CASE REPORT

SPINAL PARAGANGLIOMA: A CASE REPORT

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ABSTRACT
Paraganglioma is a neuroectodermal tumour which originates from the chromaffin cells. Paraganglioma of spinal cord is uncommon lesion and usually presents as intradural tumour within cauda equina. We present a case of 42 year old female having spinal paraganglioma which developed in lumbar segment of spinal cord. The patient complained the pain in lower back and both legs with sensory deficit in left lower limb. Preoperative MRI showed a intradural mass. Patient operated with laminectomy and durotomy with complete excision of mass. Histopathology and Immunohistochemistry study showed the Zellaballen pattern and positive markers for Viamentin, Synaptophysin, Chromogranin A and Neurone specific enolase. Paraganglioma is a rare tumor with recurrence potential so total precision excision is the best option to achieve cure and to reduce the chance of recurrence.
Key Words: Paraganglioma; Spinal Tumour; Intradural Tumour

Introduction

Paraganglioma is a neuroectodermal tumour. Chromaffin cells belong to foetal neural crest. Paraganglioma originates from the chromaffin cells. Although it develops at the sites of normal paraganglia mainly intraabdominally, glomus jugularae or carotid bodies; it may be found in other part of the body also. The majority of Paraganglioma are non-functioning and benign. However, they may produce catecholamine excess or have malignant potential.[1,2] Paraganglioma of CNS region has been seen in pineal region, petrous ridge and sella turcica.[3-5] Paraganglioma of spinal cord is uncommon lesion. It usually presents as intradural tumour within cauda equine.[2-6] Here we present a case of spinal Paraganglioma which developed in lumbar segment of spinal cord.

Case Report

A 42 year old female was admitted in department of orthopaedics, GT hospital, Mumbai, with complain of pain in lower back with bilateral radiating leg pain since last 20 years. Her pain aggravated by sitting, standing and bending. Patient also had tingling and numbness in left lower limb. On clinical examination straight leg raise test was positive at 40 degrees, and she had a definitive sensory deficit from L3 dermatatomal distribution and below for touch and pin prick. There was no motor deficit. Deep tendon reflexes were hypoactive in both lower limbs.

Preoperative anteroposterior and lateral x-ray of lumbosacral spine showed no abnormality except L5-S1 spondylolisthesis while MRI of L-S spine revealed an L2-L3 intradural mass of size (2.5X1.5X1 cm) that exhibited isointensity with respect to spinal cord on T1 weighted image and hyperintensity on T2 weighted sequence. Tumour was well circumscribed and surrounded by band of low signal on both T1 and T2 weighted images.

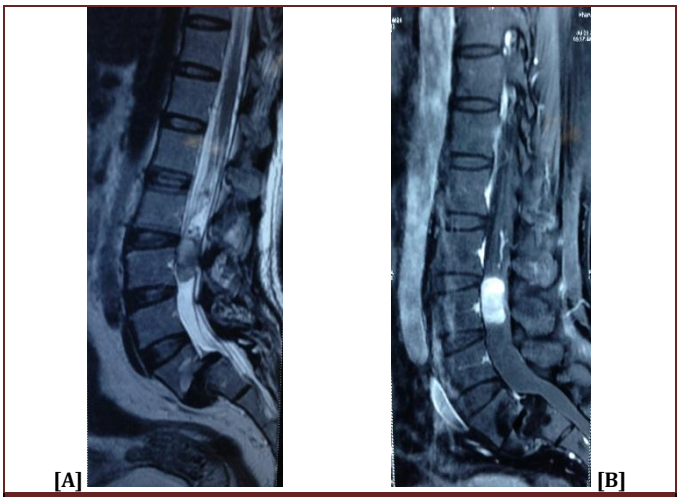

Figure-1: [A] T2 weighted MRI image showing intradural mass at L3 level; [B] MRI picture shows enhancing lesion at L3 level

Patient was operated with laminectomy of L2 and L3 in prone position under general anaesthesia with induced hypotension. Total excision of the intradural mass was done through standard durotomy. The leak of CSF during tumour excision controlled through the careful placement of neurosurgical patties beneath the dura while separating the mass from cauda. The tumour was reddish-brown, encapsulated measuring 2.5 X 1.5 X 1 cm. Postoperative she had good recovery and her leg pain was gone. Tingling and numbness in lower limb was completely resolved. On 5th post op day she was walking with frame type LS. Belt and with the help of a walker. After 10 days postoperative she was walking independently without any walking aid. Stitches removed on 14th postoperative day.

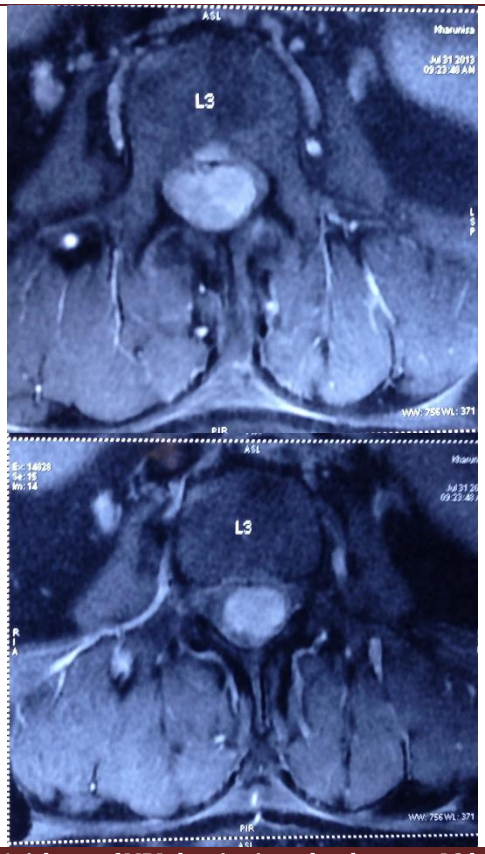

Figure-2: Axial cuts of MRI showing intradural mass at L3 level

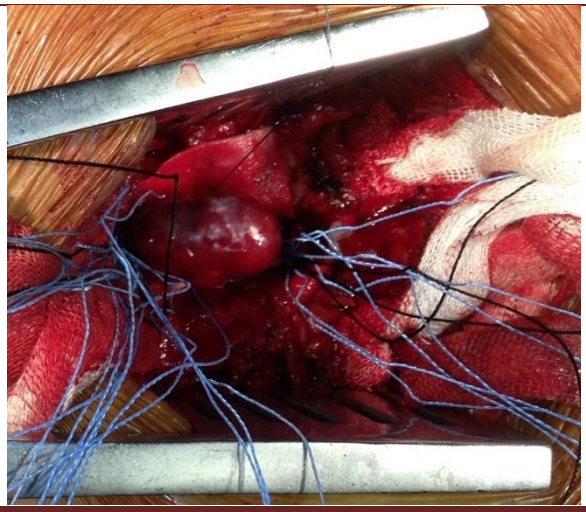

Figure-3: Intraoperative picture while excision of tumour mass.

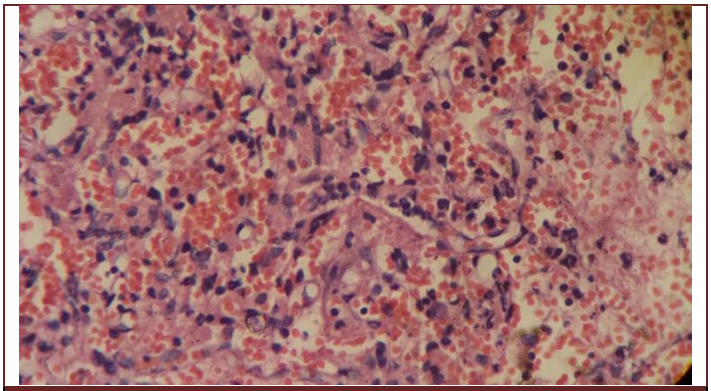

Figure-4: Histopathological picture showing chief cells separated by fibrovascular septa (Zellaballen pattern)

The histopathological examination of intra-dural tumor mass showed typical picture of Paraganglioma. The typical Zellaballen pattern meaning in German "balls of cells". Which have nest of chief cells, separated by fibrovascular septa. The immunohistochemistry of cells showed immune reactivity to Viamentin, Synaptophysin, Chromogranin A and Neurone specific enolase.

Discussion

Extraadrenal pheochromocytoma of neural crest derived sympathetic ganglia is known as Paraganglioma.[1] Paraganglioma is a rare tumour and accounts for 0.3% of all neoplasm.[7] Paraganglioma unlike pheochromocytoma are rarely functional and mostly arise in glomus jugulrae or carotid bodies (80-90% of cases).[3,4,6,8,9] However, they have been observed in numerous other sites including larynx, thyroid, mediastinum, GI, respiratory and urinary tract, vagus nerve and orbit.[10-12] In CNS they have been described in pineal and pituitary gland, cerebellopontine angle, spinal cord and cauda equine.[13,14]

Spinal Paraganglioma are rare and most of the reported cases have occurred in cauda equina or filum terminale.[2-6,10,12,15] Cauda eqina paraganglioma represents approximately 3-4% of lesion in this region.[16,17] These are benign and slow growing tumours with low proliferative activity.[6,18,19] However, malignant metastatic lesion have been reported by few authors.[20,21] Despite their neuroendocrine origin only few cases with functional hormonal activity have been found.[15,22] Spinal Paraganglioma usually presents with low backache with associated radiculopathy. Sensory and motor deficit are uncommon. Bowel and bladder dysfunction are rare.[3]

MRI is the investigation modality of choice. MRI characteristics of Paraganglioma have been well studied.[23,24] Typical Paraganglioma appear isointense to conus medullaris on T1 weighted sequence and hyperintense on T2 weighted sequence as seen in our case. Although, it may identify the lesion it is not diagnostic and it may be mistaken for schwannoma, ependymoma, meningioma or solitary metastasis.[24,25] At surgery lesion tend to arise from filum or cauda equina and are well encapsulated. Histological diagnosis are based on zellabellen pattern and positive immunohistochemistry markers.[21]

Prognosis depends on ability to achieve complete excision of mass.[4,19] Encapsulated lesion therefore have good prognosis. If lesion cannot be removed completely then radiotherapy is an option although radioresistance has

been seen.[12,26,27] Recurrence with incomplete removal, and even metastasis outside spinal canal, has been reported.[12,13,20-22,28,29] Local recurrence rate of 12% has been seen.[8,30] Hence long term follow up is needed in case of incomplete removal.

Conclusion

Paraganglioma of cauda equina or filum terminale is a rare condition. This presents with low backache and variable sensory or motor symptoms. Although MRI is not diagnostic it provides valuable information in precisely locating the tumor mass and helps in surgical management. As paraganglioma may recur, total precision surgical excision is the best option to achieve greater chance of cure and also to reduce recurrence rate. Final diagnosis comes after histopathology confirmation of nature of cells in tumor.

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