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ABSTRACT
Paragangliomas of the spine are rare neuroendocrine tumors. We present a case of spinal paraganglioma at cauda equina region. Patient presented with low back pain with monoradiccular symptoms. Total excision of the intradural extramedullary lesion provided complete relief of symptoms postoperatively. Histopathological examination of the resected tumor mass, revealed features suggestive of paraganglioma. Although paragangliomas are rare in spinal region, they should be considered as a differential diagnosis in intradural extramedullary lesions. They are benign tumors and can be successfully treated by total resection.

Keywords: Spine, Paraganglioma, Cauda equina, Zellballen, Chromogranin, Neuroendocrine.

INTRODUCTION
Cauda equina paragangliomas are extremely rare tumors. The incidence of spinal paraganglioma is 0.07/100000. About 220 cases of paraganglioma of cauda equine have been reported in literature. Extra-adrenal paragangliomas are benign neuroendocrine tumors. Commonest extra-adrenal site is jugular glomus of carotid body (90%). Paragangliomas are mostly nonsecreting in nature except in a few cases where they present with endocrine and vascular symptoms due to catecholamine release. Contrast-enhanced magnetic resonance imaging (MRI) scan usually reveals a intradural extramedullary lesion. The preoperative diagnosis of paraganglioma is difficult to reach. These neoplasms have considerable similarity with ependymomas, and the diagnosis can be easily missed in absence of special techniques.

CASE REPORT
A 35-year-old male presented to the neurosurgery department with history of low back pain since last 1.5 months with radiation of pain to right lower limb. Pain was increased on sneezing and coughing. There was no associated bowel or bladder dysfunction, and the anal tone was normal. The general physical and systemic examinations were normal. The neurological examination was normal. Spine showed no deformity and the X-ray of the spine was unremarkable. Magnetic resonance imaging lumbosacral spine (Figs 1A to C) was suggestive of a 1.5 × 1 cm uniformly enhancing intradural extramedullary lesion at upper end of L2 vertebra. Patient underwent L1-L2 laminectomy and microsurgical total excision of a 1.5 × 1 cm cherry red, firm well encapsulated, highly vascular tumor (Figs 2A and B) was done. There was no adherence of the tumor to the dura or nerve root. Patient had an uneventful postoperative course and was discharged on the 3rd postoperative day. Patient became asymptomatic postoperatively. Microscopic examination (Figs 3A to C) of the hematoxylin-eosin staining revealed nests of round to polygonal tumor cells separated by fibrovascular septa, arranged in Zellballen pattern. The tumor cells had moderately abundant eosinophilic cytoplasm, central round nucleus with finely stippled chromatin, occasional nucleolus and intranuclear inclusion. Immunohistochemical staining was positive for chromogranin and synaptophysin.

DISCUSSION
Paragangliomas are neuroendocrine tumors of neural crest origin. They arise from neuroepithelial cell groups called
paraganglia. Majority of paragangliomas are adrenal in origin (85-90%), whereas the extra-adrenal type arises from jugular glomus of the carotid body.\(^5\) Paragangliomas with an extraspinal localization may be multiple. Within the central nervous system, sella turcica, petrous ridge and pineal gland are the commonly encountered sites.\(^6,7\) In the spine, they are intradural extramedullary in location. They are most commonly encountered in the lumbar region followed by cervical and thoracic regions.\(^8,9\) This pathological entity was first described by Miller and Torack in 1970,\(^10\) who called it a secretory ependymoma. Lerman and Kaplan\(^11\) were the first to coin the term

Figs 2A and B: Intraoperative images: (A) Cauda equina paragangliomas after opening of dural sac and (B) paraganglioma resected specimen

Figs 3A to C: (A) Gross specimen of paraganglioma, (B) H&E stain at 40× shows tumor cells arranged in Zellballen pattern and (C) diffuse positivity of tumor cells to chromogranin
paraganglioma of the cauda equina in 1972. Lumbar paragangliomas are usually located in the cauda equina and filum terminale region. Spinal paragangliomas are found in adolescent and the middle-aged population with a male preponderance. It clinically presents as low back pain as the main symptom in 50 to 80% patients with monoradicular symptoms in approximately 20 to 70% patients. The incidence of sensory or motor deficits or sphincter dysfunction is very low. These present as intradural extramedullary tumors and are often misdiagnosed as ependymoma based on clinical and radiological appearance. However, histologically ependymomas have a different immunohistochemical pattern (positive for glial fibrillary protein and negative for neuron specific enolase). Magnetic resonance imaging features on T1-weighted sequence is low/intermediate-signal intensity and intermediate/high-signal intensity on T2-weighted sequences. They enhance intensely with gadolinium contrast. These are vascular tumors and the blood flow causes serpigenous areas of signal voids. In addition to the typical MRI findings, the lesion has been known to present with an atypical hourglass configuration with flat superior portion and bulbous lower end. The size of the tumor varies from 1.5 to 10 cm. Histology coupled with immunohistochemistry forms the mainstay of diagnosis. Microscopic examination shows nests of round to polygonal tumor cells separated by fibrovascular septa, arranged in Zellballen pattern. Areas of hemorrhage and necrosis may be noted. Immunohistochemical staining is confirmatory and is positive for chromogranin, synaptophysin, neuron specific enolase and S-100 markers. Cauda equina paragangliomas are classified as World Health Organization Grade I spinal tumors. Definitive treatment is complete excision of the tumor.Incomplete surgical resection tumors or locally invasive tumors are benefited with radiotherapy. A recurrence rate of 4% is noted in these patients after gross total excision of tumor.

CONCLUSION

Paraganglioma of the cauda equina is an extremely rare condition. It clinically presents with low back pain with monoradiculer pain. These are benign neoplasm with total resection being the treatment of choice. Histopathology and immunohistochemistry form the basis of diagnosis. Patients with incomplete removal need postoperative radiotherapy and long-term follow-up.

REFERENCES