Thoracic exophytic ependymoma masquerading as a benign extra-axial tumor

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ABSTRACT

Spinal tumors are conventionally differentiated based on location in relation to the spinal cord. Benign spinal tumors such as schwannomas and meningiomas are typically extra-axial (intradural extramedullary) lesions, whereas more aggressive primary spinal tumors such as ependymomas are typically intramedullary masses. Rarely, ependymomas can have both intramedullary and extramedullary components (typically referred to as exophytic ependymomas). We report a case of a spinal exophytic ependymoma that radiographically masqueraded as a benign intradural extramedullary lesion causing cord compression and neurologic deficit in a 47-year-old man. The diagnosis of exophytic ependymoma was made intra-operatively, with resultant gross total resection of the extramedullary portion and subtotal resection of the intramedullary portion. Histopathological examination confirmed ependymoma with World Health Organization grade II/IV. Pre-operative suspicion of an exophytic ependymoma influences operative planning and clinical management. We review the literature and discuss clinical management strategies for these interesting spinal tumors.

1. Introduction

Intradural spinal cord tumors represent histopathologically distinct neoplasms that differ in cellular origin and pathophysiology, which impact prognosis and clinical treatment [1–3]. However, diagnosis based on clinical presentation is often difficult, as symptoms (back pain and progressive myelopathy) are nonspecific and slow clinical progression is the norm [4]. As such, MRI plays an important role in distinguishing spinal cord tumors preoperatively. Intradural tumors are classified based on their location in relation to the spinal cord. Among adults, benign meningiomas or nerve sheath tumors are the most common intradural, extramedullary (IDEM) masses, while the most common intramedullary lesions are the more aggressive ependymomas and astrocytomas. Ependymomas can rarely present as an IDEM tumor; even more rare are exophytic ependymomas with both intramedullary and extramedullary components [5–10]. We report an unusual case of a 47-year-old man with a thoracic exophytic ependymoma that on imaging appeared most consistent with meningioma or schwannoma.

2. Case report

A 47-year-old man presented to our institution with a 9-month history of progressive bilateral leg paresthesia and weakness, gait difficulty, and back pain. Neurological exam revealed bilateral lower extremity weakness with strength of 3–4/5 on the left and 4+/5 on the right, and hyperreflexia with up-going toes. Sensation was diminished starting from the T4 level. Findings were consistent with thoracic myelopathy. MRI revealed an enhancing IDEM mass at the T2 vertebrae level with spinal cord compression (Fig. 1). The slow progression of symptoms and the imaging findings were deemed most consistent with schwannoma or meningioma. A posterior T1 to T3 laminectomy was performed for resection of the intradural lesion (Fig. 2). Following midline durotomy, a well-encapsulated, avascular mass was immediately visualized in the extramedullary space, with the spinal cord compressed to the right and anteriorly, and cerebrospinal fluid under pressure superior and inferior to the mass (Fig. 2A). The majority of the extramedullary lesion was minimally adherent to, and easily separated from, the underlying cord and nerve roots. No exiting nerve root was identified in the mass, which would have been expected for a nerve sheath tumor. Interestingly, the superior portion of the mass was found to be arising from the spinal cord with no clear plane of dissection between the tumor and the cord (Fig. 2B), concerning for an exophytic lesion arising intramedullarly. The exophytic component and a lobule of the intramedullary tumor were resected (Fig. 2C, D). Gross total resection of the extramedullary component and subtotal resection of the intramedullary component were achieved. Somatosensory and motor-evoked potentials, monitored throughout the procedure, remained unchanged. Histopathological examination revealed the diagnosis of World Health Organization grade II/IV ependymoma (Fig. 2E, F). The patient regained full strength in the lower extremities bilaterally by postoperative day four. Lower extremity paresthesia remained unchanged. Postoperative MRI revealed re-expansion of the spinal cord, with no radiographic evidence of residual lesion (Fig. 3). Follow-up imaging at 6 months after surgery showed no residual tumor.

3. Discussion

This patient had an exophytic thoracic spinal ependymoma diagnosed pre-operatively as an IDEM mass based on radiographic appearance. Pre-operative recognition of an exophytic versus IDEM spinal lesion would have aided treatment and operative planning. First-line treatment for spinal ependymomas is gross total resection [3]; however, in exophytic ependymomas, the absence of a clear plane of dissection may limit the extent of resection, while the removal of the intramedullary portion involves a myelotomy,

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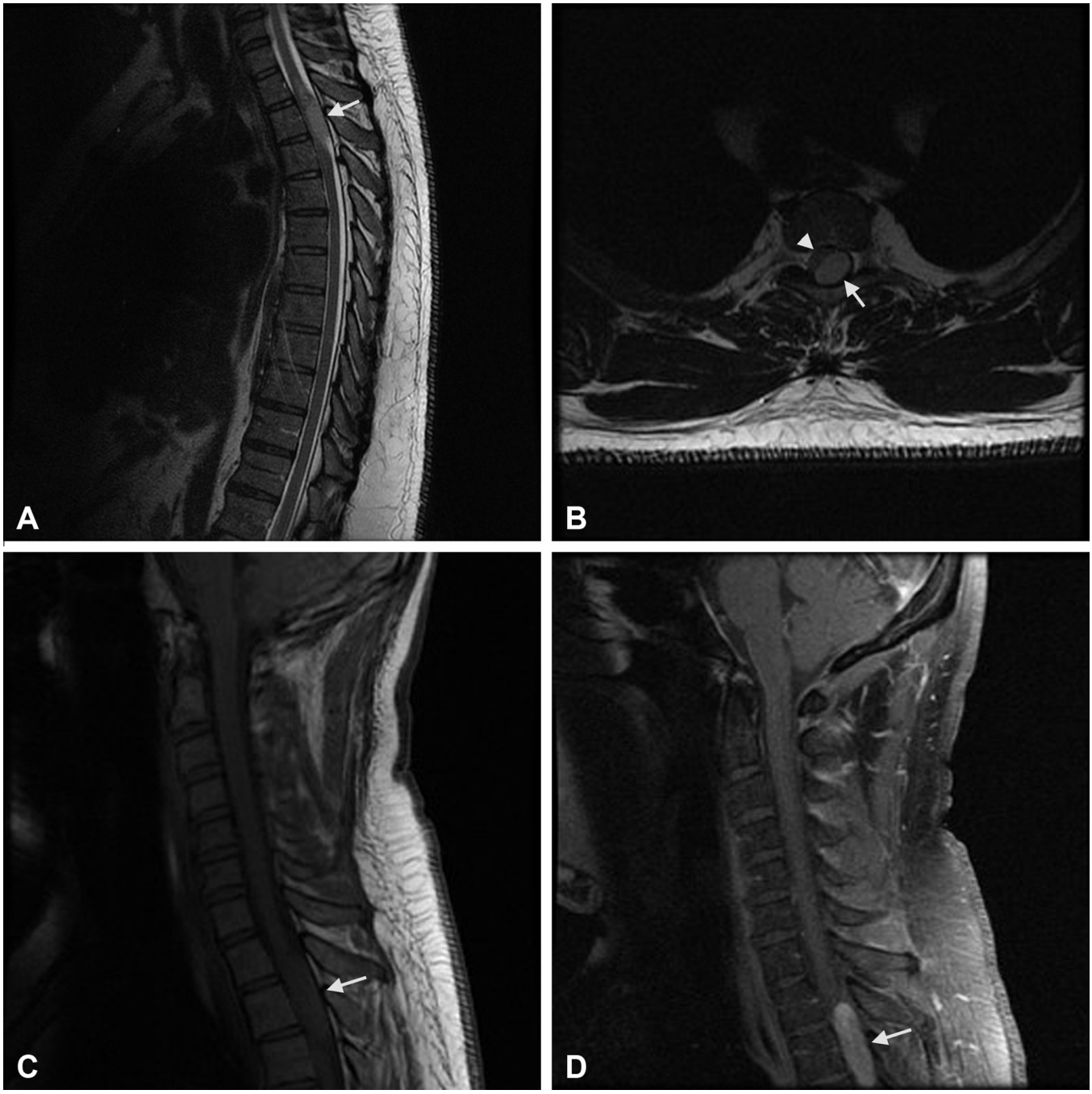

Fig. 1. MRI of the thoracic and cervical spine at initial presentation. (A) Sagittal and (B) axial (at the T2 level) T2-weighted MRI of the thoracic spine, and sagittal T1-weighted imaging of the cervical spine (C) prior to and (D) following gadolinium injection are shown. A 3.6 × 1 × 1.2 cm T1-isointense, T2-hyperintense, heterogeneously enhancing intradural extramedullary mass (arrow) at the T2 level is demonstrated, compressing the spinal cord (arrowhead) anterolaterally and to the right. Adjacent T2 hyperintensity within the cord is suggestive of edema.

risking motor and sensory deficits. Permanent surgical morbidities from resection was shown to be highest for ependymoma compared to other intramedullary lesions, postulated to result from more involved dissection of spinal cord vasculature [11,12]. As such, suspicion for an exophytic ependymoma would necessitate intra-operative monitoring of somatosensory-evoked and motor-evoked potentials as well as specific surgical strategies to minimize morbidities [12]. For our patient, subtotal resection of the lesion was performed to avoid causing permanent neurologic impairment, and intra-operative neuromonitoring revealed no changes. The rapid return to full strength post-op suggested that our patient's motor weakness largely resulted from cord compression by the extramedullary portion of the lesion. According to current recommendations for the treatment of intramedullary ependymomas [3], adjuvant radiation therapy will be considered.

Literature review revealed 25 reported patients with IDEM ependymomas since 1951, eight of which had exophytic lesions, but none masquerading as a meningioma or schwannoma as in our patient. Radiographically, intramedullary ependymomas cause focal cord enlargement and appear as an enhancing mass that is hypo- or isointense on T1-weighted images and hyperintense on T2-weighted images [2]. With the exception of the focal cord