Juvenile Nasopharyngeal Angiofibroma with intradural extension

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INTRODUCTION

The juvenile Nasopharyngeal Angiofibroma (JNA) is a benign vascular tumor of the skull base, which affects almost only male adolescents. It is a rare tumor, making up less than 0.05% all head and neck tumors. Although histologically benign, it may have a very aggressive behavior, extending to adjacent tissues and causing bone destruction by compression. The progression of such extension may cause intracranial involvement, which is relatively frequent, involving about 10% to 36% of the cases. Nonetheless, it rarely goes beyond the duramater.

CASE REPORT

The patient, a 16-year-old male, was referred to our service in August 2007 with a 4-month history of severe epistaxis, and also of pain and swelling in the left nasopharyngeal region. The patient had a past of epistaxis episodes and left nasal obstruction for 5 months; and a nasopharyngeal lesion seen upon nasal endoscopy. CT scan showed signs suggestive of an Andrews/Fisch IIB JNA. In August of 2006 he was submitted to embolization of the maxillary artery, ascending pharyngeal artery and sphenoidal sinus, cavernous sinus and temporal lobe. Angiography showed a major vascular component coming from the petrous and cavernous branches of the internal carotid artery (ICA); nonetheless, in some cases the MRI is able to identify the cleavage plane between the tumor and the cerebral parenchima vessels irrigating the tumor, allowing the tumor to be completely excised. While still under the care of the neurosurgery clinic, the patient developed dizziness upon manipulation of the pterygopalatine fossa, the pterygoid fossa, sphenoidal and infratemporal portions were digitally dissected and removed superiorly through the middle meningeal; nevertheless, there still was mild tumor irrigation from the cavernous branches of the internal carotid artery. Three days later we removed the tumor endoscopically. During the surgery, the patient developed an important bradycardia upon manipulation of the pterygopalatine fossa; thus, the procedure was interrupted, with an apparent complete tumor removal.

After 1 year of follow up, the patient had regained the lost weight, and the nasopharyngeal lesion was seen upon nasal endoscopy. CT scan and MRI showed an Andrews/Fisch IVB JNA, extending to the pterygopalatine fossa, the pterygoid fossa, sphenoidal sinus, cavernous sinus and temporal lobe. Angiography showed a major vascular component coming from the petrous and cavernous branches of the internal carotid artery (ICA); after embolization of external carotid artery branches, the main tumor feeding artery was coagulated with the bipolar. Considering the aforementioned aspects, in October of 2007 we decided for a craniotomy approach through the left frontotemporalzygomatic via for tumor resection, since the tumor may be detached from the ICA. Under microscopic view, the tumor was dissected from the middle fossa floor, which was opened by a high speed pneumatic burr, exposing the orbit, the round foramen with the maxillary nerve (V2), the oval foramen with the mandibular nerve (V3) and the cavernous sinus. The vascular connections between the ICA and the angiofibroma were coagulated with the bipolar cautery. Following that, we noticed that the angiofibroma was invading beyond the duramater, and after that, we saw that the tumor was in close contact with the temporal lobe, with a clivage plane. After removing its intradural part, the dural gap through which the tumor had passed was closed with a temporal fascia graft. The nasopharyngeal, sphenoidal and infratemporal portions were digitally dissected and removed superiorly through the middle fossa floor gap, which was later rebuilt by a temporal muscle. The histopathology analysis of the intradural specimen showed a close relation between the JNA and the dura mater, with invasion of the latter (Figure 1).

The patient developed left abducens nerve palsy in the post-operative, which lasted for 1 month. After 1 month of follow up, there were no signs of recurrence upon nasal endoscopy and MRI, or neurologic deficit, and the patient resumed his regular activities.

DISCUSSION

JNA with skull base erosion and intracranial extension courses with an increase in operatory risks, as well as the likelihood of significant vascular contributions of the ICA, which pre-operative embolization is not feasible. For these reasons, it is associated with higher hemorrhage rates of difficult control, neurologic deficits, subtotal resection and recurrence. Preoperative pre-operative embolization is also associated with recurrence, since the reduction of tumors with deep invasion of the sphenoid makes it difficult to completely excise it, with quick revascularization of the residual tumor in the immediate postop, especially by ICA branches. Based on the assumption that the bone destruction mechanism by the angiofibroma results from a compressive growth pattern instead of an infiltrative one, we see that intracranial extension is usually extradural.

Dura penetration is a rare phenomenon, with very few reports in the literature. Upon MRI, the transdural lesion suggestive signs are an absence of the cleavage plane between the tumor and the duramater, or the circunferential involvement of the ICA; nonetheless, in some cases the MRI is unable to distinguish the invasion as extradural or intradural. The presence of colateral branches of cerebral parenchima vessels irrigating the tumor, seen upon angiography, may confuse the duramater involvement.

Jafek et al. was the first to report on a case of JNA with dura penetration, treated by a combined otorhinolaryngological-neurosurgical approach. Lyons & Donald reported a case of intracranial surgical treatment of the JNA which penetrated the dura and the piaphragma of the temporal lobe. Butagun et al. reported three cases of dura transgression and cavernous sinus invasion (two of these were recurrent tumors), showing that prior manipulation predisposed the patient to dura invasion.

According to Danesi et al. the possibility of intradural extension cannot be denied; nonetheless, in order to prove it, the surgeon must show, histologically, that the tumor is invading the duramater or, alternatively, the intracranial tumor through an initial neurosurgery. Having said that, the case hereby reported fills both criteria concerning intradural involvement.

CONCLUSION

Although intracranial invasion by JNA is relatively common, its intradural extension is very rare, happening mainly in tumor recurrences. Usually the cleavage plane between the JNA and the cerebral tissue is well defined. Total tumor resection is possible with minimum neurological deficits, but dura reconstruction is necessary.

REFERENCES