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Spinal accessory nerve meningioma in a paediatric patient: Case report

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Abstract

Accessory nerve meningiomas are exceedingly rare. We present a case of a nine-year-old patient with neurofibromatosis type 2 who had radiologic evidence of spinal cord compression from an upper cervical/foramen magnum lesion. He was asymptomatic from this lesion, but it progressed in size. The tumor was resected and histologic investigation revealed frequent tight whorls and psammoma bodies consistent with meningioma. To the authors' knowledge, this is the first reported spinal accessory nerve meningioma in a pediatric patient.

Key words: *foramen magnum, nerve sheath neoplasm, children, spine*

Introduction

Schwannomas and meningiomas comprise the majority of intradural, extramedullary tumors arising at the craniocervical junction [7]. Schwannomas originate from Schwann cells of cranial and peripheral nerves, while meningiomas typically arise from meningothelial cells [2]. Optic nerve sheath meningiomas are well described, but meningiomas originating from other cranial nerves have been sparsely reported. These tumors are hypothesized to arise from arachnoid mater within the cranial nerve sheaths [6]. To our knowledge, only two patients with spinal accessory nerve meningiomas have been described in the literature. In this unique case, we discuss the characteristics of a spinal accessory nerve meningioma that was removed in a pediatric patient.

Case Report

We report a nine-year-old white male with a history of neurofibromatosis type II. Blindness in the right eye had resulted from an optic nerve meningioma. In addition to the child's bilateral eighth nerve tumors, a left Meckel's cave trigeminal nerve schwannoma, left anterior clinoid meningioma, and a dorsal T4-T5 meningioma had been identified. Examination revealed right eye blindness, nystagmus on the left with full extraocular eye movements. His hearing was

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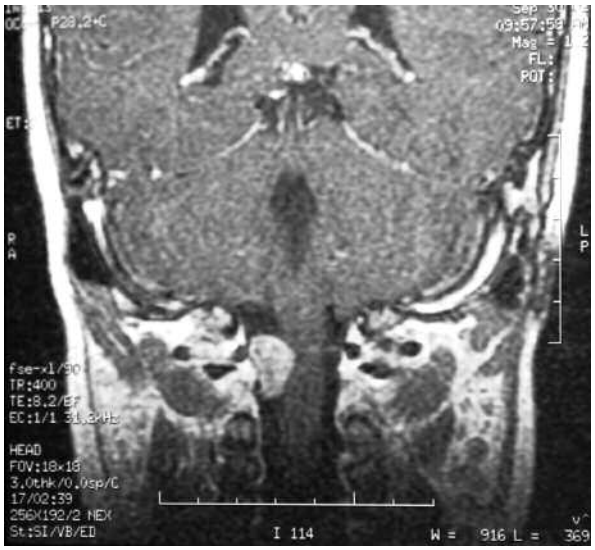

Fig. 1. Preoperative coronal T1-weighted MRI with contrast demonstrates a right-sided homogeneously enhancing upper cervical/foramen magnum mass with spinal cord compression

intact to finger rub bilaterally. His face was symmetric with intact sensation. He had good motor strength and coordination. He could perform a tandem gait normally. His reflexes were normal and symmetric. MRI revealed no change in his previously documented tumors except for an enlarging C1/foramen magnum lesion causing cord compression. There was no shoulder pain. This lesion enhanced vividly and measured 1.1 x 1.2 x 1.5 cm (Fig. 1).

At surgery, the patient was placed in the prone position and a C1 laminectomy was performed via a midline incision and the dura mater was opened. The tumor was visualized on the right indenting and shifting the cord to the left. Using microsurgical techniques, the perimeter of the tumor was dissected free from the surrounding tissue and found to arise from the vertical spinal portion of the spinal accessory nerve posterior to the denticulate ligaments. The tumor was debulked internally and its capsule was manipulated allowing it to be removed in its entirety. No injury to surrounding vessels, regional spinal nerves, or the spinal portion of the spinal accessory nerve was noted. The dura mater was closed primarily followed by routine soft tissue/skin closure.

Histologically, proliferation of sheets of epithelioid tumor cells with vesicular nuclei and eosinophilic cytoplasm with indistinct cell borders were enmeshed in dense connective tissue. The neoplasm was nodular

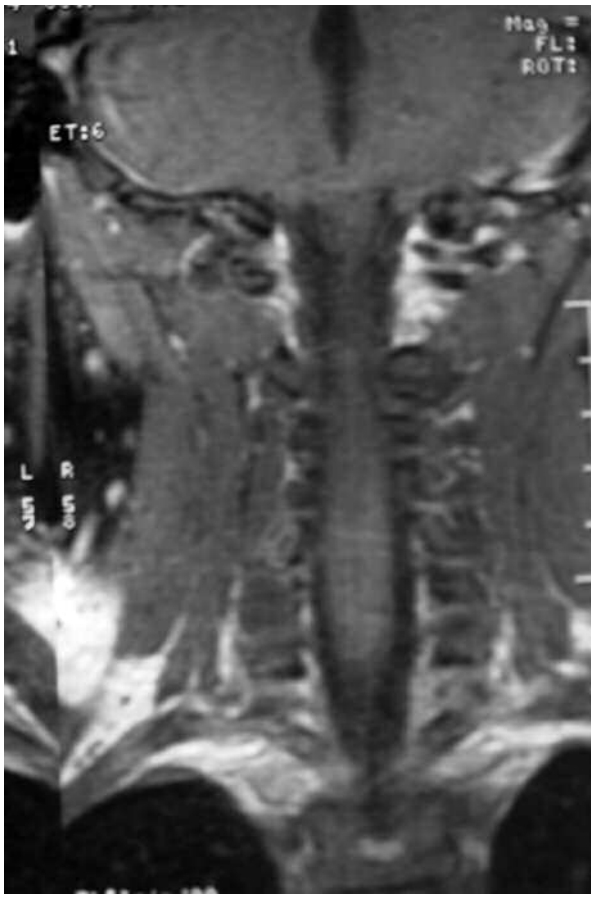

Fig. 2. Postoperative coronal T1-weighted MRI with contrast reveals complete resection of the tumor seen in Figure 1

in areas and contained frequent tight whorls and psammoma bodies. No mitotic figures were noted. Focal areas of fibrosis and mild inflammation were noted. The final diagnosis of this lesion was meningioma.

The patient's uneventful postoperative course was followed by discharge and follow-up MRI three months later (Fig. 2). Imaging revealed complete tumor resection with no evidence of recurrence. Function of this patient's spinal accessory nerve was intact. At three years follow-up the patient remains well with no symptoms of spinal accessory nerve dysfunction or recurrence of tumor of this nerve on repeat imaging.

Discussion

Excluding optic nerve sheath meningiomas [1,5], there is a paucity in the literature regarding meningio-

mas of other cranial nerves. Optic nerve sheath meningiomas are thought to account for 1-2% of all meningiomas, although they are quite rare in children [5]. Hart et al. [4] presented a unique case of a malignant meningioma of the oculomotor nerve. These authors also noted the rarity of meningiomas without dural attachment [5]. Chung et al. [3] have reported a case of a geniculate ganglion meningioma. We identified only two other cases reported in the literature of spinal accessory nerve meningiomas and both were in adults [6,7]. Only one of these reports was symptomatic due to spinal accessory nerve involvement and both involved only the intracranial segment of this nerve.

Extracranial meningiomas are hypothesized to occur by a variety of mechanisms. Some have posited that they originate from an intracranial location and then spread [6]. Some have theorized that these meningiomas may arise from arachnoid within cranial nerve sheaths [3,6]. Additionally, extracranial meningiomas could possibly arise from ectopic arachnoid cell rests [3,6]. Mesenchymal and blastomatous transformation has also been noted to be possible etiologies [3].

To our knowledge, this case illustration represents the only reported pediatric spinal accessory nerve meningioma. This unique lesion should be considered in the differential diagnosis of tumors arising from cranial nerve XI. With the potential for meningiomas to recur, proper diagnosis is essential to insure adequate long-term follow-up.

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