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Paraganglioma of the Lumbar Spinal Canal

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Abstract: Paragangliomas are rare neuroendocrine tumors arising from neural crest cells. Spinal cord being the rare site of occurrence, common being jugulo-tympanic and carotid region. In the spinal cord they present as intradural extramedullary lesion. On imaging they mimic neurofibroma and histopathology can be confused with ependymoma. We report a case spinal paraganglioma in a 39 years old male patient presenting at L2 - L3 level, which was clinically diagnosed as neurofibroma. Histopathological examination revealed the tumor to be a paraganglioma.

Keywords: Paraganglioma, spinal cord.

INTRODUCTION

Paraganglioma are neuroendocrine tumors that have origin from specialized neural crest cells. Their occurrence in the spinal cord is rare and is usually found in intradural extramedullary compartment in the lumbosacral region. Most frequent sites are the jugulotympanic region or the carotid region, but can develop at any site, where chromaffin cells are present, extending from base of cranium to bladder [1]. Chromaffin cells are specialized cells of the neural crest which are associated with sympathetic ganglia in fetal life. After birth these cells degenerate and most of the residual chromaffin cells will remain in adrenal medulla [2]. Paragangliomas of the central nervous system are rare with most of them presenting in the region of cauda equine of the spinal cord. Extraspinal localization of these tumors in the central nervous system is sella turcica, pineal gland and petrous ridge [3].

CASE REPORT

A 39 year-old male patient reported to the neurosurgery department with a history of weakness of left leg since 10 days. Patient had history of tingling and numbness. There was no history of trauma. His general examination, physical and systemic examination was normal. Motor examination and sensory examination were normal. Magnetic resonance imaging (MRI) of lumbosacral spine revealed a mixed intensity lesion on T2W images at the level of L2-L3 vertebral body. The lesion was enhancing after contrast administration (Figure-1). L2-L3 laminectomy was performed. There was grayish white small 2.5X1.8 cm, vertically oval, firm, moderately vascular tumor which was present in intradural extramedullary plane and was adherent to the rootlets and cords, compressing the cord more on right side. Subtotal decompression of tumor was done. The part adherent to the rootlets and thecal sac was left over. Post-operative period was uneventful. The tumor removed was sent to the pathology department for histopathological examination. Microscopically, the lesion showed polygonal to round tumor cells arranged in nests (Zellballen pattern) separated by thin fibrovascular septa and sustentacular cells (Figure 2). On immunohistochemistry, tumor cells are positive for chromogranin (Figure 3). Due to above features, the diagnosis of paraganglioma was made.

DISCUSSION

Spinal paragangliomas are rare “non secretary” tumors. Miller and Torack first described this tumor in 1970 as secretary ependymoma. In 1972 the term paraganglioma was coined by Lerman [4]. Most of the paragangliomas arise in the adrenal gland (85%-90%). Extraadrenal tumors are located in jugular glomus of the carotid body. Tumors arising in glomus region and carotid body are parasympathetic whereas the tumors arising in the spinal cord are typically sympathetic [5].

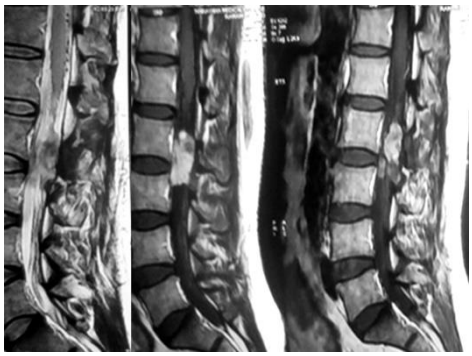

Fig-1: MRI of lumbosacral spine showing mixed intensity lesion at the level of L2-L3 vertebral body

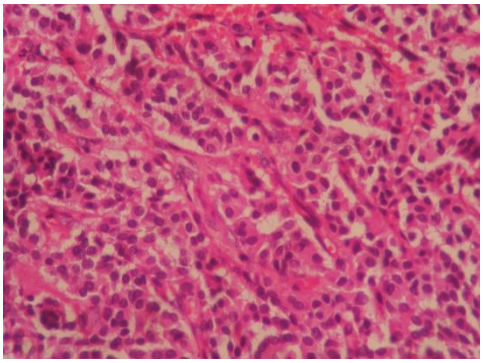

Fig-2: Tumor cells with nuclear monomorphism arranged in nesting pattern (H&E, X400)

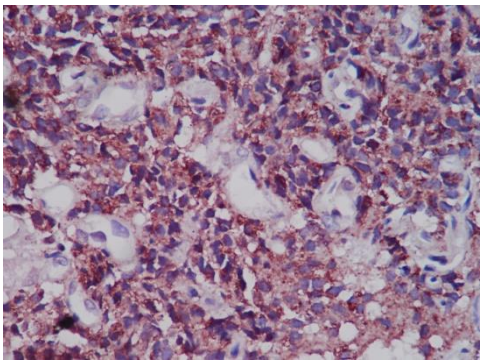

Fig-3: Tumor cells showing Chromogranin positivity (Chromogranin, X400)

Paragangliomas occurring in the spinal canal are usually found below the L1 vertebral level. Thoracic and cervical regions are less common sites. Clinical presentation of patients with spinal apraganglioma depends on the degree of spinal cord compression and the level of the cord affected. Most common presentations are motor or sensory deficits, back pain radiating down to the leg or fecal and urinary incontinence. Few tumors secrete catecholamines producing high blood pressure in patients [6].

On MRI, T1 weighted images show sharply circumscribed hypo or isointense mass which is markedly contrast enhancing on T2 weighted imaging it is hyperintense. The presence of low signal intensity rim known as “cap sign”, serpentine, ecstatic and congested vessels give diagnostic clue [7]. However these features are non-specific and similar MRI appearance can be seen in other tumors like schwannoma or ependymoma [8].

Grossly the tumors are encapsulated, grey-brown, and soft to firm in consistency with areas of hemorrhages and congestion. Microscopically the characteristic feature of paraganglioma is nesting pattern or “zell ballen” pattern. Typical nuclear monomorphism is noted. Tumor cells are polygonal in shape with round to oval nuclei showing stippled chromatin and inconspicuous nucleoli. Cytoplasm is usually finely granular and eosinophilic. Other cell type is spindle shaped Sustentacular or supporting cells. Perivascular pseudorosettes also can be seen areas of necrosis and hemorrhage may also be present. On immunohistochemistry tumor cells are positive for S-100, neuron specific enolase, chromogranin, synaptophysin [9].

Spinal paragangliomas are histologically similar to ependymoma which is mostly documented in the region of cauda equina. Epyndymomas are characterized by perivascular pseudorosettes in which the tumor cells are radially oriented around the small vessels. These tumors also have stippled chromatin. Due to these features histopathologically ependymomas can be confused with paragangliomas. Immunohistochemistry helps in differentiating tumor from paraganglioma. Ependymomas show positivity with Glial Fibrillary Acidic Protein (GFAP) staining and are negative for chromagranin, synaptophysin and neuron specific enolase which are positive in paraganglioma.

Treatment of choice for the spinal paraganglioms is surgical excision. But as these tumors are highly vascular, total surgical resection is achieved with difficulty. Subtotal resection is followed by local recurrence. Distant metastatic spread has also been described. In our case as the tumor was adherent to spinal rootlets, it could not be excised completely. Radiotherapy is preferred for locally invasive tumors and tumors with subtotal excision, though there is resistance for the treatment. Long term follow up is required as these tumors may recur after 20 years of surgical tretament [4].

CONCLUSION

Spinal paragangliomas are rare benign neoplasms which should be considered in the differential diagnosis of the intradural and extramedullary tumors occurring in spinal column. The radiological features are similar to schwanomma or ependymoma. Preoperative correct diagnosis is important because complete surgical resection is curative and subtotal resection leads to recurrence which may not be prevented by postoperative radiotherapy.

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