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% of all head and neck tumors. Although histologically benign, it may have a very aggressive behavior, extending to adjacent tissues and causing bone destruction by compression1. The progression of such extension may cause intracranial involvement, which is relatively frequent, involving about 10% to 36% of the cases2. Nonetheless, it rarely goes beyond the dura mater3.

CASE REPORT

A 16 years old, male soccer goalie, with a past of epistaxis episodes and left nasal obstruction for 5 months; and a nasopharyngeal lesion seen upon nasal endoscopy. The CT scan showed signs suggestive of an Andrews/Fisch IVF JNA. In August of 2006 he was submitted to embolization of the maxillary artery, ascending pharyngeal artery and sphenoid branches of the internal carotid artery (ICA). 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 symptom recurrence; and a nasopharyngeal lesion was seen upon nasal endoscopy. CT scan and MRI showed an Andrews/Fisch IVF 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.

Considering the aforementioned aspects, in October of 2007 we decided for a craniotomy approach through the left frontotemporal zigomatic via for tumor ressection, since the tumor may be detached from the ICA. Under microscopic view, the dura mater was dissected from the middle fossa floor, which was opened by a high speed pneumatic burr, exposing the orbit, the roof fornmen 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 dura mater, and after opening it we saw that the tumor was in close contact with the temporal lobe, with a clivage plane. After removing its intradural portion, 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 rotating the 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 paralysis 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 operator 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. Previous pre-operative embolization is also associated with recurrence, since the reduction of tumors with deep invasion of the sphenoidal makes it difficult to completely excise it, with quick revascularization of the residual tumor in the immediate postop, especially by ICA branches3.

Based on the assumption that the bone destruction mechanism by the angiofibroma results from a compressive growth pattern instead of an infiltrative one4, we see that intracranial extension is usually extradural5.

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 dura mater, or the circumbential involvement of the ICA; nonetheless, in some cases the MRI is unable to distinguish the invasion as extradural or intradural. The presence of collateral branches of cerebral parenchima vessels irrigating the tumor, seen upon angiography, may reveal the dura mater involvement.

Jafek et al.6 was the first to report on a case of JNA with dura penetration, treated by a cervicocranial approach otolaryngological-neurological surgical approach. Lyonet et al.7 reported on a case of transcranial surgical treatment of the JNA which penetrated the dura and the piamater of the temporal lobe. Butugan et al.8 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.9 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 dura mater or, alternatively, see the intradural tumor through or, alternatively, see the intradural tumor through an intradural surgical treatment of the JNA which penetrated the dura mater. Hernando et al.8 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.

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 parenchima is well defined. Total tumor resection is possible with minimum neurological deficits, but dura reconstruction is necessary.

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