Case of Conus Paraganglioma: Case Report with Review

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Abstract

Paragangliomas of spine are rare neuroendocrine tumors. They are World Health Organization grade 1 tumors with common location in cauda equina and filum terminale. We report a case of conus paraganglioma. A 57-year-old lady was admitted with low back pain with radiculopathy without bowel and bladder complaints. Magnetic resonance imaging (MRI) lumbosacral spine showed well-defined round-to-oval extramedullary lesion isointense on T1 and hyperintense on T2-weighted images at the level of L1 vertebra (suggestive of) neurogenic tumor. Intraoperatively reddish-brown capsulated tumor arising from conus with extension to filum and compressing the nerve roots was seen. Gross total excision of tumor was done. Microscopic examination showed well-encapsulated cellular tumor composed of sheets, anastomosing lobules, and nests of small, oval-to-round cells interspersed with blood vessels with mild nuclear atypia and occasional mitosis. Immunohistochemistry analysis was positive for synaptophysin and chromogranin suggestive of paraganglioma. Postoperative period was uneventful. MRI of dorso-lumbar spine showed no residual tumor. Patient was asymptomatic 5 months after surgery. The clinical, radiological, and pathological features of this rare tumor entity is presented.

Keywords
- paraganglioma
- spine

Introduction

Paragangliomas are neuroendocrine tumor. These tumors were first described by Miller and Torack in 1970.[1] Extra-adrenal paragangliomas mostly occur in carotid body and glomus jugulare. In spine, they are common in cauda equina and filum terminale and are nonfunctional. Clinical presentation is usually nonspecific. Conus is very rare site for these tumors. Contrast-enhanced magnetic resonance imaging (MRI) is the imaging of choice. We searched English literature and till now four cases of conus paraganglioma have been reported and we add one more to it. The details of the previous reported cases have been summarized in ►**Table 1**.

Case Report

A 57-year-old lady was admitted with complaints of low back pain with radiation to left lower limb for the last 3 months. No bowel and bladder complaints. On examination, patient was conscious oriented with power 5/5 in all four limbs with no motor and sensory deficit. Plantars were bilaterally flexor. MRI lumbosacral spine showed well-defined, round-to-oval extramedullary lesion measuring around $20 \times 15 \times 10$ mm isointense on T1 and hyperintense on T2-weighted images at the level of L1 vertebra with indentation on conus and cauda equina, features suggestive of neurogenic tumor (►**Figs. 1** and **2**). Patient was operated and D12 to L2 laminectomy was done.

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Table 1 Previous published cases

Sl. no.	Authors/Year	Age/sex	Location	Resection	Histopathology	Follow-up
1	Shuangshoti et al 1984[2]	24 y/female	Conus and cauda equina	Total	Paraganglioma and glioma	22 months
2	Singh et al 2005[3]	28 y/male	Conus	Total	Paraganglioma	5 year
3	Agrawal et al 2012[4]	50 y/male	Conus	Subtotal	Paraganglioma	Expired after surgery
4	Diyora et al 2015[5]	39 y/male	Conus	Total	Paraganglioma	2 year
5	Present study	57 y/female	Conus	Total	Paraganglioma	5 months

Intraoperatively on durotomy, reddish-brown capsulated tumor arising from conus with extension to filum and compressing the nerve roots was seen. Tumor was dissected from conus and gross total excision was done. Tumor tissue and filum was sent for Histopathological Examination (HPE). Lumbar pedicle screw fixation was done. Postoperative period was uneventful.

Histopathology—Microscopic examination (**A**) of tumor tissue showed well-encapsulated cellular tumor composed of sheets, anastomosing lobules, and nests of small, oval-to-round cells interspersed with blood vessels. Mild nuclear atypia and occasional mitosis were present. No necrosis or significant pleomorphism was seen. Immunohistochemistry analysis was positive for synaptophysin and chromogranin and was negative for Epithelial Membrane Antigen (EMA), Smooth Muscle Actin (SMA) and CD34. These features were suggestive of paraganglioma. (**B**) Filum showed no neoplasia or granuloma (►**Figs. 3** and **4**). MRI of dorso-lumbar spine was done that showed no residual tumor. Patient was asymptomatic 5 months after surgery (►**Fig. 5**).

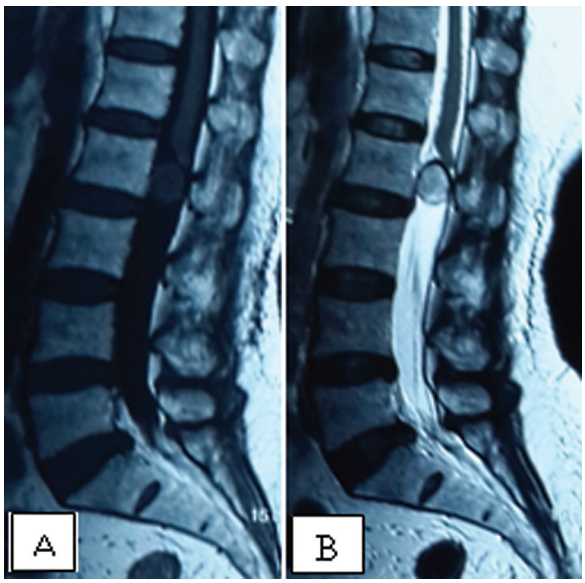

Fig. 1 (**A**) T1 and (**B**) T2 sagittal magnetic resonance imaging showing round-to-oval lesion isointense on T1 and hyperintense on T2 images.

Discussion

Paragangliomas are neuroendocrine tumors. These tumors have a neuroectodermal origin and are derived from the embryonic sympathetic and parasympathetic nervous system.[1,4] These tumors in spine were first described by Miller and Torack in 1970 and were termed as secretory ependymoma. Lerman was the first to coin the term paraganglioma of the cauda equina in 1972.[1,4] Paragangliomas are uncommon in cranium and are located in the petrous ridge, pineal gland, and sella turcica.[1,6] They comprise 3 to 4% of all spinal tumors and are mainly intradural, extramedullary in location. Most common site is cauda equina and the filum terminale, followed by thoracic and cervical regions.[8] Conus is a rare location for these tumors.[2–5] Previously cited reports show a male preponderance and occurance in third to fifth decade.[2–5] In conus, these tumors are generally nonfunctional. One case of functioning paraganglioma at conus has been reported.[4] Clinical presentation at conus may vary from low back pain with radiculopathy to motor and sensory deficits with bowel and bladder involvement.

MRI is the imaging study of choice for diagnosis. On MRI, paragangliomas appear as sharply circumscribed mass that is hypo- or isointense on T1-weighted images, hyperintense on T2-weighted images and show marked

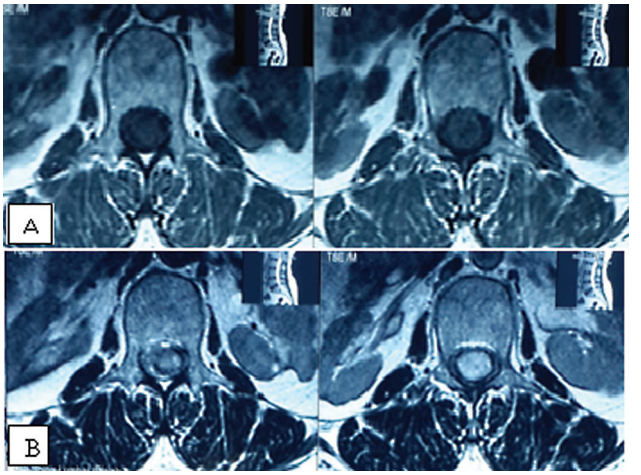

Fig. 2 (**A**) T1 axial and (**B**) T2 axial magnetic resonance imaging showing tumor displacing the cord.

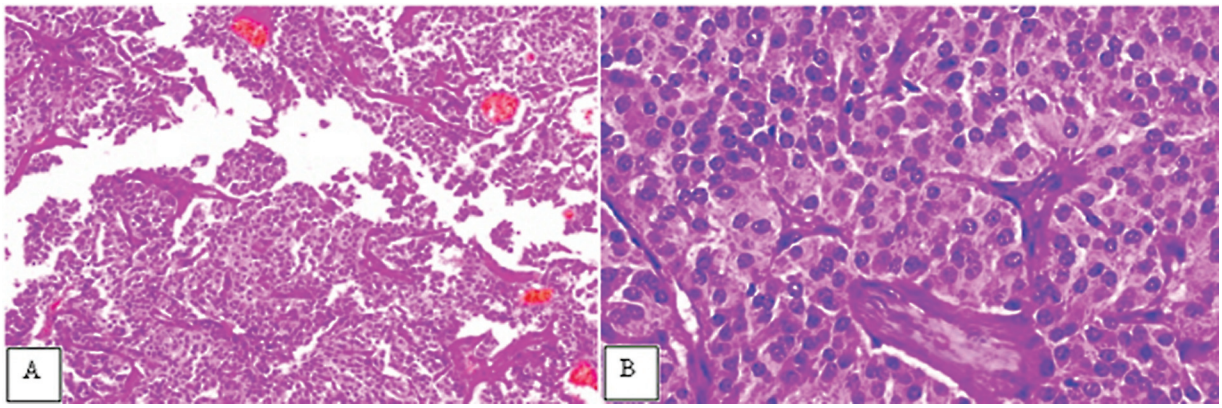

Fig. 3 Photomicrograph (**A**; hematoxylin and eosin [H&E] 10X) and (**B**; H &E 40X) showing well-encapsulated cellular tumor composed of sheets, anastomosing lobules, and nests of small, oval-to-round cells.

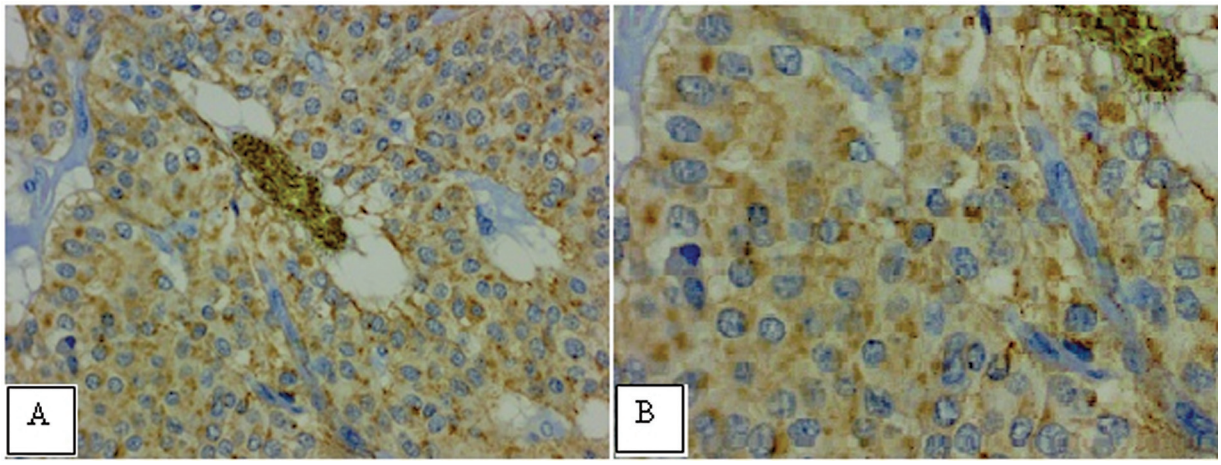

Fig. 4 Immunohistochemistry showing (**A**) positivity for chromogranin, 40X; (**B**) positivity for synaptophysin, 40X.

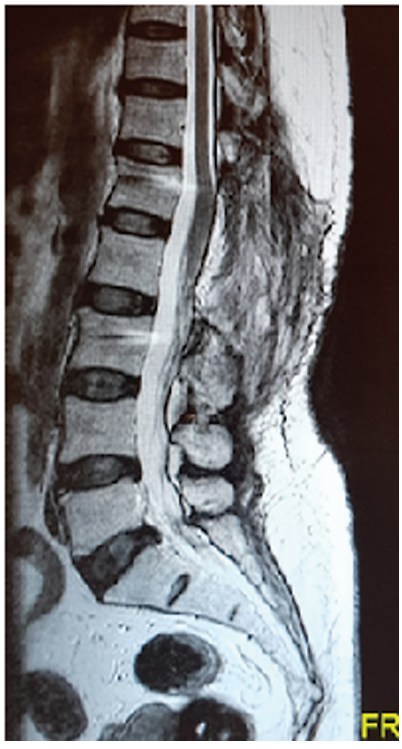

Fig. 5 T2 sagittal image showing no residual tumor.

contrast enhancement. The presence of serpentine, congested, ectatic vessels, and a low signal intensity rim ("cap sign") are considered diagnostically helpful clues.[5–7] Radiologically differential diagnosis includes neurinoma and ependymoma.[5–7]

Microscopic examination shows nests of round-to-polygonal tumor cells separated by fibrovascular septa, arranged in zellballen pattern.[3,8] Immunohistochemically paragangliomas are sensitive to neuron-specific enolase, but lack specificity, but synaptophysin and chromogranin are sensitive and reliable for chief cells.[3,9] Sustentacular cells show uniformly reactivity for S-100 protein. Differential diagnosis from ependymoma can be made by Glial Fibrillary acidic Protein (GFAP) positivity in ependymal tumors. Ependymoma may also show positivity for mucin and S100 protein in tumor cells rather than that in sustentacular cells of paraganglioma.[10,11] Paragangliomas are World Health Organization grade 1 tumors and prognosis is good if complete excision is done.[12] Recurrence after complete excision is rare.[3,5,13] Incomplete surgical resection tumors or locally invasive tumors are benefited with radiotherapy.[6,8]

Conflict of Interest
None declared.

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