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An intradural extramedullary spinal paraganglioma: Case report

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Abstract

Background: Paragangliomas originate from peculiar type of cells derived from neural crest origin, and are relatively sparse in presence. Most often these are originated with in the adrenal glands. Origin with in the spinal canal is rare, however when present, they are found in the intradural extramedullary compartment in lumbosacral region. They are usually indolent and considered WHO grade I lesions. Patient presents with symptoms of lower back pain which radiates into the lower extremities. Patients may also present with sensory or motor neural impairment.

Case Report: We present a case of a 33-year-old male initially presented with bilateral sciatica and recumbent back pain. Mild urinary urgency for over last one month was also noted. MRI scan performed which demonstrated a homogenously enhancing intra dural, extra medullary nodule at the level L2 vertebral body. Radiologically this was interpreted as ependymoma versus meningioma. Neurosurgical consult recommended due to mass effect on the adjacent cauda equina nerve roots. Laminectomy was performed and excision was done of this space occupying lesion. The histopathology turned out to be paraganglioma versus neuroendocrine tumor.

Practical Implications: Paragangliomas and neuroendocrine tumors of spine are a rare entity. They can be misdiagnosed on imaging as they have similar or overlapping features of that of the meningioma or ependymoma. The purpose of this case report is to make radiologists aware regarding the rare entities come under differential diagnosis of intradural extra-medullary tumors. To a greater extent a combination of clinical, pathological and imaging correlation should be done for better diagnosis.

Keywords: Paraganglioma, spinal neoplasms, intradural, extramedullary

Introduction

Intra-dural extra-medullary tumors comprises up to 70 to 80% of the spinal canal tumors [1]. Most commonly the intra-dural extra-medullary neoplasms comprises of meningiomas, schwannomas and neurofibromas with relatively less common entities of hemangiopericytoma or fibrous tumors [1]. Paragangliomas are neuroendocrine tumors which are usually low grade and are considered as WHO grade I tumors. These are most commonly arise from the adrenal glands; however extra adrenal locations though rare are still appreciable in clinical practice [2]. Glomus tumors (type of paragangliomas) are reported to be up to 90% of all the extra adrenals paraganglioma sites [3]. In particular to the neural axis most of the paragangliomas are associated within the spinal canal. The spinal paragangliomas mostly present with age of 40-60 years and are slightly more common in the male gender [4]. Relatively more common lesions in these location like ependymomas or meningiomas, the spinal paragangliomas are often misdiagnosed. Clinically they present with back pain or radiculopathy [5]. Here we represent a case of a 33 years old male initially presented with bilateral sciatica and recumbent back pain: the targeted magnetic resonance imaging (MRI scan) showed a well-defined intra-dural extra-medullary tumor in the upper lumbar spine which turned out to be a paraganglioma on pathological evaluation.

Presentation of case

A 35 years old male with no comorbids presented with recumbent lower back pain with associated bilateral sciatica for two months. Symptoms of mild urinary urgency along with gradual loss of urinary control was also noted. Bilateral lower extremity numbness making it difficult to walk. No motor deficit was noted.

He was not associated with any history of smoking or drug abuse. On magnetic resonance imaging (MRI) through the spine there was noted an intra-dural extra-medullary nodular lesion measuring 1.8 x 1.2 cm in craniocaudal and transverse dimensions located at the level of L2 vertebral body. On post contrast scan, the lesion shows homogeneous enhancement. There was noted displacement of adjacent cauda equina nerve roots due to mass effect; however no significant expansion of the thecal sac seen. The lesion was closely applied to the filum terminale without any associated bony remodeling. The epidural fat recesses were also preserved. No exit neural foramen and compression noted. Initially, the finding was interpreted as meningioma versus ependymoma of the spine. Neurosurgical consultation was

suggested as there was mass effect on the cauda equina nerve roots. While the brain MRI scan was unremarkable. The patient underwent laminectomy and excision of the intra-dural extra-medullary space occupying lesion at the level of L1-L2. Complete excision was achieved which turned out to be paraganglioma versus neuroendocrine tumor favoring the former. However, owing to its strong CK expression. The differential of neuroendocrine tumor remained. After the surgery the patient was hemodynamically stable without any neurological deficit. CT scan through the neck, chest, abdomen and pelvis to look for any other primary or secondary focus was also performed which was negative.

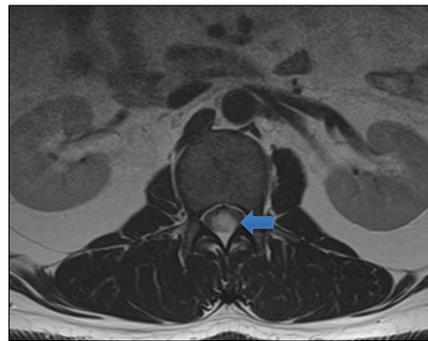


Fig 1A.



Fig 1B.

Figure 1A (Axial slice T2 sequence) and Figure 1B (Sagittal slice T2 sequence) showing a hyper-intense intra dural extra medullary lesion (arrows) at the level of L2 vertebral body having a mass effect on the adjacent cauda equina nerve

roots. The spinal cord tapers to form conus medullaris at L1 vertebral body level. Note that there is no significant expansion of the thecal sac. Background degenerative disc changes also noted.

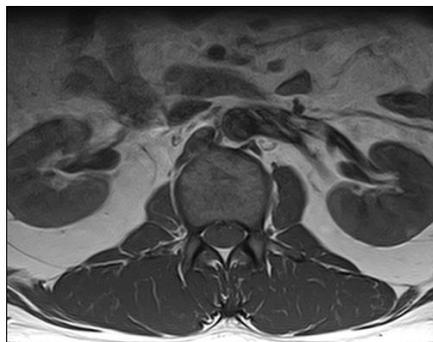


Fig 2A.

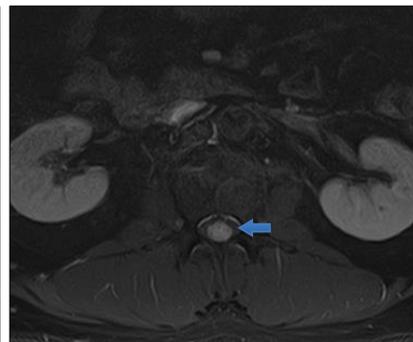


Fig 2B.



Fig 2C.

Figure 2A: Axial slice pre-contrast T1 sequence shows a hypointense signal intensity of the lesion.
Figure 2B: Axial slice post contrast from same level and

Figure 2C Sagittal slice T1 post contrast sequence showing homogenous contrast uptake (Arrows) can easily be mistaken with ependymoma or meningioma.



Fig 3A.



Fig 3B.

Figure 3A: Axial slice contrast enhanced CT scan (CECT) acquired after surgical removal of the lesion and Figure 3B: Sagittal slice contrast enhanced CT scan show post operative changes (Arrows). There is no residual disease seen on CT scan. Rest of the covered sections were negative for any possible primary or metastatic disease.

Discussion

Paraganglioma or the neoplasm having neuroendocrine origin which are mostly well encapsulated and highly vascular in morphology [6]. These can be further grouped into para-sympathetic and sympathetic types [7]. The sympathetic paragangliomas secrete catecholamines into the blood stream, while non-secretory trend have been seen associated with para-sympathetic paragangliomas [8]. The spinal para-gangliomas are typically associated with lower back pain and radiculopathies owing to its strong affiliation with the cauda equina and filum terminale [9]. As per literature there are only 2.5 to 3.8% cases reported for cauda equina or filum terminale para-gangliomas [10]. The tumor size ranges from 1.5 to 10 cm; however, these are usually not associated with distant metastases. Men have slightly more predilection towards the disease with most of the cases are sporadic in origin [11]. MRI scan is a modality of choice for initial diagnosis as well as follow up of the spinal paragangliomas; however these tumors have no peculiar features and are thus frequently misinterpreted as ependymomas meningiomas and schwannomas. Spinal teratomas and hemangiomas could also come in the differentials of this [12]. Spinal paragangliomas can be associated with intra-tumoral chronic bleed or subarachnoid hemorrhage [13]. On MRI sequences, the lesion appears iso to hyper intense on T2 sequences, while show iso intense signals to the spinal cord on T1 sequence. Associated bony remodeling due to tumor expansion can be better visualized on CT scan. While the angiographic studies show early enhancement with persistent contrast uptake on the delayed phases [14]. There are scanty case reports on the imaging findings in particular to that of para-gangliomas in the literature; however some shows salt and pepper appearances as well as internal hemorrhage. Rare cases of syringohydromyelia was also seen associated with the paraganglioma are also reported [15]. In our case the lesion particularly appears as homogenous solid mass which was well encapsulated with smooth enhancement on post contrast sequences. There was not associated cystic lesion or internal hemorrhage. No metastatic lesion identified within the neural axis, while the screening CT scan acquired from the neck, chest, abdomen and pelvis was also clear of any suspicious neuroendocrine lesion/primary or metastatic deposit.

Conclusion

Due to its scarcity the spinal paragangliomas are considered down the line in the spinal imaging and are usually misdiagnosed to be intra-dural ependymoma or meningiomas. There is no pathognomonic radiological signs to govern its presence. It is important for the Radiologist to have a knowledge of various intra-dural extra-medullary lesions to have a better understanding; however, the histopathological diagnosis remains gold standard. Complete surgical resection is often curative.

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