Cauda Equina Neuroendocrine Tumor - A Case Report

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Abstract
Cauda equina neuroendocrine tumors are rare tumors; very few cases have been reported in literature. The presenting report is a case of 53 years old male hailing from Kurigram district. He had low back pain for 2 years radiating to both lower limbs. The physical examinations were within normal limit. The MRI showed an oval intradural lesion in L_2-L_3 level with T_1 and T_2 signal intensity L_2 and L_3 Laminectomy followed by total resection of tumor was done. Routine H&E stain and immunostain confirmed the case as Cauda equina neuroendocrine tumor.

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Keywords: Paraganglioma, Cauda equina.

Introduction

Paraganglioma is benign and slow growing neuroectodermal tumor commonly found in the adrenal medulla, carotid body and glomus jugular,[1] resulting from the migrated cells from the neural crest to the level of autonomic nervous system. The cauda equine paraganglioma are very rare.[2]

In 2022 WHO neuroectodermal tumor classification it is designated as cauda equina neuroectodermal tumor.[3]

Case Report

A 53 years old male patient having low back pain for 2 years with radiation to both lower limbs. Locally he was treated treatment with nonsteroidal anti-inflammatory drugs and common analgesics. But no significant improvement achieved. On admission in NINS examined thoroughly. All of his physical and routine examinations were within normal range. The MRI showed an oval intradural extramedullary lesion. The patient was operated through posterior approach; laminectomy and total resection of tumor was done. Grossly, the tumor was a single nodule measuring 1.5x1 cm. The cut surface was dark brown. Under microscope it was a capsulated lesion composed of numerous endothelial lined vascular structures containing blood. The interstitium reveals monomorphous polygonal cells with spherical nuclei and abundant eosinophilic granular cytoplasm. The cells were disposed in nodules, trabeculae and interconnecting bands and cords. On the basis of microscopic examination it was diagnosed as paraganglioma or cavernoma. Immunostain was done for Pan-CK, Vimentin, Ki67, Synaptophysin, Chromoganin A and S-100 protein. The tumor shows defuse positivity for S-100p and Pan-CK. Also tumor cells are positive for Vimentin, Synaptophysin and Chromoganin A. Ki-67 was positive in 4-5% cells.

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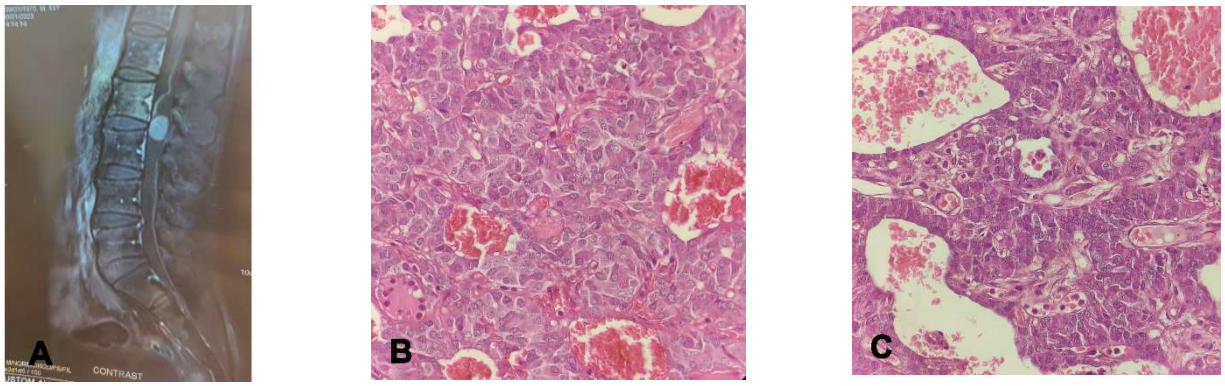

Figure 1. (A) Sagittal contrast enhancement (MRI) demonstrating inhomogenous enhancement after gadolinium enhancement. (B) & (C) H & E stain histopathological slide (x40).

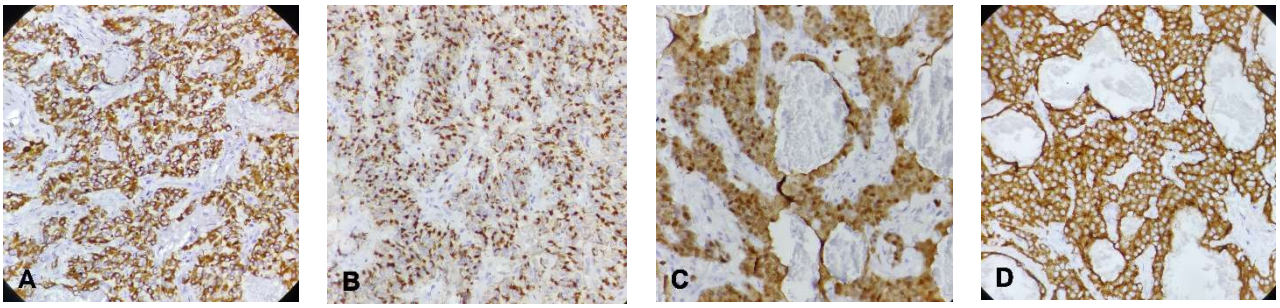

Figure 2. Showing (A) positive for Pan-CK (40x), (B) Diffuse expression of Chromoganin A by tumor cells (40x), (C) diffuse expression by S-100 (40x), (D) tumor cells are diffusely positive for Synaptophysin.

Discussion

Cauda equina paragangliomas are rare tumors representing 2.5 to 3.8% of Cauda equina tumors.[2]The presenting case is a middle aged male of 53 years Mr. Landi reported a slight male predominant in his study on cauda equina tumor.[4]The clinical presentation of cauda equina NET is non-specific. Though paraganglioma has the potential to secrete catecholamines, most of the tumor may appear to be clinically nonfunctional. The most common complaints of cauda equina NETs were back pain and sciatica.[5]In the presenting case, main complaint was radiating low back pain. Magnetic Resonance Imaging (MRI) plays an important role in addressing the cauda equina NETs. The seusually present as inhomogeneous, isointenseT1 and hyper intense on T2, with heterogenous on Gadolinium contrast enhancement.[6]In this case, there was an oval to rounded intradural lesion in T1 isointense, and in homogenous contrast enhancement after injection of Gadolinium. Myxopapillary ependymoma, schwannoma, meningioma and hemangioblastoma are the differential diagnosis of cauda equina.[5] The diagnosis was not established until the end of histological and immunohistochemicalstudy.[2,3,7,8] The present therapeutic approach consists of

complete surgical resection, incomplete resection exposes to a risk of recurrence in the first year.[9,10] The index case had complete resection of the tumor without any complication and follow-up after 3 months was eventless.

Conclusion

Cauda equina neuroendocrine tumor (Paraganglioma) in an isolated tumor in WHO 2022 classification of neuroendocrine tumor. It is a benign neuroendocrine tumor. Clinical features and MRI is not sufficient for diagnosis. The confirmatory diagnosis is mainly depends on histopathology and immunohistochemistry. Complete surgical excision remains the ideal treatment to minimize the risk of recurrence.

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