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PARAGANGLIOMA OF LUMBAR SPINE – A RARE CASE REPORT

Sumaiya Irfan, Parul Gupta, Kshama Tiwari, Noorin Zaidi, Nirupma Lal, Sharique Ahmad

Department of Pathology

Era's Lucknow Medical College & Hospital, Sarfarazganj Lucknow, U.P., India-226003

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Address for correspondence
Dr. Parul Gupta
Department of Pathology
Era's Lucknow Medical College & Hospital, Lucknow-226003
Email: Drpg0679@gmail.com
Contact no: +91-8853100914

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 specalized 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 lumbosacral 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

taken and tissue was processed. H&E stained sections were examined under microscope. Section showed tumor cells arranged in zellballen(nested)pattern surrounded by delicate network of capillaries. The tumor cells were uniform round to polygonal with central round nuclei with stippled chromatin and inconspicuous nucleoli (Figure 1,2,3). A diagnosis of spinal paraganglioma was made after histopathological examination. IHC was positive for synaptophysin & chromogranin. Patient was discharged from hospital after an uneventful post-operative period.

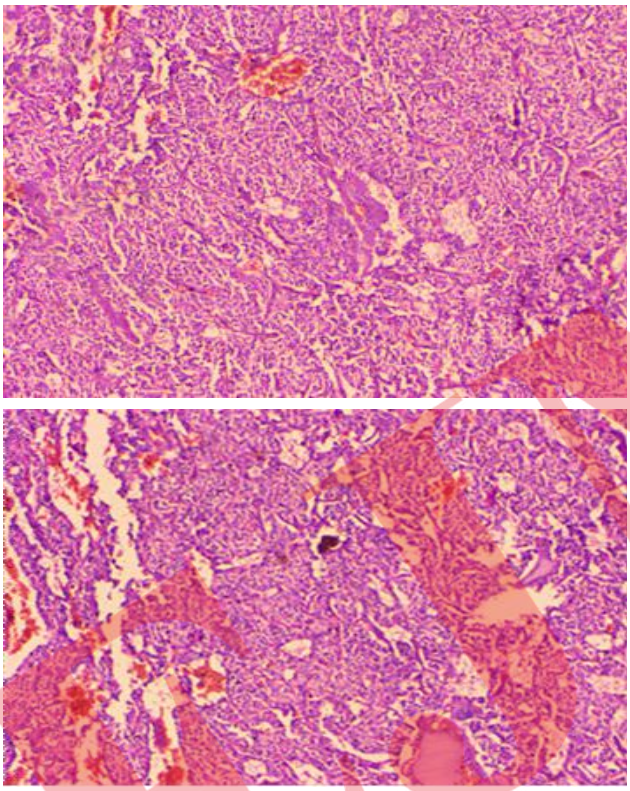

Fig. 1&2: (h&e ;10x)photomicrograph Shows Tumor Cells Arranged In Zellballen Pattern Surrounded By Delicate Network Of Capillaries

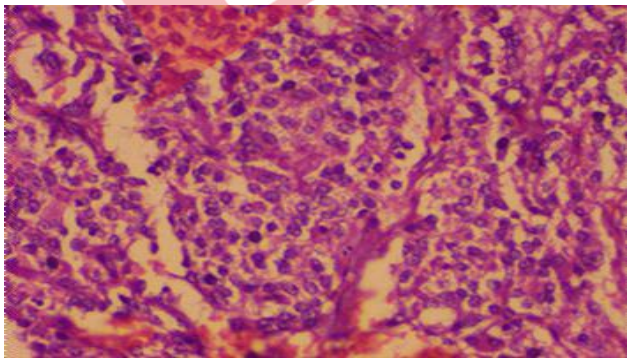

Fig. 3: (h&e ; 40x) Photomicrograph Shows Uniform Round To Polygonal Tumor Cells With Central Round Nuclei With Stippled Chromatin And Inconspicuous Nucleoli

DISCUSSION

Paraganglioma are benign neuroendocrine tumors that arise from specialized neural crest cells associated with paraganglia (3). The juglotympanic region paragangliomas comprise 80%-90% cases, although they can grow anywhere from base of cranium to bladder where there are chromaffin cells.Spinal paraganglioma are bengin encapsulated tumors, most of them are spinal intradural tumor in cauda equine region. Lerman et al. in 1972 for the first time used the term paraganglioma. Most commonly involved is cauda equine region of spinal cord (3-4 %).5Mean age at presentation is 46 years, more common in males. Most of the paragangliomas of cauda equine are non-secreting. Rarely these tumors are functional very few cases have been reported. Most of the patients present with nonspecific symptoms due to pressure on spinal cord.

Radiologically paraganglioma resembles other lesions, such as schwannomas, ependymomas, meningiomas, dermoid tumors or lipomas which appear T1-weighted image are isointense or hypointense, on T2- weighted images are hyperintense and with Gadolinium administration show enhancement (2).

Histopathologically, Paragangliomas are well differentiated tumors showing zellballen architecture surrounded by a delicate vascular network in which chief cells having fine granular eosinophilic cytoplasm and round to oval nucleus. These cells are disposed in nest or lobules surrounded by a single layer of undistinguishable spindle shaped sustentacular cells .Occasionally paragangliomas of cauda equina region show features like angiomatous, adenomatous and pseudorosette pattern which is similar to carcinoid tumor .Foci of hemorrhagic necrosis may be seen. Immunohistochemical staining of paraganglioma shows positivity for synaptophysin and chromagranin, these are sensitive and reliable markers; neuron specific enolase is a sensitive marker but lacks specificity. Other neuroendocrine markers expressed by paraganglioma are CgA, Syn, CD56 and S-100.

Considering the differential diagnosis of ependymoma a well documented WHO grade I-III glial neoplasm in cauda equine region which charcterstically 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,paraganglioma shows a 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 [12]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.

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