

A Series of 2 Rare Cases of Filum Terminale Paraganglioma – A Case Report

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Abstract: Paraganglioma is a neuroendocrine tumour arising from non chromaffin paraganglion cells anywhere in the body. Most common site is adrenal gland. In central nervous system, they are found at petrous ridge, pineal gland, sella turcica and spine. In the spine, cauda equine localization of paraganglioma is common however it is much rarer in filum terminale site. We are presenting two cases of middle age gentlemen with complaints of radicular pain of lower limb. MRI lumbar spine was suggestive of intradural tumor arising from filum terminale at lumbar site. The tumor was completely excised and histopathology revealed to be paraganglioma. We hope this case will be documented in the literature and help in understanding this entity more clearly.

Keywords: Paraganglioma, Filum terminale.

INTRODUCTION

Paraganglioma of the filum terminale is an uncommon neuroendocrine tumour and is classified as WHO grade I [1]. The term paraganglioma was coined by Lerman in 1972 [2]. Most commonly it arises from adrenal medulla. Most common extra adrenal site of paraganglioma is carotid body as glomus jugulare tumour [3]. Paragangliomas of central nervous system are very rare. Their origin in the spinal canal is even more rare [4]. In the spinal canal most commonly they arise from cauda equina and very rarely in the filum terminale [5]. Low back pain and sciatic type of pain are the main presenting symptoms [6]. Diagnosis of choice is Magnetic Resonance Imaging and they are mostly Intradural Extramedullary in location [7]. However, diagnosis depends on pathological confirmation of typical Zellballen pattern and immunohistochemical positivity for chromagranin and S100 proteins [8]. Complete removal of the tumour is the ultimate goal of neurosurgery and as the tumor is benign in nature prognosis is usually good [9]. When complete removal is not possible, radiotherapy may be considered [10]. Here we report the rare case of filum terminale paraganglioma which was treated successfully in our institution.

CASE HISTORY

Patient 1- 49 years old gentleman was admitted with complaints of pain in the right side of lower back in September 2015 since 1 month which was sharp electric like sensation radiating posteriorly up to mid-thigh region. Bladder and bowel movements and sensation were normal. On examination, there was no tenderness and no sensory loss or motor weakness. Tendon reflexes were present with normal anal tone. MRI of spine showed well enhancing and well

circumscribed tumour at L3-L4 level. Pre-operative diagnosis was kept as intradural tumor and total excision of the tumor was done.

Patient 2) 32 years old gentleman was admitted with complaints of pain in the lower back radicular type radiating to both lower limbs, right more than the left since 8 days. Bladder and bowel movements and sensation were normal. On examination, there was no tenderness and no sensory loss or motor weakness. Tendon reflexes were present with normal anal tone. MRI of spine showed well enhancing and well circumscribed tumour at inferior border at L2 level. Pre-operative diagnosis was kept as intradural tumor and total excision of the tumor was done. Intra operatively, tumour was found to be arising from filum terminale which was identified as fibrous septae which was bluish whitish in colour with a vessel on its surface. Post operatively patient pain reduced significantly and was later on discharged.

Histopathological report in both patients showed classical Zellballen pattern and immunohistochemistry was positive for synaptophysin and chromagranin in both cases. However, there was evidence of high MIB-1 index in second case with proliferative index of 16%. Thus the diagnosis of Filum terminale paraganglioma was made in both cases.



Fig-1: MRI Lumbar spine showing L2 level well circumscribed hypointense lesion (Filum Terminale) in patient 1



Fig-2: post contrast MRI lumbar spine showing enhancement of the tumor in patient 1



Fig-3: Case 2- MRI Lumbar spine sagittal view showing an intradural tumor at L2-S1 level (Filum Terminale) in patient 2

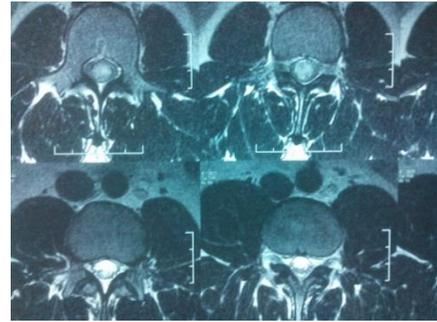


Fig 4: Axial MRI sequence showing complete excision of the tumor



Fig-5: post-operative MRI showing complete excision of tumour.

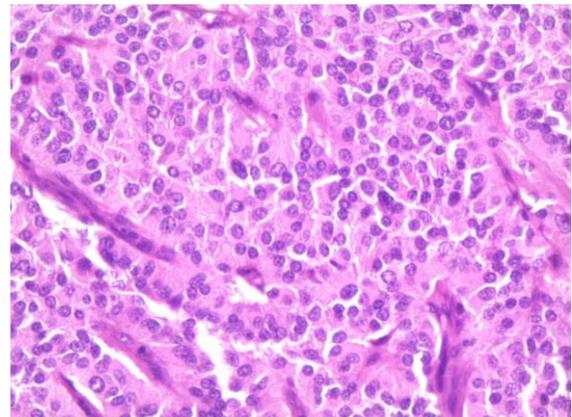


Fig 6 : Histopathology slide showing Zellballen Pattern

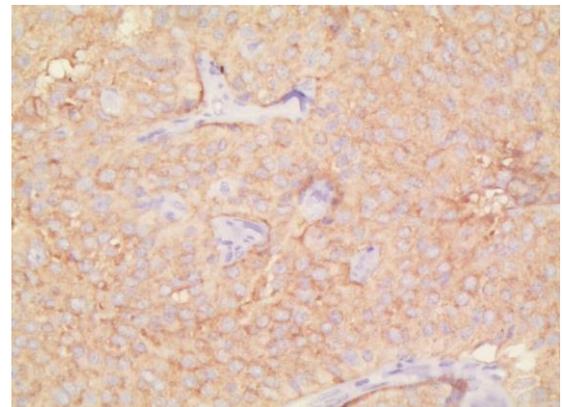


Fig 7 – Slide showing Chromagranin A positivity.

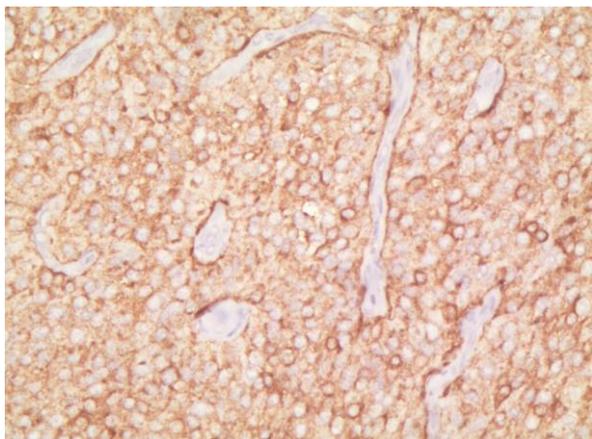


Fig 8 – Slide showing Synaptophysin positivity.

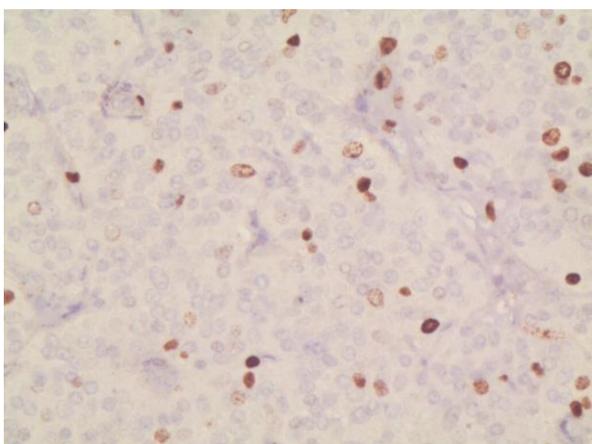


Fig 9 – Slide showing High MIB – 1 index

DISCUSSION

Paragangliomas arise from non chromaffin cells present anywhere in the body. In about 80-85% cases, adrenal medulla is the most common site. Extra adrenal paragangliomas can be found in organ of Zuckerkandl. In Head and Neck region they are most commonly seen in carotid bodies. In central nervous system they are found at petrous ridge, pineal gland, sella turcica and spine.

Spinal paragangliomas are rare pathology with incidence estimated to be 0.07 per 1, 00,000 population. Although tumours of the extra spinal region are parasympathetic, paragangliomas of spinal origin are mostly sympathetic.

Some of this spinal paragangliomas have familiar occurrence with chordomas, leiomyosarcomas and pituitary adenomas.

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