherapy, to improve overall outcome of the patient.

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