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The Spine Journal 14 (2014) e9–e12

THE SPINE JOURNAL

Case Report

Interesting case of subependymoma of the spinal cord

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Received 30 June 2012; revised 20 September 2013; accepted 31 October 2013

Abstract

BACKGROUND CONTEXT: Subependymomas are rare, slow-growing, and usually noninvasive/nonaggressive World Health Organization Grade I tumors that tend to occur in the ventricles. Their most common site of occurrence is the fourth ventricle followed by the lateral ventricles. Spinal cord subependymomas typically manifest as cervical and cervicothoracic intramedullary or, rarely, extramedullary mass lesions. They often present clinically with pain and neurologic symptoms, including motor, sensory, urinary, and sexual dysfunction. Histologically, there are hypocellular areas with occasional clusters of cells and frequent microcystic changes, calcifications, and hemorrhage. Radiologically, subependymomas generally manifest as eccentric well circumscribed nodular lesions with mild-to-moderate enhancement.

PURPOSE: To highlight an interesting and rare presentation for subependymoma of the spinal cord.

STUDY DESIGN: This is a case report of a single patient in whom a subependymoma was resected from the cervical spinal cord with return to normal functioning.

METHODS: Clinical examination, magetic resonance imaging evaluation, surgical resection, and histological analysis were performed for diagnosis and treatment of this patient.

RESULTS: The patient experiencing myelopathy symptoms underwent a surgical resection of cervical spinal cord subependymoma that resulted in return to normal function.

CONCLUSIONS: Subependymoma should be included in the differential diagnosis of atypical presentations for myelopathy, as discrete surgical resection can result in good outcome.

Keywords: Subependymoma; Ependymoma; Glioma; Non-aggressive; Cervical; Spinal cord

Introduction

Subependymomas are rare, slow-growing, and usually noninvasive/nonaggressive World Health Organization Grade I tumors that tend to occur in the ventricles [1–4]. Their most common site of occurrence is the fourth ventricle followed by the lateral ventricles [5–7]. Involvement of the third ventricle and along the septum pellucidum is less common. Spinal cord subependymomas typically manifest as cervical and cervicothoracic intramedullary or, rarely, extramedullary mass lesions [5–7]. They often present clinically with pain and neurologic symptoms, including motor, sensory, urinary, and sexual dysfunction [5–7]. Histologically, there are hypocellular areas with occasional clusters of cells and frequent microcystic changes, calcifications, and hemorrhage [5–7]. Radiologically, subependymomas generally manifest as eccentric well circumscribed nodular lesions with mild-to-moderate enhancement [5–7].

Case report

History

A 29-year-old male presented in 2002 with spasms in his left leg after watching a college football game. In 2004,

FDA device/drug status: Not applicable.

Author disclosures: ***LMC:*** Nothing to disclose. ***CRH:*** Nothing to disclose. ***AMB:*** Nothing to disclose. ***LLS:*** Nothing to disclose. ***ACH:*** Nothing to disclose.

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1529-9430/$ - see front matter
http://dx.doi.org/10.1016/j.spinee.2013.10.056

while in the Peace Corps in Africa, he developed spasms in both legs and severe pain in his left hand after riding his bike for 50 km. The patient was transferred to Washington, D.C. where he was seen by a neurologist. A cervical magnetic resonance imaging (MRI) revealed an enhancing tumor in the spinal cord with associated myelomalacia. The patient was recommended to consult a neurosurgeon, but he opted not to initially. The patient noted progressive pain and clumsiness in his legs, especially with activity, and a periodic discomfort in his left arm. Consequently, he was referred to the neurosurgery department at our institution where a repeat MRI showed that the spinal cord lesion had grown.

Examination

On physical examination, signs of left cervical myeloradiculopathy were found with clumsiness of his right leg and occasional numbness in his feet. The MRI performed at our institution demonstrated an isointense T1 expansion of the cervical spinal cord extending from C5 to T2 levels with associated increased T2 signal intensity consistent for perilesional edema (Fig. 1A and B). A well circumscribed nodular focus of enhancement with hemorrhagic components was identified within the central spinal cord at C6–C7 (Fig. 1C, D, and E).

Operation

The patient underwent a C6–C7 laminectomy, with microscopic removal of the tumor guided by ultrasound localization. Intraoperatively, an edematous cord was found with marked hypervascularity to the left of the midline. Abnormal tissue was identified under high power magnification within a millimeter or so of the cord surface. The tissue was in some parts gray and soft and in other parts more pale beige/yellow and soft and bubbly. While pulling on the tumor, it was noted that it had a firm consistency and that any sort of retraction would lead to motor changes in the left hand. Complete removal of the lesion was successfully performed/obtained, with resultant marked decompression of the spinal cord. The patient tolerated the procedure well and no postoperative complications were encountered. In addition, the patient continued to be symptom free with the last contact made approximately 2 years after the surgical procedure.

Histological findings

Small clusters of cells surrounded by a dense fibrillary and microcystic matrix were found on pathologic examination of the submitted surgical specimen (Fig. 2, Top Left and Top Right). Glial fibrillary acidic protein positivity

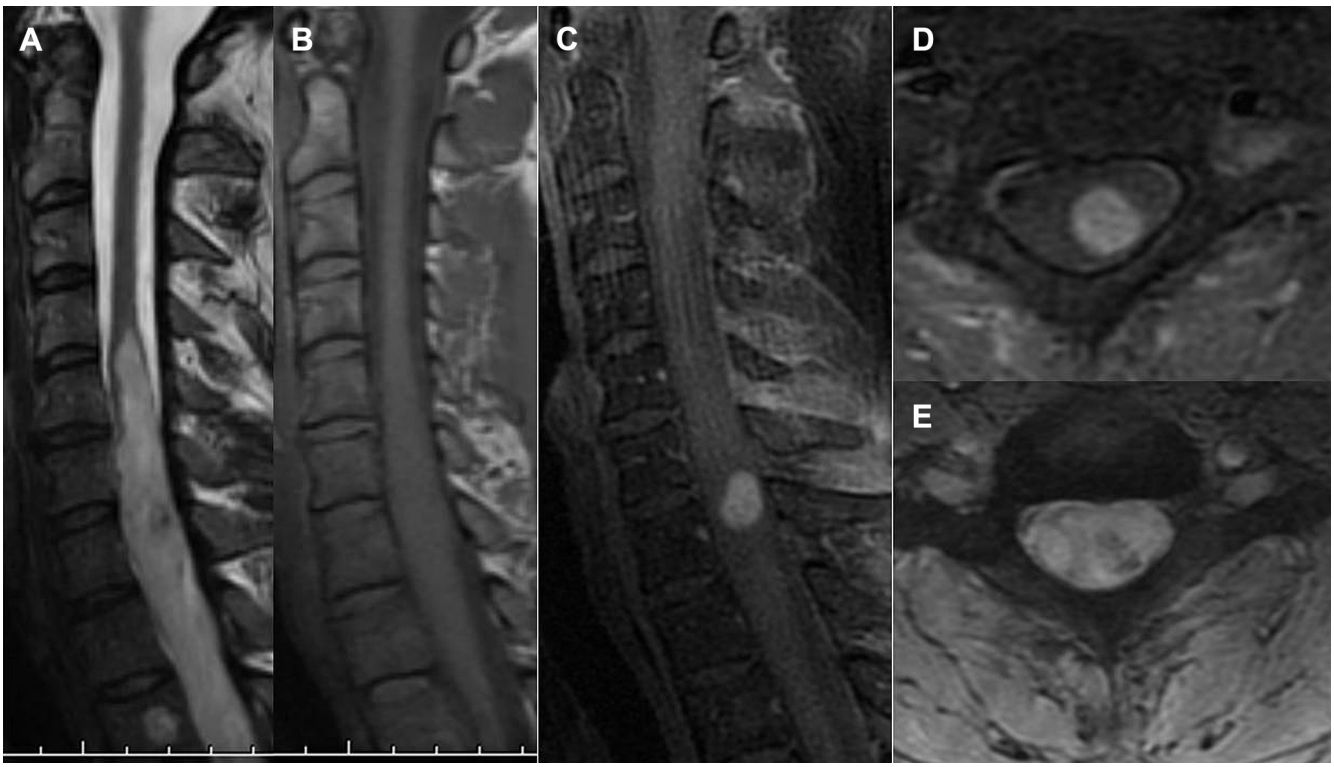

Fig. 1. (A) Sagittal T2-weighted magnetic resonance imaging (MRI) demonstrates cord expansion with increased signal intensity from C4 through the visualized T2 level. A T2 nodular hypointense focus is identified at C6–C7. (B) Sagittal T1-weighted MRI demonstrates isointense fusiform expansion of the cervical spinal cord from C5 through T1 levels. (C) T1 fat suppression postgadolinium sagittal and (D) axial MRIs demonstrate a well circumscribed eccentric nodular focus of enhancement at the C6–C7 level. (E) Gradient echo axial sequence exhibits susceptibility artifact consistent for a small hemorrhagic component.