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Case Report

Primary Neuroendocrine Tumor of the Lumbar Spine: Rare Tumor Mimicking Nerve Sheath Tumor of the Spine

Abstract

Neuroendocrine tumors (Carcinoid tumors) generally arise from enterochromaffin cells of gut and bronchi. Primary carcinoid tumors of spine are extremely rare and have been described in sacrum and coccyx. Primary carcinoid tumors involving the spinal cord are still rarer, and review of literature revealed only two cases reported. Our patient a 39-year-old male had diagnosed as a case of nerve sheath tumor (intradural extramedullary) at LV4 region on neuroimaging. However, postoperatively, the tumor turned out to be a rare primary spinal carcinoid tumor on histopathological examination and immunohistochemical staining. Work up to rule out any other site in the body was negative. To the best of our knowledge, this is the first case of primary carcinoid tumor of the lumbar spine.

Keywords: *Intradural extramedullary, lumbar, primary neuroendocrine tumor*

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Introduction

Neuroendocrine tumors (Carcinoid tumors [CTs]) generally arise from enterochromaffin cells of gut and bronchi.[1] The incidence of the disease has been reported to be 1 in 100,000.[2] Metastasis to central nervous system involving the brain[3] and spinal column are infrequent.[4,5] Primary CTs of spine are extremely rare and have been described in sacrum[6] and coccyx.[7] Primary CTs involving the spinal cord are still rarer, and review of literature revealed only two cases reported.[8,9] To the best of our knowledge, this is the first case of CTs of the lumbar spine which was in intradural extramedullary (IDEM) mimicking a nerve sheath tumor in the preoperative contrast-enhanced magnetic resonance imaging (MRI).

Case Report

A 39-year-old male presented to our outpatient department with complaints of low backache of 6 months' duration associated with radiculopathy and paresthesia of the right lower limb of 4 months' duration. The patient did not give any history of trauma, weakness of lower limbs, or any sphincter disturbances. Neurological examination did not reveal any focal neurological deficits except for sluggish deep tendon jerk of the left ankle. Contrast-enhanced MRI of the lumbosacral spine with a screening of the whole spine revealed well-defined 10 mm × 12 mm × 15 mm IDEM lesion at the level of LV4 level which was mildly hyperintense on T1W and showed uniform postcontrast enhancement [Figure 1a and b]. The lesion was displacing the cauda equina circumferentially and had no foraminal extension. Based on the patient's clinical history, examination findings, and the character of the lesion on contrast-enhanced (CE) MRI, a provisional diagnosis of nerve sheath tumor arising from the left L4 nerve root was entertained.

The patient underwent laminectomy of LV4–LV5, durotomy, and total excision of the lesion. Post operatively, the patient recovered well without and fresh neurological deficits. Postoperative CE MRI revealed complete excision of the lesion [Figure 2a and b]. Cells positive for chromogranin neuroendocrine marker [Figure 3a ×400] and cells positive for CD56 neuroendocrine marker [Figure 3b ×400]. HPE was consistent with low-grade neuroendocrine tumor. In view of this rare diagnosis on HPE, the patient underwent further evaluation with CE computed tomography (CECT) chest and abdomen,

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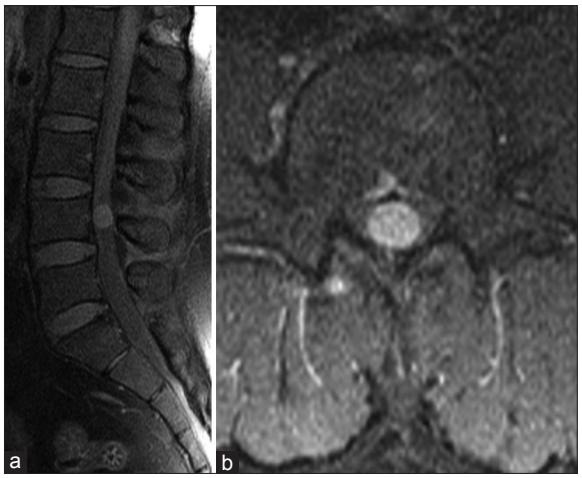

Figure 1: (a) Preoperative postcontrast sagittal image showing homogeneous enhancement of the lesion, (b) preoperative postcontrast axial image showing homogeneous enhancement of the lesion

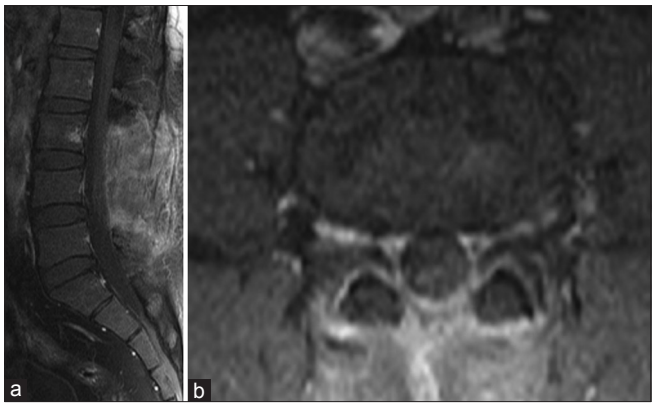

Figure 2: (a) Postoperative postcontrast sagittal image showing complete excision of the lesion, (b) postoperative postcontrast axial image showing complete excision of the lesion

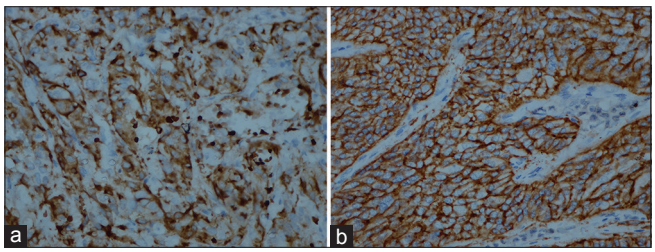

Figure 3: (a) Cells positive for chromogranin neuroendocrine marker (×400), (b) cells positive for CD56 neuroendocrine marker (×400)

positron emission tomography (PET) scan to rule out any other lesion in the body. No other lesions were seen on any further evaluation. A diagnosis of primary neuroendocrine tumor of the spine was made, and the patient has remained on regular follow-up without any fresh complaints of neurological deficits on examination.

Discussion

IDEM of the spinal cord is the most common lesion of which nerve sheath tumors and meningioma being the most common pathologies. CTs presenting as metastasis to the spine have been reported in literature.[4,5] However, primary CTs of the spine is a very rare entity, and review of literature revealed only two cases, Narayan *et al.* reported a case of primary cervical spine CT in a 50-year-old African–American woman[8] and Zhang *et al.* reported a case of primary CT of medulla spinalis in a 33-year-old male.[9] There are no reported cases of primary CT of spine of lumbar spine mimicking an IDEM as was the case in our patient.

CTs are rare entities arising from enterochromaffin cells of gut and bronchi with an incidence of 0.28 to 0.8 cases per 100,000 per year.[10] Hormonally, active tumors release serotonin which the cause of carcinoid syndrome in these patients characterized by vasodilatation, flushing, diarrhea, and bronchoconstriction. Most commonly CTs present as metastasis to vertebral column which itself is rare and seen in fewer than 2% of cases.[5,11] The differential diagnosis of IDEM of spine includes nerve sheath tumors (30%), meningioma, ganglioneuromas, and ganglioneuromas. Typical morphology of Schwannoma is a well-circumscribed round or lobulated lesion with homogeneous enhancement on contrast MRI as was the case in our patient. Radiologically, it is difficult to differentiate nerve sheath tumors from primary carcinoid tumors of the spine if the lesion if well circumscribed and lobulated presenting as an IDEM. Since primary CTs are rare tumors, most commonly they are diagnosed on histopathological examination which reveals cells arranged in ribbons and festoons separated by thin fibrovascular septae with abundant dense pink cytoplasm with nuclei vesicular to having salt and pepper chromatin. Tumor cells are positive for Chromogranin neuroendocrine marker and cells positive for CD56 neuroendocrine marker.

The workup to rule out metastatic disease of the spine includes CECT of chest and abdomen, PET scan and octreotide scintigraphy which was performed in our patient and did not reveal and other lesions. The contrast-enhanced MRI of the lesion in our case which was hypointense on T1W, hyperintense on T2W, and showed homogeneous contrast enhancement suggestive of IDEM and consistent with nerve sheath tumor. Hence, it is difficult to diagnose primary CTs of the spine preoperatively just on imaging features, and definitive diagnosis is on HPE and positive immunohistochemical (IHC) staining for chromogranin A as was demonstrated in our case.[12]

The treatment of choice remains complete excision of tumor which will provide definitive cure in primary CT of spine. Radiotherapy is recommended for patients having metastatic CTs of the spine; however radiotherapy and chemotherapy have limited evidence in the management of primary spinal CTs.[9]

Conclusions

Primary spinal CT is rare tumors and is difficult to diagnose preoperatively just on imaging characteristics. Definitive diagnosis is only after HPE and IHC. However, once diagnosed on HPE, patients need to be investigated with CECT abdomen and chest, PET scan and octreotide scanning to rule out metastatic CT of spine. The total excision of the lesion is curative in primary spinal CTs with no significant role of adjuvant radiotherapy and chemotherapy in these patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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