Meningioma with dural venous sinus invasion and jugular vein extension

Case report

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Meningiomas represent the most common benign intracranial neoplasm in adults, with a considerably lower incidence in children. The authors present the case of an intracranial meningioma with invasion of, and intraluminal extension into, the transverse and sigmoid sinuses, jugular bulb, and internal jugular vein, resulting in venous occlusion in a 14-year-old girl. Computed tomography scanning, MR imaging, and conventional angiography were performed preoperatively. The patient underwent a 2-stage resection: the supratentorial component was resected first, and the infratentorial and venous sinus and jugular vein components were subsequently removed using a combined skull base approach. Gross-total resection was achieved by opening the lateral dural sinus and removing the meningioma from within the transverse and sigmoid sinuses, the jugular bulb, and the internal jugular vein. The patient remained neurologically intact after the staged tumor resections. Postoperative imaging confirmed the gross-total resection. This case illustrates the unusual property of an intracranial meningioma to invade the intrasinusoidal space and extend into the jugular vein without adherence to the underlying venous endothelium of the jugular vein.

KEY WORDS • dural venous sinus invasion • jugular vein • meningioma • skull base

Meningiomas represent ≤ 3% of the intracranial tumors in the pediatric population and < 2% of meningiomas in all age groups. A report from our institution by Drake et al. has revealed only 13 cases of meningioma among the 1283 intracranial tumors in the children encountered over 51 years, which constitutes an incidence of 1%. A literature review by Mendiratta et al. has indicated an incidence of 1.5% among 2620 cases of intracranial tumors in a pediatric population. Worldwide reports have indicated an incidence ranging from 1.3 to 2.4%. Meningiomas in children also tend to occur in the posterior fossa with an incidence ranging from 19 to 46%, whereas only 10% of these lesions in adults appear infratentorially. A primary intracerebral location without an obvious dural attachment has also been reported in cases of meningiomas in children. Finally, meningiomas in children are more commonly cystic in nature compared with those in adults. All of these features unique to the pediatric age group make the preoperative diagnosis of meningiomas in children more difficult than in adults even with the advent of advanced MR imaging sequences.

Here we describe the diagnosis and treatment of a teenager harboring a meningioma that originated from the dura mater at the junction of the transverse and sigmoid sinuses and that had both intracranial supratentorial growth and extension into the lateral dural venous sinuses to the level of the mid–jugular vein.

Abbreviation used in this paper: GTR = gross-total resection.

This article contains some figures that are displayed in color online but in black and white in the print edition.
Case Report

History and Examination. This 14-year-old girl presented with a 6-month history of headache, dizziness, and neck pain; previously, she had been well. She reported a persistent machine-like noise in her right ear. She also described a noxious burning smell manifesting itself 3–4 times per day over the month prior to her presentation. There was no associated clinical seizure activity. The results of general physical and neurological examinations were normal.

Computed tomography and MR imaging of the brain revealed a right-sided, noncalcified, extraaxial dural-based mass measuring 3.3 × 4.2 × 2.7 cm (Fig. 1). The lesion was based supratentorially but traversed the tentorium and filled the distal transverse sinus, sigmoid sinus, and jugular bulb, extending through the jugular foramen into the internal jugular vein. Distally, venous occlusion was observed to the level of C-2. Mild mass effect was noted in the right posterior temporal lobe. A chest radiograph was performed and did not reveal any pulmonary lesions.

Cerebral angiography demonstrated a tumor blush in the right posterior temporal region, with the ipsilateral occipital, middle meningeal, and ascending pharyngeal arteries identified as feeding vessels. The distal half of the right transverse sinus and the sigmoid sinus and internal jugular vein were occluded. A small arteriovenous fistula between the right occipital artery and transverse sinus was also identified (Fig. 2). The differential diagnosis included meningioma, or an extraaxial neurogenic tumor such as Ewing sarcoma or paraganglioma. Intracranial mass effect and intrasinusoidal and intravenous extension of the tumor to C-2 accounted for the patient’s headache and neck pain. The associated arteriovenous fistula may have explained the machine-like noise she heard in her right ear.

First Operation. A staged approach to the tumor was undertaken. Resection of the supratentorial component was performed first via a posterior temporal craniotomy with the aid of image guidance. The tumor was debulked superoinferiorly, leaving a small cuff attached to the adjacent transverse-sigmoid sinus junction. Pathological analysis revealed a WHO Grade I meningioma with rare mitoses and a low MIB-1 proliferative index (Fig. 3).

First Postoperative Course. The patient was discharged home without neurological deficit on the 3rd postoperative day. In the follow-up period, she remained well neurologically, and neuroimaging studies revealed limited regrowth of residual tumor.

Second Operation. Six months after the first procedure, a posterior temporal craniotomy was again performed, this time in conjunction with an extended posterior fossa craniotomy and a retrolabyrinthine approach to the residual tumor. A lumbar CSF drain was used at the outset. The facial nerve was identified and preserved in its osseous canal, and the horizontal segment of the carotid artery was exposed in the carotid artery canal of the petrous bone. A high anterior cervical dissection was performed to identify the enlarged and occluded internal jugular vein. The vein was opened and the tumor was dissected free from the endothelium of the vein to which it was not attached and was removed en bloc up to the jugular bulb (Fig. 4). The jugular vein distal to the tumor was suture ligated, as were the common facial vein and other venous tributaries that were unblocked once the tumor had been removed in this location.

Our attention was then turned toward the tumor at the transverse-sigmoid sinus junction. Here, the lateral dural sinuses were opened, and tumor was removed from within the sigmoid sinus and down to the jugular bulb by using a Cavitation ultrasonic surgical aspirator. No attempt was made to reconstruct the already occluded lateral venous sinuses once the tumor had been removed. Piecemeal removal of tumor within the jugular bulb was then performed down to the jugular vein. Bleeding from within the jugular

Fig. 1. Axial (left) and coronal (right) T1-weighted MR images with Gd demonstrating a homogeneously enhancing extraaxial, dural-based mass with transtentorial, intravenous, and transcranial extension into the retrostyloid region of the infratemporal fossa.
bulb, emanating from the inferior petrosal sinus and the occipital sinus, was controlled with Gelfoam, which was used to occlude the jugular bulb. Tumor was then removed from within the transverse sinus from the junction of the sigmoid and transverse sinuses proximally to a distance approximately one-third the length of the transverse sinus after it had been unroofed. Gelfoam was used here to pack the transverse sinus distal to a large-caliber draining cerebral vein, which drained retrograde to the confluence of sinuses. Following GTR of the tumor, the opened and occluded jugular vein was closed primarily.

Second Postoperative Course. Postoperatively, the patient remained neurologically intact. A postoperative imaging study has confirmed GTR of the tumor (Fig. 5). Mild CSF otorrhea ceased within 5 days of lumbar CSF drainage. The patient was discharged home in good condition on the 10th postoperative day, and she remained neurologically intact 12 months after discharge. Pathological analysis of the intrasinusoidal tumor revealed a low-grade meningioma very similar in features to those in the lesion from the first procedure (data not shown).

Discussion

We described an adolescent with an intracranial meningioma that originated from the dura of the lateral venous sinuses. The tumor transgressed the dura and grew intrasinusooidally to the level of the upper cervical jugular vein. Interestingly, the tumor grew down from its origin within the confines of the sigmoid sinus, jugular bulb, and jugular vein without invasion of the venous endothelium of the jugular vein. A 2-stage approach to the lesion was undertaken, and the patient has fared well without neurological deficit.

As discussed, the most striking differences between meningiomas in adults and those in children are the radiological and pathological characteristics. The radiological differential diagnosis is extremely broad and includes primary CNS tumors and secondary lesions from osseous and hematological neoplasms. Assigning the pathology grade is difficult in cases of lesions in pediatric patients, and the presence of sclerosing and sarcomatous variants poses problems in the histological diagnosis. Note that MIB-1 staining indices correlate with tumor grade and signify tumor recurrence. In a direct comparison of lesions in adults and children, there was no difference in the MIB-1 levels between the 2 patient groups. Thus, predicting the postresection behavior of tumors in children is rendered more difficult. Close follow-up is recommended even in cases with a benign histology.

Through progressive growth over time, meningiomas not infrequently invade and ultimately occlude the dural venous sinuses, rendering them difficult to extirpate completely. In a retrospective study by Sindou and Hallacq, 18% (80 of 425) of all meningiomas involved major dural sinuses, and 5 of these lesions involved the transverse sinus and 3 the confluence of sinuses. Sindou has also devised a classification system for venous sinus involvement by these tumors, modified from the previously published schema by Bonnal et al. Three potential strategies for tumor resection have been espoused for meningiomas affecting the major dural sinuses. The first is removal of the meningioma and resection of the outer dural layer of the sinus while coagulating its inner layer at the site of dural attachment. The second involves resection of the invaded walls of the sinus together with excision of the intraluminal tumor fragment and subsequent primary venous repair, autologous patch grafting, or venous bypass. The third option involves complete resection of the invaded sinus without venous reconstruction. Overall, the com-
The complication rate in such cases has been as low as 3% but also has been quoted as high as 50%.\textsuperscript{5,30,31} Preservation of the cortical draining venous structures is arguably the most important step in the surgical removal of meningiomas of all types. Some authors have recommended reconstruction of the dural sinuses even in cases of complete venous occlusion as it allows for the development of collateral venous drainage with progressive occlusion of the primary venous repair.\textsuperscript{30}

Because the meningioma we encountered in the present case involved the jugular foramen, the preoperative differential diagnosis included lesions that typically arise from this region, including glomus tumors, schwannomas, or meningiomas. Meningiomas comprise \textasciitilde 10% of the tumors in this region, and patients with benign lesions present with cranial nerve deficits, whereas those with malignant lesions often present with ear and mastoid pain.\textsuperscript{24} A CSF cleft between the tumor, cranial nerves, and cerebellum is usually present, thus facilitating resection. Extracranial extension of jugular foramen meningiomas into the carotid sheath is common, with resultant encasement of the carotid artery and occlusion of the internal jugular vein.\textsuperscript{13} Primary jugular foramen meningiomas create a permeative, sclerotic change to the foramen with foraminal expansion. Intraosseous extension into the skull base is not uncommon.\textsuperscript{13,32} Secondary jugular foramen schwannomas extend to the foramen from other neighboring regions and are thought to be responsible for the higher recurrence rates related to incomplete resection.\textsuperscript{13,15,16} The lesion in the case we present is clearly different from either the primary or secondary jugular foramen meningiomas described in the literature as the tumor did not expand the jugular foramen, cause osseous alterations, or transgress the carotid sheath.
Meningioma with jugular vein extension

Prabhu and DeMonte have described a case of a glomus jugulare tumor that displayed remarkable venous extension similar to that observed in our case. They described a lesion that started at the confluence of sinuses and extended intraluminally to the junction of the internal jugular vein and innominate vein. In the cephalad direction, the lesion grew adjacent to the temporal lobe, mesotympanum, and middle ear. The authors also used a 2-stage approach for resection. The first stage involved tumor removal via a temporal bone resection, posterior fossa craniotomy, radical neck dissection, and an infratemporal fossa approach. The second stage consisted of a median sternotomy with ligation of the innominate vein and the distal internal jugular vein.

A 2-stage approach promotes maximal tumor resection with minimal neurological morbidity. Although identified in its osseous canal, the facial nerve did not need to be mobilized, hence completely preserving its function. The lower cranial nerves within the jugular foramen and jugular bulb areas also were not placed at high risk in our case as the meningioma extended within these venous structures, distending them to a certain degree but not invading the venous endothelium. It is the intraluminal extension of the meningioma here that we found unique and of which there are few if any cases reported in the literature. Such an unusual phenomenon may relate to the biology of meningioma in the pediatric versus the adult population. One can argue that it was unnecessary to preserve the walls of the jugular vein in our case as it was already occluded and its retention could conceivably diminish long-term tumor control; however, we noted a relative lack of involvement of the endothelium within the jugular vein by the meningioma. Furthermore, we were concerned that a circumferential dissection of the jugular vein could lead to a lower cranial nerve injury. In most cases of extensive skull base meningiomas, adult or pediatric, the risk of recurrence is directly related to the extent of resection. As with many skull base meningiomas, Simpson Grade 3 and 4 resections are performed almost 10 times more often than Simpson Grade 1 and 2 resections. In the present case, given our concerns regarding a future recurrence within the jugular bulb or foramen, fractionated radiotherapy or radiosurgery may be an option to achieve tumor control.

Conclusions

We presented the first reported case of a transcranial meningioma with extensive intraluminal involvement and consequent venous occlusion. A 2-stage GTR was performed without venous reconstruction, and the patient suffered minimal perioperative and neurological morbidity. We believe that such a management strategy provides the best opportunity for maximal reduction of the tumor burden and optimal neurological results.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References


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