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Case Report

Apoplectic presentation of a cauda equina paraganglioma

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Abstract

Background: Cauda equina paragangliomas (CEPs) are rare spinal tumors that are mostly misdiagnosed preoperatively as ependymomas or schwannomas on magnetic resonance imaging (MRI). Clinically, they usually present with the gradual onset of back pain and radiculopathy rather than an acute cauda equina syndrome.

Case Description: A 36-year-old female presented with an acute flaccid paraparesis/cauda equina syndrome. Based upon MRI studies, the predominant differential diagnoses included ependymoma or schwannoma. The intraoperative findings revealed an acute intralesional hemorrhage or apoplexy, responsible for the acute clinical deterioration. Histopathology and immunohistochemistry (IHC) revealed that the tumor was a paraganglioma.

Conclusion: CEPs commonly present with mild symptoms and signs rather than the acute-onset of a flaccid paraparesis/cauda equina syndrome as seen in this case. Here, the authors review the radiological and histopathological characteristics of CEP and emphasize the role of IHC in differentiating "CEP" from the more common ependymomas.

Key Words: Apoplexy in spinal tumors, cauda equina paraganglioma, intradural extramedullary spinal tumor, intralesional hemorrhage in spinal tumors, tumors of the filum terminale

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INTRODUCTION

Paragangliomas are rare, representing approximately 3% of cauda equina tumors.[1,6] Clinically and radiologically, they mimic schwannomas or ependymomas.[3,7] They often present insidiously, with back pain and radiculopathy, but rarely with the acute-onset of paraparesis or a cauda equine syndrome.[1] Here, a 36-year-old female presented with an acute flaccid paraparesis attributed to a hemorrhage within a paraganglioma cauda equina paragangliomas (CEP) involving the cauda equina.

CASE REPORT

Clinical and radiographic presentation

A 36-year-old female, presented with a 4-day history of the acute-onset of a flaccid paraplegia with urinary retention, accompanied by partial sensory loss below L1, and complete sensory loss below L3. On magnetic resonance imaging (MRI), the lesion was well-demarcated, extending from T12 to L2 [Figure 1a-g]. It was isointense on T1-weighted images with specks of hyperintensity, was hyperintense on T2-weighted images, and on MR with

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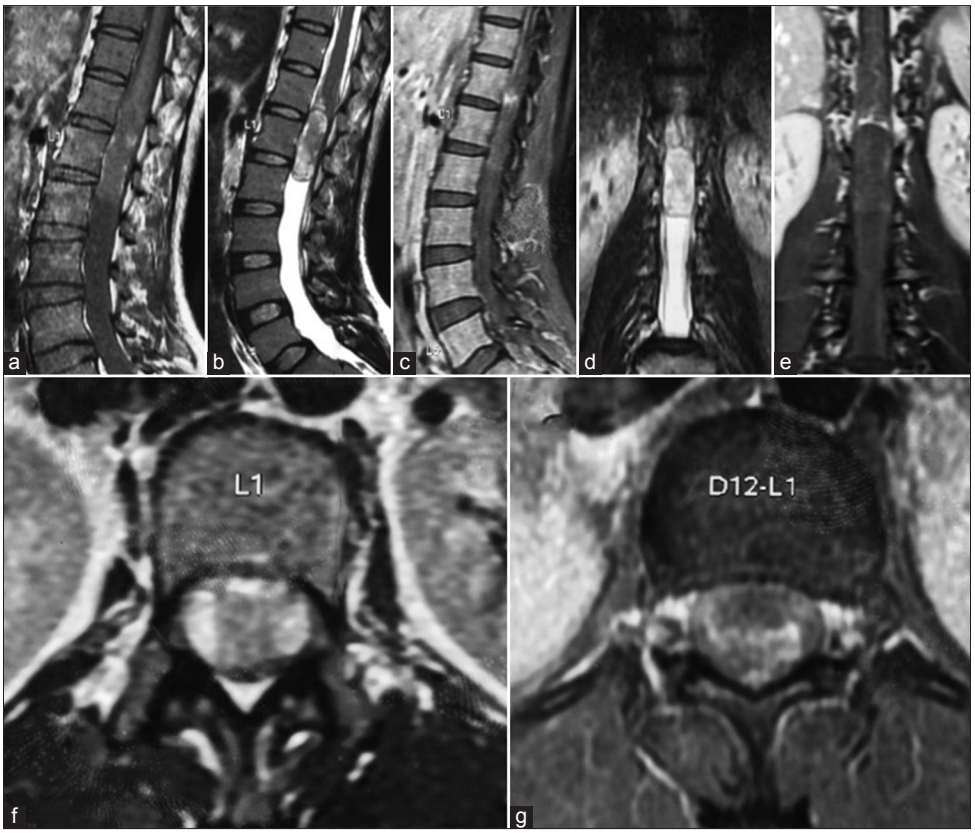

Figure 1: Magnetic resonance imaging showing (a) T1-weighted sagittal, (b) T2-weighted sagittal, (c) contrast-enhanced T1-weighted sagittal, (d) STIR sequence coronal images (hyperintense signal, suggestive of intralesional bleed), (e) contrast-enhanced T1-weighted coronal image (small rim of enhancement at the superior pole), (f) T2-weighted axial image, and (g) contrast-enhanced T1-weighted image at the superior pole (D12-L1 level showing rim-enhancement)

gadolinium-diethylene-triamine pentaacetic acid showed thin rim of peripheral enhancement at the superior pole. In addition, the short tau inversion recovery (STIR) MRI demonstrated uniform hyperintensity, suggestive of an intralesional bleed [Figure 1d].

Surgical intervention and follow-up

She emergently (within 24 h) underwent a T12-L2 laminectomy for tumor excision. Intraoperatively, the lesion was dark red in color, originated from the filum terminale, was ventral to the cauda equina, and had a well-defined, tense capsule due to the intralesional hemorrhage. The histopathology was consistent with a paraganglioma containing areas of fresh hemorrhage [Figure 2]. On immunohistochemistry (IHC), the intracytoplasmic granules showed uptake of chromogranin, also consistent with the diagnosis of a CEP. Nine months postoperatively, the patient exhibited a partial postoperative recovery; the motor examination showed 3/5 function (vs. the preoperative 0/5), with residual patchy sensory loss in the L5 and S1 dermatomes, and mild residual urinary retention (postvoid residual urine <100 ml) but without bowel dysfunction. The follow-up MRI showed no evidence of residual or recurrent tumor [Figure 3].

DISCUSSION

Origin and magnetic resonance findings of cauda equina paragangliomas

Paragangliomas are neuroendocrine tumors arising from specialized neural crest cells (chief cells) and are commonly encountered in the fifth and sixth decades of life.[1,6] They typically originate from the proximal filum terminale, rather than the cauda equina nerve roots.[1] They often present as solid, well-encapsulated, highly vascular, slow-growing tumors.[5] On MRI, they have no pathognomonic features and are frequently misdiagnosed as schwannomas or ependymomas. They are typically iso- to hypo-intense on T1-weighted studies, hyperintense on T2-weighted images, and exhibit intense contrast enhancement.[3] Serpentine, congested, ectatic vessels, and a low signal intensity rim ("cap sign" due to hemosiderin deposition) on T2-weighted images [Figure 1b] may be helpful in correctly establishing the diagnosis of CEP.[3,5,7] In this case, the tumor only showed contrast enhancement at the superior pole [Figure 1c and e], due to recent intratumoral hemorrhage.

Histopathology of cauda equina paragangliomas

On histopathological examination, CEPs are well-encapsulated, benign tumors (WHO Grade I),

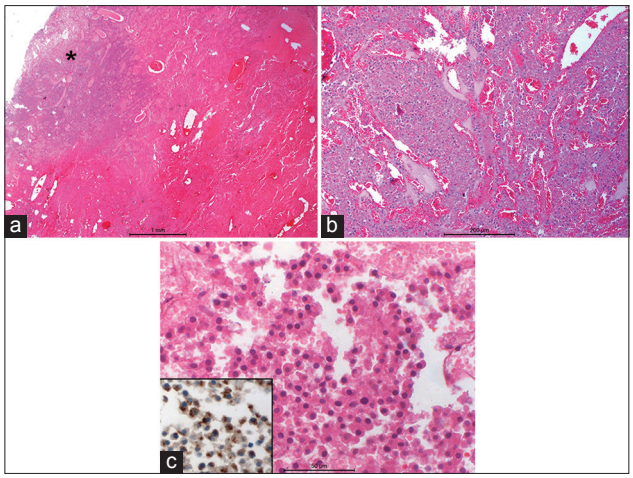

Figure 2: Photomicrographs of paraganglioma showing (a) well-circumscribed lesion (starred) with predominant hemorrhage. (b) The cellular component is arranged in sinusoidal pattern separated by thin-walled blood vessels. (c) Cells have a plasmacytoid appearance, with abundant cytoplasm. (c - inset) The intracytoplasmic granules express chromogranin (a: H and E, ×50; b: H and E, ×100; c: H and E, ×400; c-inset = immunoperoxidase, chromogranin)

characterized by the presence of "zellballen," or a nesting of cell groups, and trabecular cords of cells within thin fibrovascular stroma.[1] The predominant cell type is the "chief cell," which is round to polygonal in shape, and possesses central round to oval nuclei with finely stippled chromatin and inconspicuous nucleoli.

Immunohistochemistry of cauda equina paragangliomas

On IHC, the "chief cell" within the tumor is glial fibrillary acidic protein stain (GFAP) negative, while being neuron-specific enolase, synaptophysin, and chromogranin positive.[6] The second cell type is the "sustentacular or supporting cell," which is spindle-shaped and shows uptake for S-100 protein on IHC.[6] IHC is vital in differentiating paragangliomas from ependymomas (as ependymomas are GFAP positive and chromogranin negative on IHC). CEPs can occasionally be malignant and can rarely be functional. The outcome is excellent after complete excision of the capsulated tumor.[1]

SUMMARY

CEPs are solid, well-encapsulated, highly vascular benign lesions, which commonly present in an insidious fashion. Although CEPs have been reported to present with spinal subarachnoid hemorrhage, presentation with an acute flaccid paraparesis/cauda equina syndrome attributed to an intratumoral hemorrhage has not been previously documented.[2,4,5] Although often confused with schwannomas or ependymomas, due to the lack of pathognomonic MR findings, histopathological and IHC play a vital role in uniquely diagnosing these lesions. Emergency microsurgical excision of these tumors improves the outcome as observed in this case.

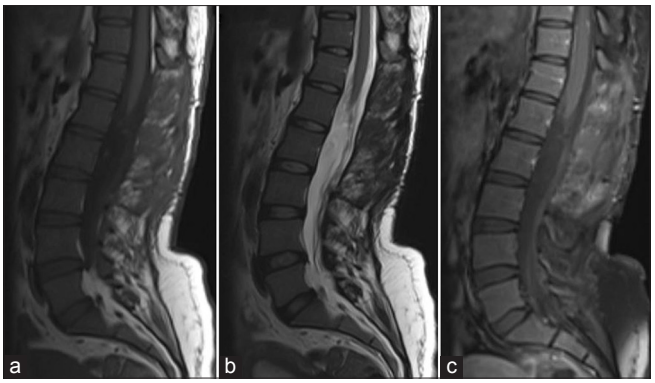

Figure 3: Magnetic resonance imaging lumbar spine at 6 months follow-up (a) T1-weighted sagittal, (b) T2-weighted sagittal, (c) contrast-enhanced T1-weighted sagittal images, showing no evidence of residual or recurrent tumor

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Conflicts of interest

There are no conflicts of interest.

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