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RECURRENT EPITHELIOID SARCOMA OF THE LUMBAR SPINE: A RARE CASE REPORT

Neurosurgery

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ABSTRACT

Epithelioid sarcoma is a rare[1,2] soft tissue sarcoma, uncommon in females[3,4] with less chances of occurrence in spine, lumbar region being the rarest[13] location. Initial diagnosis is often misleading[6,13]. Only few cases involving spine[5] are reported in literature, most of which have distant metastasis at presentation. This is a case of a 34 year old lady with abdominal lump, low backache and weakness of both lower limbs since last 2 years. Neurological examination showed decreased power in bilateral lower limbs. MRI of spine revealed a soft tissue tumor arising from lumbar spine causing extradural compression and having a large retroperitoneal component. Initially, resection of the tumor was done followed by histopathology which suggested a benign tumor. Soon, the growth reappeared alongwith neurological deficit. Then, immunohistochemistry review of the previous slides revealed epithelioid sarcoma of the spine. Whole body ^{18}F-FDG PET/MRI revealed a bulky metabolically active recurrent mass in the previously operated bed with compression of nerve roots and no distant metastasis. The authors have thus reported a rare case of recurrent epithelioid sarcoma of lumbar spine and its clinical course along with immunohistochemical and imaging findings.

KEYWORDS

Epithelioid sarcoma . Recurrent . Lumbar spine .

INTRODUCTION

Epithelioid sarcoma is a rare type of soft tissue sarcoma. The subtypes are categorized as rare proximal type and common distal type with the overall incidence of this tumor being less than 1%[1,2].

The distal type[16] usually presents with multiple nodules in the dermis of upper extremities, especially the hand and the wrist. On the other hand, the proximal or the axial type is more aggressive[5,14] and occurs more commonly in pelvis, perineum, and proximal extremities.

Initial diagnosis is often not correctly ascertained[6,13] due to histopathologically benign features such as epithelioid or necrotic granuloma-like appearance. It is more common in adolescent males than females[3,4]. Not much is known about the etiopathogenesis and geographical distribution of this tumor.

Accurate diagnosis is provided mainly by immunohistochemistry due to limitations of gross microscopic features in predicting the nature and subtype of the tumor. These tumors have a tendency for local recurrence[6,7,8] after initial surgical resection and metastasis[9,10,11,12] to regional lymph nodes, lung, bone, brain, and other locations, including the scalp.

According to the available literature, only few cases of epithelioid sarcoma have been reported in spine involving cervical[5,13] and thoracic[13] vertebrae with only two case reports showing involvement of the lumbar spine[17,18].

CASE REPORT

A 34 year old female presented with complaints of gradually progressive abdominal lump with low back ache and progressive weakness of both lower limbs since two years. MRI spine revealed soft tissue mass at L4-L5 level extending to involve psoas muscle. Image-guided FNAB was done which was suggestive of Malignant peripheral nerve sheath tumor.

Further management was proposed in the form of staged surgical procedure. The first stage being posterior instrumentation, fusion, laminectomy, decompression and partial tumor removal. Second stage involving anterior retroperitoneal approach to remove the tumor mass and corpectomy with reconstruction of L5 body. The final stage being complete tumor resection depending upon clear margin.

Repeat MRI spine revealed large soft tissue mass in the lumbar region of low intensity in T1w and high intensity in T2w images (Fig.1 and 2). L5 body was compressed with partial involvement of L4. CECT abdomen revealed heterogenously enhancing soft tissue retroperitoneal mass involving right para- and peri-vertebral regions with destruction of L4 and collapse of L5 body with medial extension causing obliteration of thecal sac and posteriorly extending to involve the erector spinae. Initially, she underwent right L5 hemilaminectomy with L2,L3 and S1 posterior inter fusion alongwith biopsy of the tumor. Histopathological examination revealed schwannoma and no evidence of malignancy. Repeat CECT abdomen done post-operatively revealed unchanged huge lobulated retroperitoneal mass lying between the right common iliac artery and vein. The mass invaded the spinal canal at L4 and L5 at right paravertebral level with no evidence of metastasis. Shortly, patient underwent next stage surgery with post-operative course being uneventful.

However, rapid recurrence was noted after four months which lead to follow up on histopathology. The final diagnosis was a soft tissue sarcoma(epithelioid type) of the spine of distal variety(grade II).Patient followed up with us for further management. There were scars (of previous surgery) in both abdominal and lumbar regions. There was visible right paraspinal(lumbar) fullness extending towards right flank and abdominal lumbar region, crossing the midline to extend beyond the umbilicus towards left side. Neurological examination showed marked motor weakness of both lower limbs (3/5 MRC grade) affecting the gait with sensations intact and SLR in both lower limbs of around 10 degrees. Rest of the clinical examination was unremarkable. PET/MRI was planned to evaluate the extent of disease of spine, cord, root or nerve involvement and distant metastasis. It revealed a large metabolically active recurrent tumor with extension to retroperitoneal and pelvic regions with no evidence of distant metastasis.

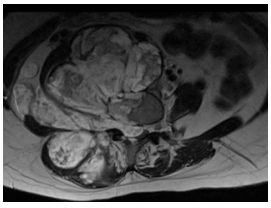

Figure 1. T2w image showing large lobulated soft tissue paravertebral mass in the L5 lumbar region involving the vertebral body with extension into the spinal canal.

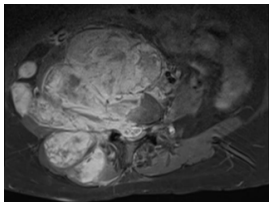

Figure 2. Post-contrast image showing heterogenous enhancement of mass

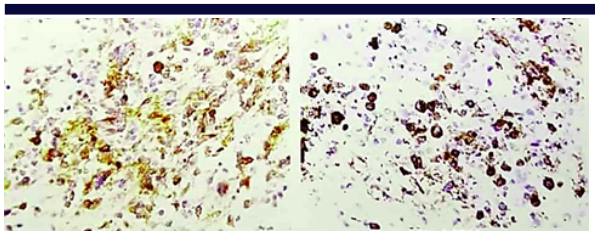

Figure3. Immunohistochemical reaction showing positivity to epithelial membrane antigen(EMA) (left side) and to CD34 (right side).

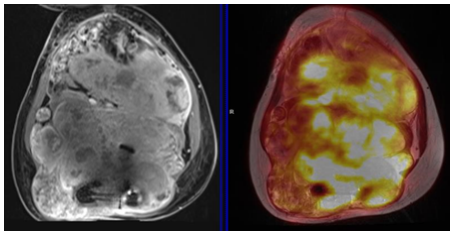

Figure 4. Fused PET/MRI image at L4-L5 level showing huge heterogenous enhancing mass (left side) filling abdominal cavity and invading the paraspinal region involving vertebrae,muscles and extending into the spinal canal. Image on the right side shows mass at the same level as metabolically active with increased ^{18}F-FDG uptake.

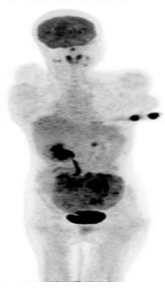

Figure 5. (right) MIP(maximum intensity projection) ^{18}F-FDG PET image showing large metabolically active mass in abdomino-pelvic region with no evidence of metastasis. ^{18}F-FDG avid foci seen in the left hand region indicates injection site.

DISCUSSION

Epithelioid sarcoma is a rare type of soft tissue sarcoma with overall incidence being <1%[1,2]. It is often misdiagnosed as benign and later on suspected to be malignant owing to its aggressive growth in a short span. It was first described by Lakowski in 1961 as "sarcoma aponeurotica" and the term "epithelioid sarcoma" was first coined by Enzinger in 1970[15]. It has two known pathological variants[5] namely proximal and distal. Proximal variant (more aggressive)[14] is more common in axial parts of body and distal variant (granuloma like) is more common in extremities[16].

The most common reason why the diagnosis is often late and misleading[13] initially is because these cases often present with rapid growth with histopathological features similar to those of inflammatory processes and benign tumours. Tumour itself has tendency to metastasize[9,10,11,12], so delayed diagnosis might affect the chance of curability. In this case, the initial histological diagnosis was a benign tumour probably schwannoma. This is the reason that no chemotherapy or radiotherapy was instituted and the tumor was debulked in the second stage of surgery. However the tumour recurred quickly in a course of 4 months that prompted re-evaluation by histopathology and radiology.

As evident in literature[5], immunohistochemical co-expression of cytokeratin and/or epithelial membrane antigen (EMA), vimentin, reactivity for CD34 are seen in around half of the cases, and an occasional reactivity to smooth muscle actin (SMA), desmin, neuron specific enolase and S100 protein. In our case, the final diagnosis was made on the basis of typical immunopositivity to EMA and CD34 (Fig.3). Spinal canal and nerve involvement is common, hence MRI was prefered over CT scan. It was combined with ^{18}F-FDG-PET to assess the avidity as well as distant metastasis. The tumor was found to be metabolically active (Fig.4) and no other active lesion was found elsewhere in the remaining whole body (Fig.5) to suggest metastasis.

Wide local excision is the standard treatment but often not possible[6] due to involvement of nerve sheath and the spinal canal. The present case is a recurrent one having huge bulk with local invasion of nerve roots with unclear tissue planes due to adhesions, hence surgery was not contemplated. Cases in literature have shown initial radical resection[19] followed by chemoradiotherapy[13] in the management, however lacks clear guidelines owing to extreme rarity of this disease. In conclusion, we report this case of epithelioid sacrcoma of the lumbar spine due to its rare location, the clinical course and presentation.

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