ORIGINAL RESEARCH PAPER
Volume - 12 | Issue - 09 | September - 2023 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

FILUM TERMINALE PARAGANGLIOMA : A REPORT ON 2 RARE CASES AND REVIEW OF LITERATURE

Neurosurgery

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ABSTRACT

Paragangliomas are neuro-endocrine tumors originating from the adrenal gland. They are usually benign and nonfunctioning, rarely seen in central nervous system. More than 90% of central nervous system paragangliomas are manifested as carotid and glomus jugulare tumors. Spinal paragangliomas are quite rare. Spinal paragangliomas are rare extra adrenal tumors with good prognosis if complete resection is done which is often possible. Presenting 2 cases of filum terminale paragangliomas examined and treated in our institution. Paraganglioma is a very rare malignant tumor. This tumor should be distinguished from ependymoma, meningioma and hemangioblastoma, to avoid misdiagnosis, and missed diagnosis and also for further evaluation and follow up.

KEYWORDS

Differential Diagnosis, Immunohistochemistry, Lumbar Spine, Neuroendocrine Tumor, Paraganglioma

INTRODUCTION

Paragangliomas are rare neuroendocrine tumors extra adrenal in origin, composed of chromaffin cells. 90% are found in jugular glomus and rest 10% associated with conus medullaris of spine. About close to 220 cases have been reported , which makes it quite a rare entity and hence the clinical presentation and further management need to be reported to enhance the knowledge about the disesase.[1,2]

This report of 2 cases discusses the presentation, clinical and radiological findings and the review of literature as well.

Case Presentation

First Case : A 64 year man, farmer by occupation, presented with 6 years history of progressive low back pain with bilateral lower limbs radicular pain and bladder involvement in the form of urinary incontinence. The reflexes and sensorimotor examinations were found to be within normal limits.

MRI revealed an irregular, intradural, T1, T2 and GRE hypointense lesion of size about 31*17*14mm with heterogenous enhancement post contrast at L3 and L4 levels displacing the cauda equina roots laterally. Also sacralization of L5 was noted. (Figure 1 and 2) The differential diagnosis included schwannoma, intradural sequestered disc material and calcified meningioma.

Further investigations for preparing the patient for surgery was carried out and no other comorbidity found. Surgery was planned. L3-L4 laminectomy was done to relieve the pain and gross total excision of tumor done and sent for histopathological examination after satisfactory dissection from nerve roots.

Histopathology revealed paraganglioma consistent with World Health Organization (WHO) grade I. Immunohistochemistry results showed that the chief cells were positive for neuron-specific enolase (NSE), Syn and CgA, and Sertoli cells were positive for S-100.

Second Case: A 26yr old male was presented with low back pain with bilateral L5 & S1 radiculopathy with lower motor neuron bladder involvement for last 6 months. There were no signs or symptoms indicating catecholamine hyper-secretion, such as hypertension, psychomotor distress or headache. Both lower limb straight leg raising (SLR) test was positive without any other neurologic deficits.

MRI of lumbosacral region revealed a large lobulated altered signal intensity enhancing intraspinal intradural extramedullary space occupying lesion at the level of L4 to S1 measuring 70 mm x 37 mm x 24 mm in size (Figure3 (a-d)). There was widening of spinal canal & scalloping of posterior cortex of L5 & S1 vertebral bodies. A provisional diagnosis of filum terminale myxopapillary ependymoma was made.

All the ancillary preoperative investigations were normal. Enblock excision of the tumor was done after dissecting the filum terminale cranial to the lesion and from the caudal roots. There was no evidence of infiltration of the caudal roots. An elongated, well encapsulated, cherry red highly vascular tumor was encompassing the cauda equina nerve roots and filum terminale. The histopathology was reported as paraganglioma (Figure 4a and 4b). Immuno-histochemical testing was gave confirmatory diagnosis as positive reaction for chromogranin A (Figure 4c) and synaptophysin.

DISCUSSION:

Paraganglioma being a unique neuroendocrine tumor, is often well encapsulated and benign. Derived from special neural crest cells in segment or lateral autonomic ganglia, it consists of nest like arrangement of chief cells along with differentiated neurons , surrounded by sertoli cells and fibres.[3] It is usually found near cauda equina regions and hence sometimes referred to as CEP, firstly described by Miller.[4] It was explicitly explained by Lerman et al in 1972, it equivalents to WHO grade I.[5,6] Overall it accounts for 3.4% to 3.8% of cases of paraganglioma in CNS. Rare reports of intracranial paragangliomas have also been reported, where tumors were located in sellar region, cerebellopontine region and cerebellar parenchyma.[7]

CT shows homogeneous iso-density lesions while on MRI, lesions appear with clear margins and occasionally displays some cystic areas. The tumor presents with low to intermediate signal intensity on T1WI, and intermediate to high signal intensity on T2WI. Since the tumor is rich in blood vessels, an empty area will appear in the background of slow blood flow with an enhanced signal of the tumor. This is called “salt and pepper disease” on T2W1 images, which is one of the features of paraganglioma.[8,9] The gross pathologic manifestations include an intact capsule, which is dark red or brown. Multifocal hemorrhage occurs after the removal of tumor surface tissues during the operation [10]

These expresses neuroendocrine markers such as CgA, Syn, CD56 and S-100. Before surgery for definite lesion is planned, the metastatic workup to rule out hepatocellular carcinoma and gastrointestinal neuroendocrine carcinoma should be carried out. When CEP presents with a pseudo chrysanthemum-like structure, it is very similar to ependymoma, and immunohistochemical staining identification is required. Ependymoma expresses GFAP and EMA but does not express neuroendocrine markers.[11] When it has an obvious nest-like or sinusoids structure, it needs to be distinguished from carcinoid tumors.

The immunohistochemical staining of carcinoid tumors is negative for S-100 protein.[11]

Spinal paragangliomas are mostly benign and rarely malignant. It is difficult to determine benign and malignant tumors by histology. In this report, immunohistochemical staining could have been used to effectively analyze the shape of paraganglioma. Malignant paraganglioma is a possibility if spread is identified to lymph nodes or distant sites. this greatly influences the treatment strategy as complete removal of tumor by 1-stage resection is the first choice for the treatment of paraganglioma in the spinal canal. But if in case, the margins are not free and hence resection is incomplete, the recurrence would easily occur.[12] Therefore, the range of excision should be extended to remove the tumor completely under the guidance of accurate pathological diagnosis. The prognosis of the 1st case is being hoped to be good as he is currently pain free, however there is no improvement in his bladder involvement and is in follow up after being discharged on 12/01/23. The second case is doing well as of now. However, pre-operative therapies such as embolization may be beneficial depending on tumor size and vascularity to help minimize intra-operative bleeding. In rare cases of recurrence, usually after suboptimal resection, radiotherapy may be warranted. In our patients, a gross-total resection was achieved and confirmed by post-operative imaging.

CONCLUSION:

It is not always possible to remove the tumour totally because of adherence to the neural structures and it's vascularity and to avoid postoperative morbidity and neurodeterioration. Gross-total surgical resection is the primary goal in ensuring an excellent prognosis. Careful attention should be given to differentiate Spinal Paraganglioma from other more aggressive spinal tumors. As such, we recommend spinal paragangliomas be considered in the diagnoses of IDEM tumors with a coexisting finding of spinal schwannomas.

Images of case 1 : Fig 1(Pre-operative) irregular, intradural, T1, T2 and GRE hypointense lesion heterogenous enhancement post contrast at L3 and L4 levels

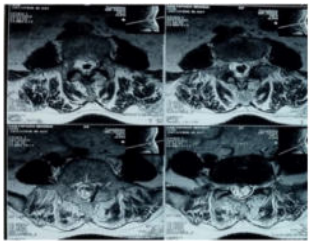

Fig 1a. Axial section LS Spine

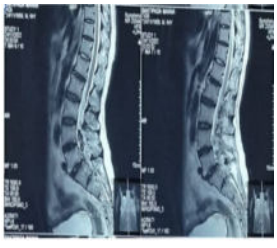

Fig 1b. Sagittal section LS spine

Fig 2. Post op images showing gross total excision of L3L4 spinal SOL

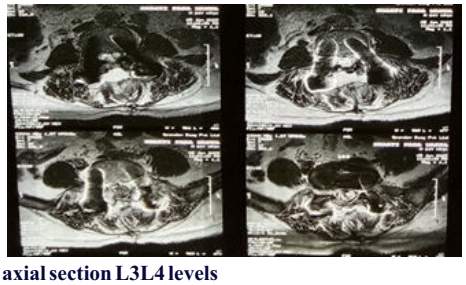

Fig 2a. axial section L3L4 levels

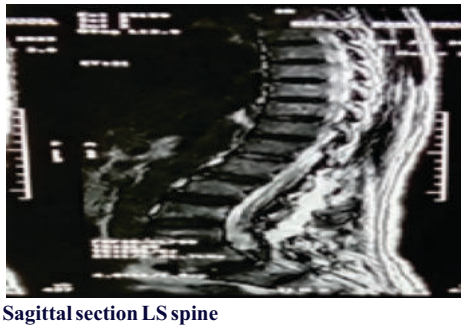

Fig 2b. Sagittal section LS spine

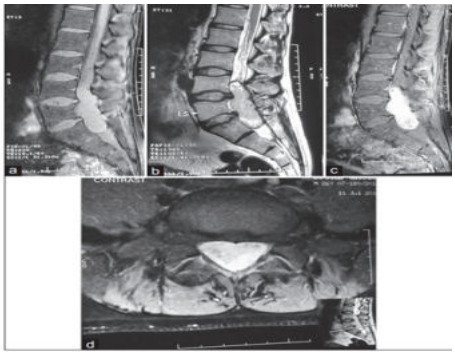

Fig 3 (a)Sagittal T1W MRI reveals an isointense intraspinal intradural tumour extending from L4 to S1, (b) In sagittal T2W MRI the lesion was mixed intensity more of hyperintense, (c) On sagittal T1W MRI with gadolinium, tumor was brilliantly contrast enhancing, (d) On axial T1W with gadolinium, the tumor occupied whole of intraspinal space

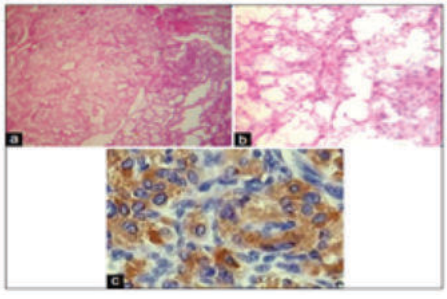

Figure 4: (a) Hematoxylin-eosin [×100] stained was showed zellballen pattern as the tumor tissue arranged in typical nests and lobules surrounded by many capillaries, (b) Hematoxylin-eosin [×400] individual cells are round to polygonal with abundant eosinophilic cytoplasm and centrally placed nuclei with stippled chromatin, (c) Immunostain [× 400] photomicrograph is showing diffuse positivity of tumour cells for chromogranin A

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