Intracranial Juvenile Angiofibroma with Intradural and Cavernous Sinus Involvement

Abstract—Juvenile angiofibroma is a benign tumor, well known for aggressive extracranial growth, that frequently spreads to involve the skull base and intracranial compartment as well. The meninges, however, provide a strong barrier against intracerebral spread. This article reports an unusual case of angiofibroma with dural penetration, cavernous sinus involvement, and adherence to the temporal lobe of the brain. Controversy exists as to how the intracranial extensions are best managed. Our experience with this case, combined with review of the literature, suggests that these lesions can be removed surgically, eliminating in most cases the need for radiation therapy. (*Skull Base Surgery*, Volume 2, Number 2, 1992, p. 87)

Juvenile angiofibroma is an uncommon benign tumor affecting almost exclusively pubertal males. It takes origin at the base of the pterygoid plates in the body of the sphenoid, in the region of the sphenopalatine foramen. By direct spread, tumor can involve the nasopharynx, nasal cavity, pterygopalatine fossa, infratemporal fossa and intracranial space. Juvenile angiofibroma is characterized by extreme vascularity, local invasion, and bony destruction. Historically, this tumor has been very difficult to treat; multiple recurrences are the norm, with an attendant significant morbidity and high mortality rate. Improved radiologic imaging, such as high-resolution computed tomography (CT) and magnetic resonance imaging (MRI) and the use of approaches providing optimal tumor exposure have increased the surgical control rates for extracranial angiofibroma to better than 90%.¹ Good results using radiation as the primary modality have been obtained;³ however, most surgeons opt for surgical excision of extracranial lesions.^{1,2} Controversy remains concerning the management of juvenile angiofibroma with intracranial extension. The authors herein report an unusual case of juvenile angiofibroma with intradural invasion managed surgically.

CASE REPORT

A 13-year-old boy was referred to the University of California Davis Medical Center Center for Skull Base Surgery in December 1989 with a 6-month history of progressive nasal obstruction and three bouts of epistaxis, two mild and one moderate in severity. Clinical examination revealed a purplish mass in the left posterior nasal cavity and nasopharynx totally obstructing the left nasal airway, and bilateral middle ear effusions. A fine cut CT scan and an MRI scan with gadolinium enhancement demonstrated a large tumor involving the left nasopharynx, sphenoid sinus, pterygomaxillary and infratemporal fossae. Tumor extended into the left middle cranial fossa, with apparent invasion of temporal lobe and cavernous sinus with displacement of the ipsilateral carotid artery superomedially (Figs. 1–4).

Biopsy under anesthesia confirmed the diagnosis of angiofibroma. He was placed on 5 mg of diethylstilbestrol for approximately 3 weeks. This promotes enhanced fibrosis, reduced vascularity, and reduction in tumor size.⁴ Four hours prior to surgery, he underwent preoperative

Figure 1. Coronal magnetic resonance imaging (T₁ weighted image) without contrast demonstrating extensive tumor with middle cranial fossa and temporal lobe involvement and extension into cavernous sinus with displacement of left internal carotid artery.

Figure 3. Sagittal magnetic resonance imaging (T₁ weighted image) demonstrating encroachment of the tumor into temporal lobe.

Figure 2. Coronal magnetic resonance imaging (T₁ weighted image) with gadolinium demonstrating encroachment of the tumor into temporal lobe and extensive infratemporal fossa involvement.

Figure 4. Axial magnetic resonance imaging (T₁ weighted image) demonstrating extensive nasal, sphenoid, infratemporal, and middle cranial fossa involvement of the tumor.

tumor embolization. The internal maxillary artery, the major arterial feeder, was occluded with Gelfoam. Care was taken to embolize the vessel beyond the supply to the temporalis muscle. The tumor was resected via a combined transpalatal and Caldwell-Luc procedure and a lateral infratemporal/middle fossa approach. The intracranial portion of the tumor was found to have caused massive destruction of the floor of the middle cranial fossa and actually penetrated the dura and the pia mater of the temporal lobe (Fig. 5). A rim of temporal lobe had to be removed to ensure complete tumor extirpation. Medially, the tumor extended into the inferior cavernous sinus and displaced the carotid artery upward and medially. In addition, it invaded the sphenoid sinus. Because of bone involvement, part of the clivus and sphenoid sinus walls had to be drilled away. There was no histologic evidence for malignancy in the tumor specimen. Using this combined approach, the entire tumor was removed macroscopically, augmented with microscopic resection and use of the carbon dioxide laser. The dural defect through which the tumor extended was closed with a 2 cm and 3 cm temporalis fascia graft. The temporalis muscle was reflected across the floor of the middle fossa and fixed by its attached pericranial cuff to the clival periosteum. A microvascular rectus abdominis free flap was used to fill the defect in the infratemporal fossa and to avoid a cosmetic deformity. Blood loss was reduced in part by the use of the carbon dioxide laser to diminish tumor bulk. Intraoperative blood loss was 3600 ml. Blood replacement was greatly aided by the use of the "cell saver." Postoperatively, the patient had one grand mal seizure and a persistent left middle ear effusion requiring a ventilation tube. Otherwise, his course was uncomplicated.

Subsequently, he has done well with no ocular or neurologic deficit. The highly invasive character of this tumor required one further transpalatal excision of tumor that actually invaded the nasopharyngeal mucosa. One

other excision was accomplished by the sinus endoscope. There has been no evidence of intracranial recurrence.

DISCUSSION

Intracranial extension has been shown to occur in 20 to 30% of patients with juvenile angiofibroma.^{1,3} Several options have been suggested in the literature concerning the management of intracranial extensions of angiofibroma. One alternative is to regard extensive disease such as this as unresectable and to use radiation therapy as the primary modality of treatment.² A dose of 3000 to 3500 rads given over 3 to 3½ weeks is the usual treatment regimen.⁵ Cummings et al³ achieved good control rates with radiation therapy, but not all centers can match these excellent results. We believe that the possible long-term effects of radiation, including impaired facial growth, cerebral damage, and induction of malignancy, are too great to advocate its use in this benign tumor. Almost all the reported malignant transformations of these tumors have occurred in those patients who have been irradiated.¹⁰ Some authors advocate a subtotal resection of tumor, leaving behind tumor that involves the cavernous sinus.^{1,6} This is usually performed using an intracranial/extradural and extracranial approach. Other authors recommend a complete extradural removal from an extracranial approach;⁷ or tumor may be amputated at the skull base, leaving the intracranial portion. Total tumor removal can be attempted via a combined intra- and extracranial approach.⁸⁻¹⁰ In the past, attempted total tumor removal has been condemned because of intraoperative deaths in early series^{3,8} and the risks of permanent damage to the optic nerves and other cranial nerves within the cavernous sinus. The argument for subtotal removal has been reinforced by occasional reports that residual tumor can spontaneously regress.¹¹ However, there have also been reports

Figure 5. Subcranial-intracranial view of angiofibroma penetrating the middle fossa dura. Temporal lobe dura is covered by Telfa to the right of the photograph. Temporalis muscle is retracted to the left of the photograph. Suction tip is pointing to the area of dural penetration. Angiofibroma is penetrating the dura of the middle cranial fossa (arrowhead) and adherent to temporal lobe (arrow).

Figure 6. Digital subtraction angiogram demonstrates arterial feeders from the intrapetrous and intracavernous internal carotid artery.

of rapid regrowth and progression despite radiation therapy.¹² Chemotherapy has also been advocated for use in advanced juvenile angiofibromas,¹³ but should only be used with the strictest of reservations for benign neoplasms.

We believe that total tumor removal can and should be performed via a combined intra- and extracranial approach. Tumors involving the cavernous sinus can often be safely removed without permanent neurologic impairment or risk to the patient.^{14,15} Many of the patients, who died as a result of the tumor itself or as a result of treatment complications, have had uncontrolled intracranial tumor at autopsy.^{8,10,11} Thus, we should strive for complete tumor removal. Undoubtedly, there will be cases in which intracranial or cavernous sinus involvement is so massive that surgical removal may endanger the patient's life or risk permanent loss of vision and other debilitating neurologic dysfunction. In these unusual situations one should consider adjunctive therapy after aggressive subtotal tumor removal. Watchful vigilance of tumor remnants in vital areas with repeat MRI scans can detect early tumor progression. However, with meticulous microdissection, total tumor removal can be achieved in most cases.

Removal of the tumor from below by peeling it off the dura⁷ should be condemned, since there are arterial feeders from the cavernous internal carotid in 93% of cases, which can be difficult to control from this approach (Fig. 6).¹ Unrecognized dural tears can result in meningitis. Intradural extension may occur, as in this case, resulting in higher recurrence rates if unrecognized. Thus, intracranial and sometimes intradural control and dissection of tumor are essential.

Dural penetration by juvenile angiofibroma is very

rare. Review of the literature revealed only five reported cases^{1,3} and in no instance was the tumor reported to be involving brain tissue, as was found in the case reported herein. Previous reports suggesting massive intracranial involvement by juvenile angiofibroma were limited by the lack of sophisticated radiologic imaging.⁸ This may be the first documented case of juvenile angiofibroma extending into the brain substance with successful surgical management.

CONCLUSIONS

1. Intracranial extension of angiofibroma often occurs. Intradural spread is rare, but needs to be excluded in all cases.
2. Total tumor removal is possible, even with cavernous sinus involvement.
3. Surgical removal should be performed via a combined intra- and extracranial approach.

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