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Acute onset of paraganglioma of filum terminale: A case report and surgical treatment

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ABSTRACT

INTRODUCTION: Paragangliomas of filum terminale are rare benign tumors, arising from the adrenal medulla or extra-adrenal paraganglia. These lesions usually present with chronic back pain and radiculopathy and only two cases of acute neurological deficit have been reported in literature.
PRESENTATION OF CASE: A case with an acute paraplegia and cauda equina syndrome due to an hemorrhagic paraganglioma of the filum terminale is described. Magnetic resonance imaging showed an intradural tumor extending from L1 to L2 compressing the cauda equina, with an intralesional and intradural bleed. An emergent laminectomy with total removal of the tumor was performed allowing a post-operative partial sensory recovery. Histopathological examination diagnosed paraganglioma.
DISCUSSION: Paragangliomas are solid, slow growing tumors arising from specialized neural crest cells, mostly occurring in the head and neck and rarely in cauda equina or filum terminale. MRI is gold standard radiological for diagnosis and follow-up of these lesions. They have no pathognomonic radiological and clinical features and are frequently misdiagnosed as other spinal lesions. No significant correlation was observed between the duration of symptoms and tumor dimension. Acute presentation is unusual and emergent surgical treatment is fondamental. The outcome is very good after complete excision and radiotherapical treatment is recommended after an incomplete resection. Conclusion: Early radiological assessment and timely surgery are mandatory to avoid progressive neurological deficits in case of acute clinical manifestation of paraganglioma of filum terminale.

1. Introduction

Paragangliomas of filum terminale are rare benign neuroendocrine tumors, arising from the adrenal medulla or extra-adrenal paraganglia, representing approximately 3% of cauda equina tumors [1]. The mean age of presentation is approximately 40–60 years with a slight male predominance [2]. Clinically and radiologically these lesions can be misdiagnosed as schwannomas or ependymomas and they often present insidiously with chronic back pain and radiculopathy. Reviewing the pertinent literature, only two cases of acute manifestation are described. We describe a case with an acute paraplegia and cauda equina syndrome because of an hemorrhagic paraganglioma of the filum terminale. We analyse the clinical, histopathological and radiological findings of these tumors, and discuss surgical treatment in line with the SCARE criteria [3].

2. Presentation of case

A 56-year-old man with a 10 years history of progressive low back pain and bilateral radicular leg pain without evidence of bowel or bladder incontinence or myelopathy, presented with a 4-day history of the acute-onset of a flaccid paraplegia with urinary retention, accompanied by complete sensory loss below L1. Emergent MRI (Magnetic Resonance Imaging) demonstrated a large intradural well encapsulated tumor extending from L1 to L2 compressing the cauda equine, measured about 4 cm in cranio-caudal diameter, heterogeneously enhancing after gadolinium injection with serpentine flow-voids in the subarachnoid space cranial to the tumor and with FrFSE (Fast Relaxation Fast Spin Echo) MR images showing uniform hyperintensity suggestive of an intralesional and intradural bleed (Figs. 1 and 2a–c). An emergent D12-L3 laminectomy was performed. Intraoperatively, prior to dural opening, the dural sac was noted to be dark blue in color and tense, because of underlying hematoma. The dura was initially opened above the tumor to

Abbreviations: MRI, Magnetic Resonance Imaging; FrFSE, Fast Relaxation Fast Spin Echo; WHO, World Health Organization.

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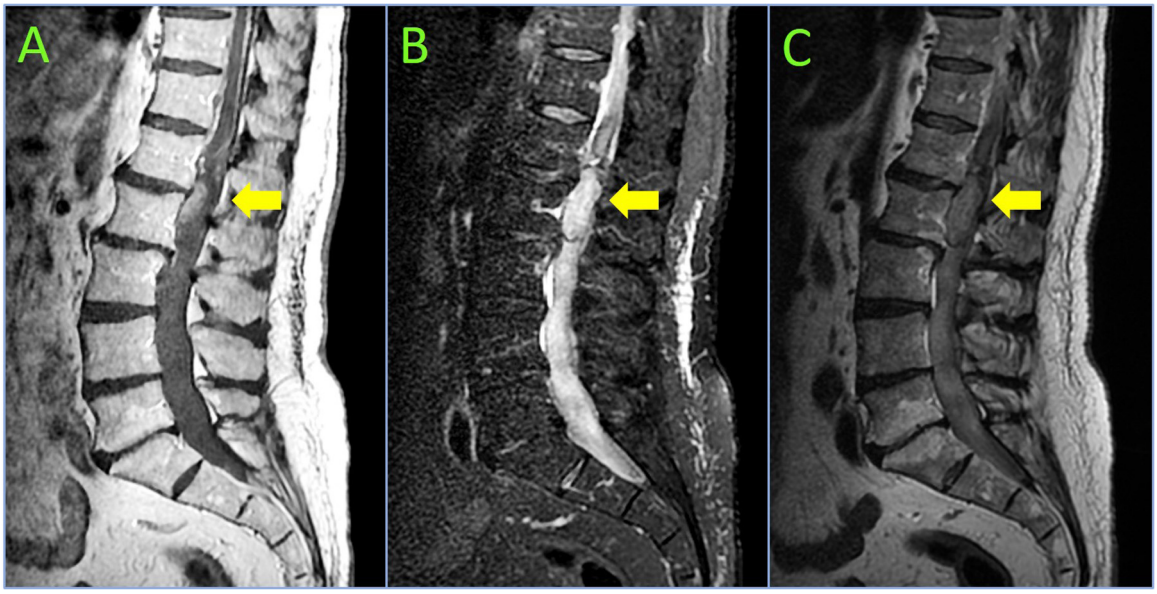

Fig 1. Sagittal (B) T1-weighted (A), T2-weighted FrFSE (B) and T2-weighted FrFSE fat sat (C) magnetic resonance images revealing a large omogeneously hyperintense intradural lesion (yellow arrow) extending from L1 to L2, with enhancement after gadolinium and flow voids cranial to the mass indicative of venous congestion or high vascularity of the tumor.

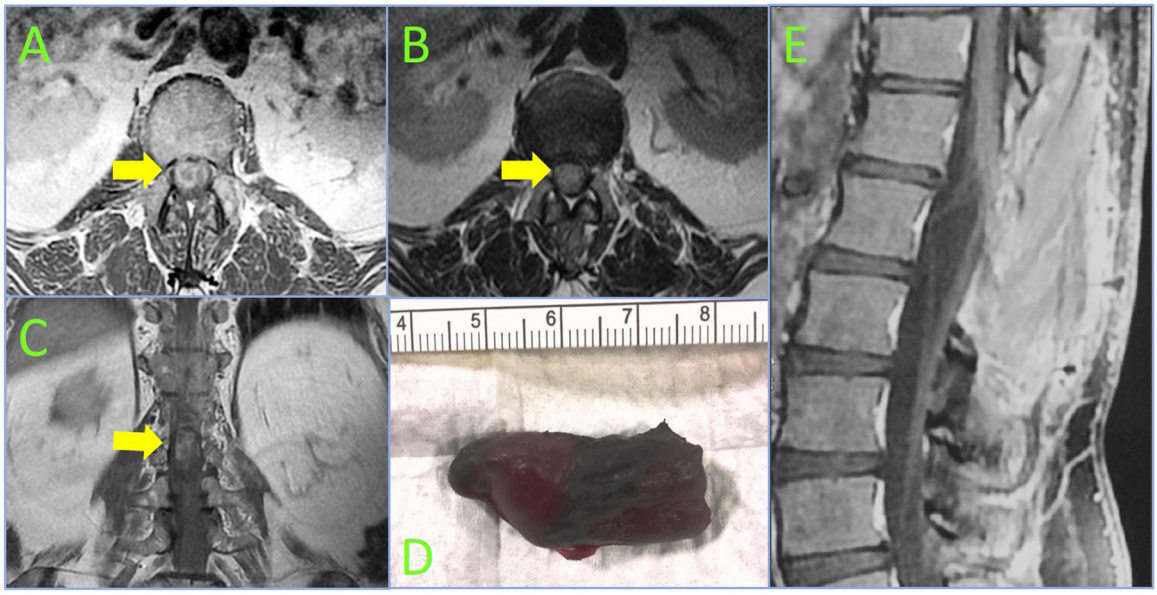

Fig. 2. Axial T1 (A) and T2-weighted (B) and coronal T1-weighted (C) images showing a large lesion (yellow arrow) taking up the vast majority of the cross-sectional area at L1-L2 level, obscuring the visibility of the cauda equina. Photograph of the hemorrhagic paraganglioma (4 × 2 cm) after excision en bloc (D). One year post-operative, sagittal T1-weighted image after gadolinim (E) demonstring no residual or recurrent contrast enhancing tumor.

prevent downward herniation of the mass, observing the egress of bloody cerebrospinal fluid under pressure and a large, well-encapsulated, dark red in color, mass pushing the dorsal nerve roots out of the dural opening. Caudally extending the midline durotomy, a shape dissection plane, retracting the nerve roots, was carefully performed around the tumor, that originated from the filum terminale, identified by the large, tortuous vein running along its length. After coagulation and cutting of this vein, the tumor was freed from its superior attachment and then removed en bloc (Fig. 2d). Histopathological examination showed paraganglioma, positive for chromogranin, synaptophysin and S-100 protein. By two months from surgery, only partial sensory returned below L2, but the patient still has no perineal sensation and has not regained bowel or bladder function. At one year post-operative MRI showed no evidence of tumor recurrence (Fig. 2e).

3. Discussion

Paragangliomas are solid, well-encapsulated, highly vascular, slow-growing neuroendocrine tumors arising from specialized neural crest cells [4]. Paraganglionic cells and the neural crest have a common origin, and during embryogenesis, they migrate along the neural tube. Paragangliomass result from dysfunction of embryonic paraganglia cell migration or non-regression. They can be found in adrenal and extra-adrenal tissues, and the extra-adrenal paragangliomas can be divided into sympathetic and