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Case Report

Intramedullary Paraganglioma of Filum Terminale: A Rare Case Report

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Abstract: Paragangliomas are neuroendocrine tumors that arise from neural crest cells of the sympathetic or parasympathetic autonomic nervous system, most frequently found in the Glomus jugulare and the carotid bodies. Paragangliomas of the Central nervous system are uncommon. In spinal cord they arise intradural extramedullarily with most common site being filum terminale. Here we presented one of such rare case of paraganglioma of spinal cord in a 45-year-old female patient arising from the filum terminale. Clinical symptoms include as that of any spinal tumor. Magnetic Resonance Imaging is the choice of investigation for spinal tumors preoperatively but histopathological examination is confirmatory. Surgical excision is the definitive treatment. As they are included in grade I category, prognosis is good.
Keywords: Filum terminale, Paraganglioma, Parasympathetic, Sympathetic.

INTRODUCTION

Paragangliomas are neuroendocrine tumors arising mostly from adrenal medulla. Extra adrenal paragangliomas are rare and 90% arise in head and neck in carotid body or glomus jugulare [1]. Paragangliomas of Central nervous system (CNS) are rare and arise commonly from cauda equina or filum terminale. They are mostly intradural extramedullary in location [2]. Diagnosis depends on both radiological and pathological correlation. They have to be differentiated from other spinal tumors, as it will aid in treatment and prognosis.

CASE REPORT

A 45 year old female patient came with chief complaints of backache since 3 months and weakness of both lower limbs since 3 days. Her past history was nil significant. Routine blood investigations and Echo Doppler were normal. MRI was done which showed altered signal intensity in region of conus medullaris and filum terminale with lesion measuring 17 x 33 mm, which was isointense on T1W (Fig. 1& 2) and hyperintense on T2W. Contrast study showed heterogenous enhancement. Based on these findings probable diagnosis of intramedullary spinal tumor possibly ependymoma was given. Surgical treatment of L1, L2 and L3 laminectomy with excision of the tumor was done and specimen sent to pathology department for histological examination.

Histological Examination

Grossly we received multiple grey white to grey brown soft tissue bits all together measuring 2.5 x 2 cms. Cut section showed grey white to grey brown areas. Tissue was processed and stained with Hematoxylin and Eosin (H&E) stain and examined under microscope. Microscopy revealed tumor tissue arranged in typical nests and lobules (Zellballen pattern) surrounded by many capillaries (Figure 3). Individual cells are round to polygonal with abundant eosinophilic cytoplasm and centrally placed nuclei with stippled chromatin (Figure 4). Based on these features diagnosis of Paraganglioma was given.

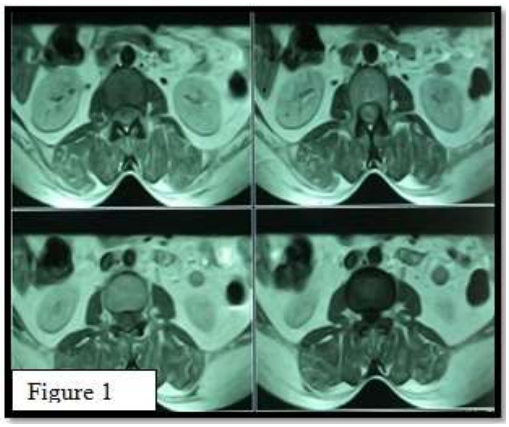

Fig. 1: Cross sectional view of MRI lumbar region showing isointense lesion

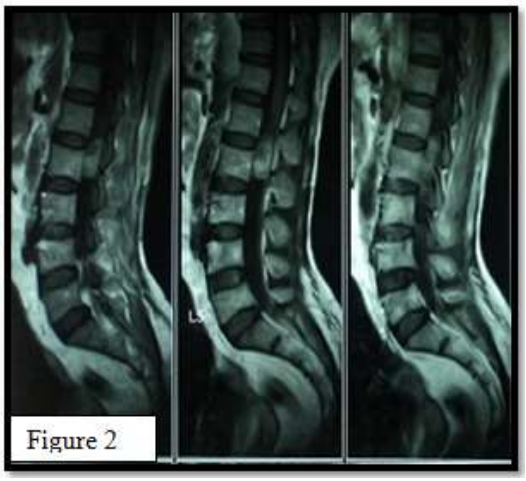

Figure 2: Longitudinal view of MRI lumbar spine with isointense lesion at L2 level

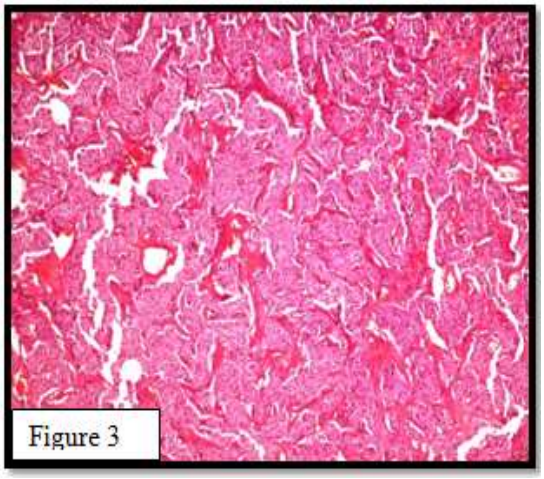

Fig. 3: H & E 10x showing tumor tissue arranged in Zellballen pattern

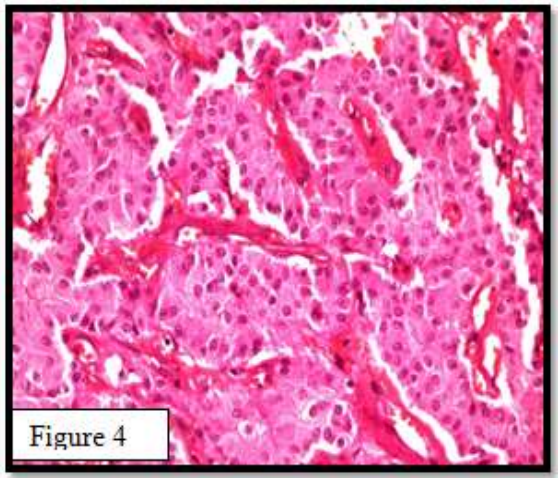

Fig. 4: H & E 40x showing tumors cells arranged in nests with abundant granular cytoplasm and centrally placed nuclei

DISCUSSION

Extra adrenal Paragangliomas are tumors that arise from paraganglia cells of the sympathetic or parasympathetic autonomic nervous system, most frequently found in head and neck region arising from Glomus jugulare and the Carotid bodies. Paragangliomas of the CNS are rare [3]. The vast majority present as spinal tumors usually located in the intradural, extramedullary compartment at the level of the cuada equina and the filum terminale, followed by thoracic and cervical regions[4,5]. Paragangliomas of cauda equina are seen mostly in third and fourth decade, with a slight male preponderance. Clinical symptoms are non-specific and include radiculopathy due to compression of mass on spinal nerve roots. Our case was female with complaints of backache and weakness. MRI is the investigation of choice for spinal tumors.

On MRI, paragangliomas are isointense on T1W image and hyperintense on T2W image with heterogenous enhancement on contrast [1,6]. They are located intradural extramedullarily but in our case location of tumour was intramedullary, which is rare. Differential diagnosis of tumors of the cauda equina region includes ependymoma, neurinoma, hemangioblastoma, meningioma [6]. Of these ependymoma is most common in that site hence, it has to be differentiated. Immunihistochemically paragangliomas are sensitive to Neuron specific enolase [NSE], but lack specificity, but synaptophysin and chromogranin are sensitive and reliable. Sustentacular cells are uniformly reactive for S-100 protein. IHC is most commonly used for differential diagnosis in case of difficulty on H&E staining.

Complete surgical excision is the treatment of choice. According to WHO classification they are classified under Grade I category hence has good prognosis when excised completely [1]. Recurrence after complete excision is rare [7]. Recently, a hereditary paraganglioma syndrome (PGL) had been classified based on molecular genetics: succinate dehydrogenase (SHD) subunit mutations which compose portions of mitochondrial complex II [4].

CONCLUSION

Paragangliomas are rare tumors of spinal cord arising commonly from cauda equina or filum terminale. They often present with non specific symptoms mainly due to mass effect on spinal roots. MRI is the investigation of choice. Though rare, they are kept in differential diagnosis of tumors of spinal cord and have to be differentiated from other tumors, as prognosis is excellent after complete surgical excision. Recurrence after excision is rare.

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