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Case Report

Intraosseous schwannoma

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ABSTRACT

Intraosseous schwannomas are rare and benign neoplasms originating from the nerve sheath cells. We present two such cases, one located in the ninth thoracic vertebrae and the other involving the sacrum. Clinicoradiological diagnosis in both the cases was a primary bone tumor. Histopathological examination revealed the characteristic morphology of schwannoma, which was confirmed by immunohistochemistry. These two cases highlight the importance of clinical, radiological and histopathological correlation in the diagnosis of these uncommon bone tumors.

KEY WORDS: Intraosseous, sacral, schwannoma, spinal

INTRODUCTION

Schwannomas are benign soft tissue neoplasms originating from nerve sheath cells. These characteristically present along the roots of sensory nerves and are frequently seen as soft tissue tumors of the extremities or thorax [1]. Intraosseous location is rare and account for <0.2% of primary bone tumors; mandible and sacrum being the most common sites affected [1,2]. Intraosseous schwannoma, clinically and radiologically mimics a primary bone tumor [3]. We present two such cases, one involving the thoracic vertebra and other one in the sacrum.

CASE REPORTS

Case 1

A 33-year-old male presented with back pain and left radiculopathy since 2 years. No genital or sphincter dysfunction was noted. He gave a history of similar episodes of pain during childhood. On local examination, a small tender swelling over the thoracic spine was seen at T8-L2 level. No neurological deficit was seen. Computed tomography (CT) scan of dorsal spine showed a well-defined mildly expansile lytic lesion involving left posterior body, pedicle and transverse process of T12 vertebra, with mildly sclerotic margins and soft tissue lesion filling the defect. Laterally the cortical bony margin was extremely thinned out. The neural foramen was normal [Figure 1]. Clinical differential diagnoses were giant cell tumor and osteoblastoma. T9 pedicle and transverse process biopsy was done. Histopathological examination revealed a cellular tumor composed of predominantly Antoni A areas with interlacing fascicles of spindle cells with buckled nuclei and numerous Verocay bodies along with areas of hyalinized blood vessels. The spindle cells showed immunoreactivity to S-100 [Figure 2]. Final diagnosis of schwannoma was made. The patient was further managed conservatively.

Case 2

A 49-year-old lady presented with low backache and right radiculopathy. Bowel and bladder continence was maintained. Local examination of the spine showed mild paresthesia and

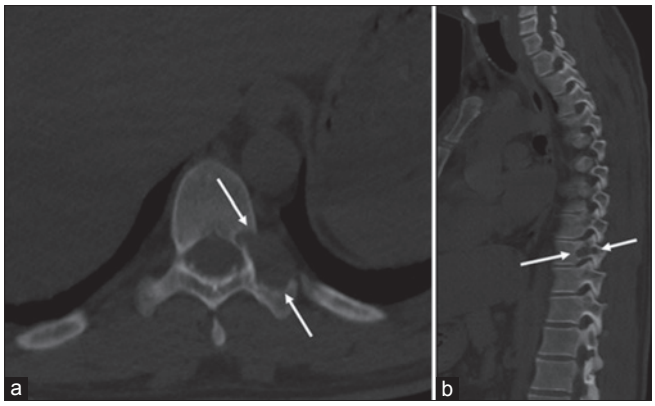

Figure 1: Computed tomography scan of dorsal spine, (a) axial and (b) sagittal views reveal an expansile lytic lesion showing thin sclerotic rim, involving the left posterior body, pedicle and transverse process of T12 vertebra with associated soft tissue lesion within (arrow). There was no evidence of any neural foraminal widening seen.

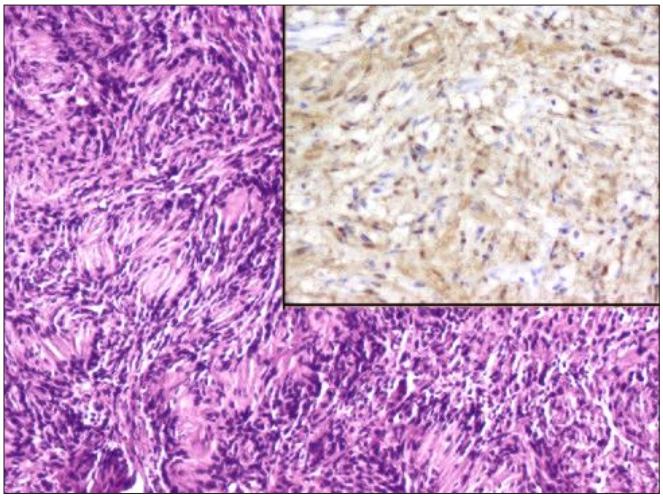

Figure 2: Photomicrograph showing schwannoma composed of fascicles of spindle cells with Verocay bodies (H&E, ×100). Inset: Tumor cells showing S100 protein immunoreactivity (Immunoperoxidase, ×200).

tenderness over the lower part. There were no symptoms of neurological deficit. Plain radiograph of the pelvis revealed a large relatively well-defined lytic lesion involving right sacral ala and the differentials put forth were chordoma and metastasis [Figure 3]. Subsequently, CT scan was done which revealed expansile lytic lesion involving the right neural foramina of S1 vertebra with an intermediate density soft tissue lesion within, extending into the pelvis from a defect in postero-inferior margin of sacral ala [Figure 4]. Diagnosis of nerve sheath tumor was proposed. A lesional biopsy was done. The biopsy showed a tumor composed of cellular Antoni A and hypocellular Antoni B areas. The former showed nests, whorls and fascicles of spindle cells, few showing nuclear pseudoinclusion along with Verocay bodies. The hypocellular areas showed loosely placed spindle cells with areas of hemorrhage and hyalinized vessels [Figure 5]. Differential diagnosis of schwannoma and meningioma was proffered. The latter was excluded by demonstrating S-100

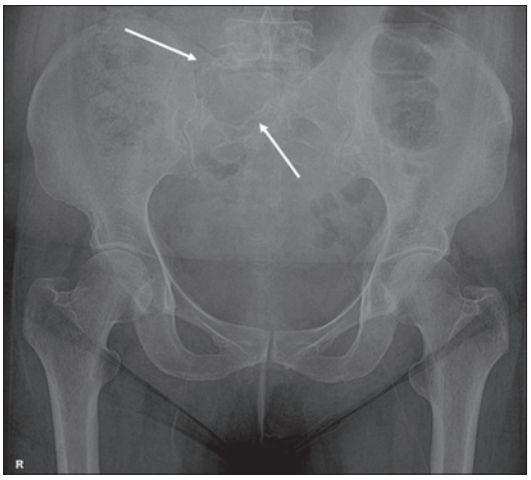

Figure 3: Frontal radiograph of pelvis reveals a large expansile lytic lesion involving the right sacral ala (arrows).

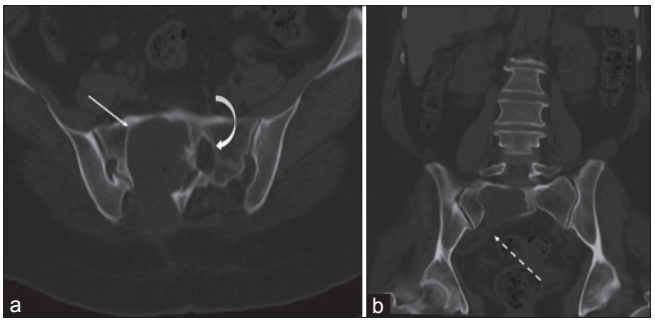

Figure 4: Computed tomography scan of pelvis, (a) axial and (b) coronal views reveal gross widening of right S1 neural foramina (arrow) showing thin sclerotic margins with an associated large soft tissue lesion completely filling it and extending into the pelvis along the dehiscent postero-inferior walls (dashed arrow). Note is made of the normal sized left S1 neural foramina (curved arrow).

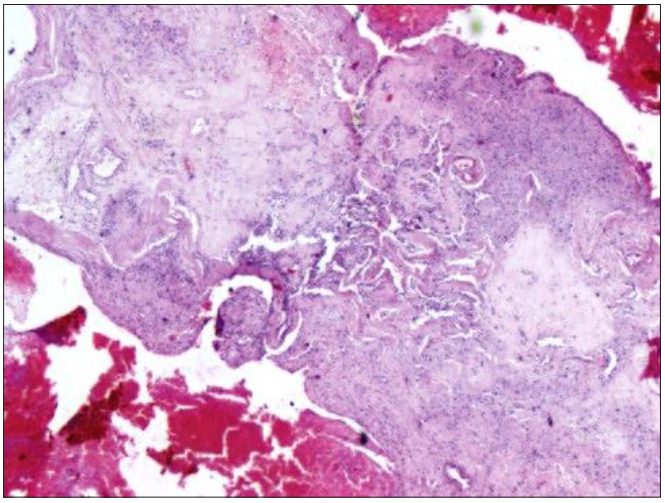

Figure 5: Photomicrograph showing hypocellular areas with hemorrhage and hyalinized blood vessels (H and E, ×40).

immunoreactivity in tumor cells which stained negative for epithelial membrane antigen. Patient was treated with local radiotherapy.

Discussion

Schwannomas are benign tumors derived from Schwann cells of the nerve sheath. Intraspinal schwannomas are usually found in the intradural extramedullary location, while their intraosseous localization is very rare. Most common sites for intraosseous schwannoma are mandible and sacrum with other rare documented sites being cervical, lumbar and dorsal spine [2-5]. The probable mechanisms postulated for intraosseous localization of nerve sheath tumors are: (1) an extraosseous tumor causing bony erosion with secondary invasion into the medullary canal, (2) tumor arising from the small vasomotor nerves along the nutrient vessels of the bone, (3) tumors arising from the central neural foramina and forming dumbbell configuration [3,5].

We present two intraosseous spinal schwannomas, one in the dorsal vertebra and the other in the sacral region. Dorsal vertebral intraosseous scahwannoma is only the second such case reported in literature, which was purely intraosseous, showing no extravertebral or spinal canal involvement, with no neural foraminal widening [5]. This morphology of the tumour favors the theory of its origin being from the vasomotor nerves supplying the nutrient vessels. Conversely the sacral schwannoma was seen as gross expansion of the neural foramina in sacral ala, with the extraosseous extension of the soft tissue into the pelvis through a defect in its cortex postero-inferiorly.

Preoperative diagnosis of intraosseous schwannoma has rarely ever been made, due to lack of awareness about the condition and primarily due to their non-specific imaging findings. Radiologic mimics include osteoblastoma, giant cell tumor, aneurysmal bone cyst and plasmacytoma [5]. Similar differentials were observed in our cases. On magnetic resonance imaging intraosseous schwannomas have been described to be hypointense on T1-weighted and hyperintense on T2-weighted images showing post contrast heterogeneous enhancement. On CT scan, they mimic any other benign primary bone tumor, presenting as variably expansile, lytic lesion with thin sclerotic margins, with occasional minimal periosteal reaction. Cortical destruction with extraosseous soft tissue extension has also been described uncommonly [3,5].

On histopathology, schwannomas are characterized by cellular Antoni A and hypocellular Antoni B areas composed of spindle shaped Schwann cells. The former shows bundle and fascicles of tumor cells with areas of peripheral palisading and central eosinophilic masses. Antoni B areas have loosely spaced, haphazardly arranged tumor cells. These are confirmed by immunohistochemical staining of tumor cells by S100 [6-8]. Schwannomas may undergo cystic change, hemorrhage and calcification. These are referred to as ancient schwannomas and can simulate a malignant process. However, the absence of atypical mitosis, increased mitotic activity and necrosis favors the benign nature of the disease [1].

Histopathological differentials of intraosseous schwannomas are discussed in Table 1 [9-12].

Table 1: Histopathological differentials of intraosseous schwannomas

Differential diagnosis	Differentiating features
Neurofibroma	Unencapsulated, absence of characteristic Antoni A areas and Antoni B areas
Malignant peripheral nerve sheath tumour	Presence of atypia, necrosis and atypical mitosis
Fibrous histiocytoma	Presence of multinucleated giant cells, foamy cells and chronic inflammation
Fibroblastic meningioma	Positive immunostaining for EMA and negative for S100
Fibrous component of fibrous dysplasia	Absence of schwannian (wavy) nature of spindle cells and negative for S100

In both our cases, the diagnosis was made based on histology and confirmed by S100 immunoreactivity.

The treatment of intraosseous tumor is surgical excision or curettage [5]. Recurrence is rare however late complications such as cystic myelopathy, spinal arachnoiditis, spinal deformity and pain have been reported [13]. The role of radiotherapy is unclear and proposed for cases where the tumor excision is limited by size, anatomic site or the risk of causing the functional defect [14]. Radiotherapy was offered in the second case presented due to the large size and site of the tumor.

CONCLUSION

Intraosseous schwannomas are rare nerve sheath tumors with an excellent prognosis. They are usually diagnosed on histopathological examination of lesions, excised or curetted with clinicoradiological suspicion of primary bone tumors. These cases emphasize the importance of a multidisciplinary approach utilizing clinical findings, radiology and pathology for accurate diagnosis of these rare bone tumors in order to provide timely and appropriate treatment.

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