ABSTRACT
Paragangliomas are neuroendocrine neoplasm. These neoplasms originate from specialized neural crest cells and have association with segmental or collateral autonomic ganglia. Spinal paraganglioma is rare tumor. We are presenting a case of spinal paraganglioma in a 58 years old male presented with low back pain, his MRI spine showed extramedullary intradural tumor at the level of L1-L2 vertebra .Tumor was excised and patient was discharged after uneventful stay at hospital. Paragangliomas are WHO grade I slow growing tumor, radiologically paraganglioma resembles other lesions, such as schwannomas, ependymomas, meningiomas. Gross total resection of tumor is treatment of choice .Prognosis is good with very rare recurrence rate.

KEYWORDS: Paraganglioma, Spinal Cord, Cauda equine, Intradural, Neuroendocrine.

INTRODUCTION
Paragangliomas are low grade neuroendocrine neoplasm arising in paraganglionic tissue. They originate from the paraganglia in specialized cells that are derived from embryonic neural crest and function as a part of the sympathetic nervous system. These cells normally act as special chemoreceptors located along blood vessels, particularly in carotid bodies and aortic bodies. These neoplasm have high vascularity and characteristically show continuation with the tissues integrated in autonomic functions. Terminology of paraganglioma is generally based on anatomic site: e.g. carotid body paraganglioma, jugulotympanic paraganglioma, spinal paraganglioma. More than 90% of central nervous system paragangliomas manifest as carotid and glomus jugulare tumors. "Pheochromocytoma" is used exclusively for histologically identical tumors arising from the adrenal medulla. Paraganglioma in the spinal canal is rare, which is mostly located in the spinal cord and cauda equina. Spinal paraganglioma comprise 3.5% of tumors in the cauda equina / filum terminale region. Jugulotympanic paraganglioma are most common tumor of the temporal bone. They mostly affect adults with peak incidence in fifth decade of life. More common in males compared to females. Most common presenting feature is low back pain. Most of the spinal paragangliomas are nonfunctional tumors, only few functional paragangliomas are described in literature. Spinal paragangliomas are most commonly intradural extramedullary tumors in lumbo sacral area. Radiologically, it is similar to ependymomas, and histopathologically, to other neuroendocrine tumors. Therefore, it is difficult to diagnose paragangliomas prior to surgery, which is why there is a need to analyze such cases. Majority of spinal paraganglioma grow slowly. Total excision of the tumor is usually curative. This case report recounts a rare case of spinal paraganglioma of the lumbar region, its morphological features, immunohistochemical characteristics. Along with the differential diagnosis of paraganglioma in the cauda equina region of the spinal cord.

CASE REPORT
A 58 year old male presented to neurosurgery out patient department of era lucknow medical college & hospital with chief complains of pain in lower back since 8 months gradually increasing in intensity with time. Since past 15 days he also complained of pain radiating to both lower limbs more towards left side. On examination he showed positive leg raise test, rest of the neurological examination was within normal limits. He was advised routine blood investigations along with MRI spine. His routine blood investigations were within normal limits. MRI of spine showed an extramedullary intradural tumor measuring 3X2.4x1.8cm at level of L1-L2 vertebra. T1 and T2 weighted images showed intermediate signal intensity. The Patient underwent laminectomy at level of L1-L2 vertebra. Dura was opened a well circumscribed reddish mass was seen. Gross total resection of tumor mass was done securing the nerve roots. Tumor mass was sent for histopathological examination. Tumor tissue received at our department grossly showed a well circumscribed reddish brown, firm mass measuring 2.4X2X1.5cm. Cut surface of encapsulated tumor showed congested areas and grey brown areas. Representative sections were
Radiologically, paraganglioma resembles other lesions, such as schwannomas, ependymomas, meningiomas, dermoid tumors, or lipomas, which appear T1-weighted images as isointense or hypointense, and on T2-weighted images are hyperintense and show enhancement with Gadolinium administration (2).

Histopathologically, paragangliomas are well-differentiated tumors showing zellballen architecture surrounded by a delicate vascular network in which chief cells have fine granular eosinophilic cytoplasm and round to oval nuclei. These cells are disposed in nests or lobules surrounded by a single layer of indistinguishable spindle-shaped sustentacular cells. Occasionally, paragangliomas of the cauda equina region show features like angiomatous, adenomatous, and pseudorosette patterns similar to carcinoid tumors. Foci of hemorrhagic necrosis may be seen.

Immunohistochemical staining of paragangliomas shows positivity for synaptophysin and chromogranin; neuron-specific enolase is a sensitive marker but lacks specificity. Other neuroendocrine markers expressed by paragangliomas are CgA, Syn, CD56, and S-100.

Paragangliomas are benign neuroendocrine tumors that arise from specialized neural crest cells associated with paraganglia (3). The juglottympanic region paragangliomas comprise 80%-90% of cases, although they can grow anywhere from the base of the cranium to the bladder where there are chromaffin cells. Spinal paragangliomas are benign encapsulated tumors, most of which are spinal intradural tumors in the cauda equina region. Lerman et al. in 1972 were the first to use the term paraganglioma. Most commonly involved is the cauda equina region of the spinal cord (3-4%). Mean age at presentation is 46 years, more common in males. Most of the paragangliomas of the cauda equina are non-secreting. Rarely, these tumors are functional in very few cases and have been reported. Most of the patients present with nonspecific symptoms due to pressure on the spinal cord.

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Considering the differential diagnosis of ependymoma, a well-documented WHO grade I-III glial neoplasm in the cauda equina region which characteristically shows perivascular pseudorosettes, radially arranged cell groups around small vessels histologically. Sometimes, tumor shows both paraganglionic and ependymal differentiation, or often the lesion contains areas with ependymoma-like morphology, but PG-like IHC shows the pseudo chrysanthemum-like structure, similar to ependymoma leading to diagnostic confusion. Then ependymoma must be ruled out. Ependymoma are positive for GFAP and EMA but are negative for neuroendocrine markers (9, 3).
Often the morphology of tumor shows an obvious nest-like or sinusoids structure in such cases carcinoid tumor must be ruled out by IHC, carcinoid tumor are S100 protein negative. Many a times paragangliomas are very rich in blood vessels and need to be differentiated from meningioma, which express EMA, PR and Vim, and hemangioblastoma, which does not express Syn. Considering the differential diagnosis, if the tumor simultaneously expresses CK and neuroendocrine markers, the metastasis of hepatocellular carcinoma and gastrointestinal neuroendocrine carcinoma should be excluded (12).

Malignant transformation is uncommon in spinal paraganglioma. However it is very difficult to histologically distinguish benign from malignant tumor. Prognosis is good & recurrence is rarely encountered after complete surgical resection of the paraganglioma.

CONCLUSION

Spinal paragangliomas are mostly benign slow growing neoplasm. Paraganglioma is a very rare malignant tumor. This tumor should be distinguished from ependymoma, meningioma and hemangioblastoma, to avoid misdiagnosis, and missed diagnosis. Possibility of paraganglioma must be considered when we get a intradural lesion in spinal cord on MRI, immunohistochemical staining should be done in doubtful cases for accurate diagnosis. Surgical resection of tumor is curative. Recurrence and malignant transformation is very rare.

REFERENCES


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