



Case of a Massive Juvenile Nasopharyngeal Angiofibroma with Cavernous Sinus and Middle Cranial Fossa Involvement

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Abstract

Introduction: Juvenile nasopharyngeal angiofibroma (JNA) is a rare, benign but locally aggressive tumor which typically affects adolescent males with an incidence of approximately 1:150,000.¹ The objective of this study is to report a case of a massive JNA that was successfully managed with pre-operative embolization and endoscopic resection and review the literature on these tumors.

Background: The underlying etiology of JNAs is unknown, but they are believed to be derivations of vasoproliferative or stromal cell neoplasms.² Because JNAs typically arise in adolescent males, a steroid hormonal influence has been suggested as a factor in the pathogenesis of this tumor.³ JNAs typically arise within the pterygoid canal and subsequently invade the sphenopalatine foramen, sphenoid sinus, and pterygopalatine fossa. Due to the highly vascular nature of JNAs, pre-operative embolization is recommended and has been found to reduce blood loss by a mean of 798 mL when compared to non-embolized patients.⁴

Results: A 17-year-old male presented to a tertiary rhinology clinic for evaluation a JNA that had been partially resected one year prior. He reported ongoing right nasal obstruction and epistaxis. Nasal endoscopy was notable for a large, hypervascular mass within the right nasal cavity. MRI demonstrated a large, enhancing sinonasal mass with skull base involvement. Pre-operative embolization was performed the evening prior to definitive surgery. Resection was performed with initial wide exposure via posterior septectomy, medial maxillectomy, and sphenoid drill-out. Intra-operative blood loss was estimated at 2 L, requiring multiple blood products.

Conclusions: Pre-operative embolization of JNAs is imperative to reduce blood loss, which may still be high. Exposure of the tumor prior to resection is key to controlling potential hemorrhage.

Methods

- The patient's medical record was reviewed, specifically examining the patient's perioperative course, imaging, and operative reports.
- A literature search was conducted on PubMed on the epidemiology, presentation, diagnosis, treatment, and outcomes of JNAs.

Case Report

- A 17-year-old with a history of JNA presented to a tertiary rhinology clinic for recurrent unilateral epistaxis and progressive ipsilateral nasal congestion.
- The patient had undergone partial tumor resection at an outside facility one year prior to presentation and noted subsequent rapid onset of nasal obstruction and epistaxis.
- CT sinus demonstrated a large sinonasal mass centered in the right sphenopalatine foramen (Figure 1).
- MRI face demonstrated a 8.9 x 5.0 x 5.9 cm vascular tumor that extended to the nasopharynx, sphenoid sinus, cavernous sinus, infratemporal fossa, foramen rotundum, and middle cranial fossa (Figure 2).
- The night before the planned surgery, pre-operative embolization was performed by interventional radiology (Figure 3). Percutaneous transnasal tumor embolization of the distal branches was also performed using n-BCA glue. There was substantial reduction of tumor vascularity after embolization.
- Endoscopic transnasal resection was performed as a joint surgical procedure with the rhinology and neurosurgery teams. Wide exposure of the tumor was obtained via posterior septectomy, medial maxillectomy, and sphenoid drill-out (Figure 4). Near-total resection of the tumor was obtained.
- Despite embolization, intra-operative blood loss was estimated at over 2 L and the patient required 4 units of red blood cells in the OR and 1 unit postoperatively.
- Postoperative MRI demonstrated near-total resection of the tumor (Figure 5).
- Histopathology was consistent with a sinonasal angiofibroma.
- The patient is five months post-surgery and is recovering well. He has not had any additional episodes of epistaxis and has no neurological deficits.



Figure 1. (A) Sagittal, (B) coronal, and (C) axial cuts from a non-contrast CT sinus demonstrating tumor extension into the right pterygopalatine fossa and the classically described Holman-Miller sign (anterior displacement of the posterior maxillary sinus wall) as denoted by the red arrow.

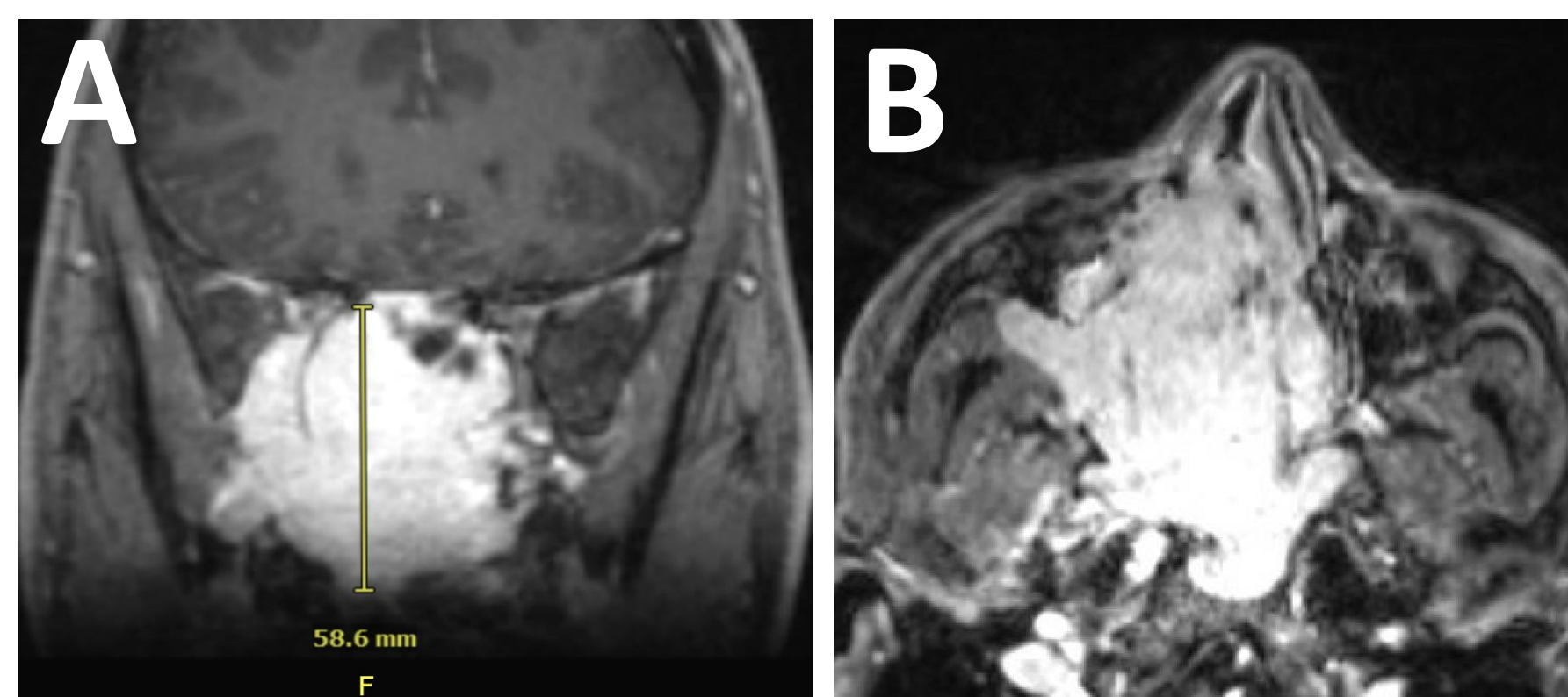


Figure 2. (A) Coronal and (B) axial cuts of a T1 post-contrast MRI brain demonstrating a contrast-enhancing mass measuring 8.9 x 5.0 x 5.9 cm with infratemporal fossa and middle cranial fossa involvement and extension into the anterior aspect of the right cavernous sinus with partial encasement of the right cavernous internal carotid artery.

Figure 3. Cerebral angiogram before (A) and after (B) embolization of the distal right internal maxillary artery, proximal right middle meningeal artery, right ascending pharyngeal artery, and distal left internal maxillary artery using PVA particles and coils.

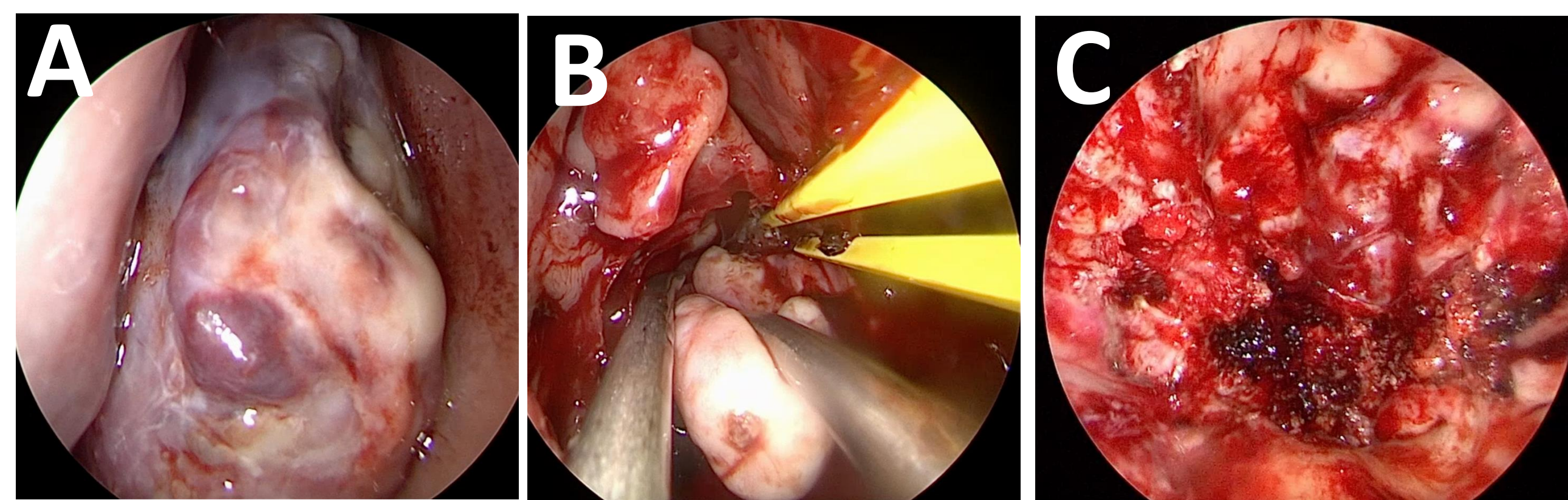
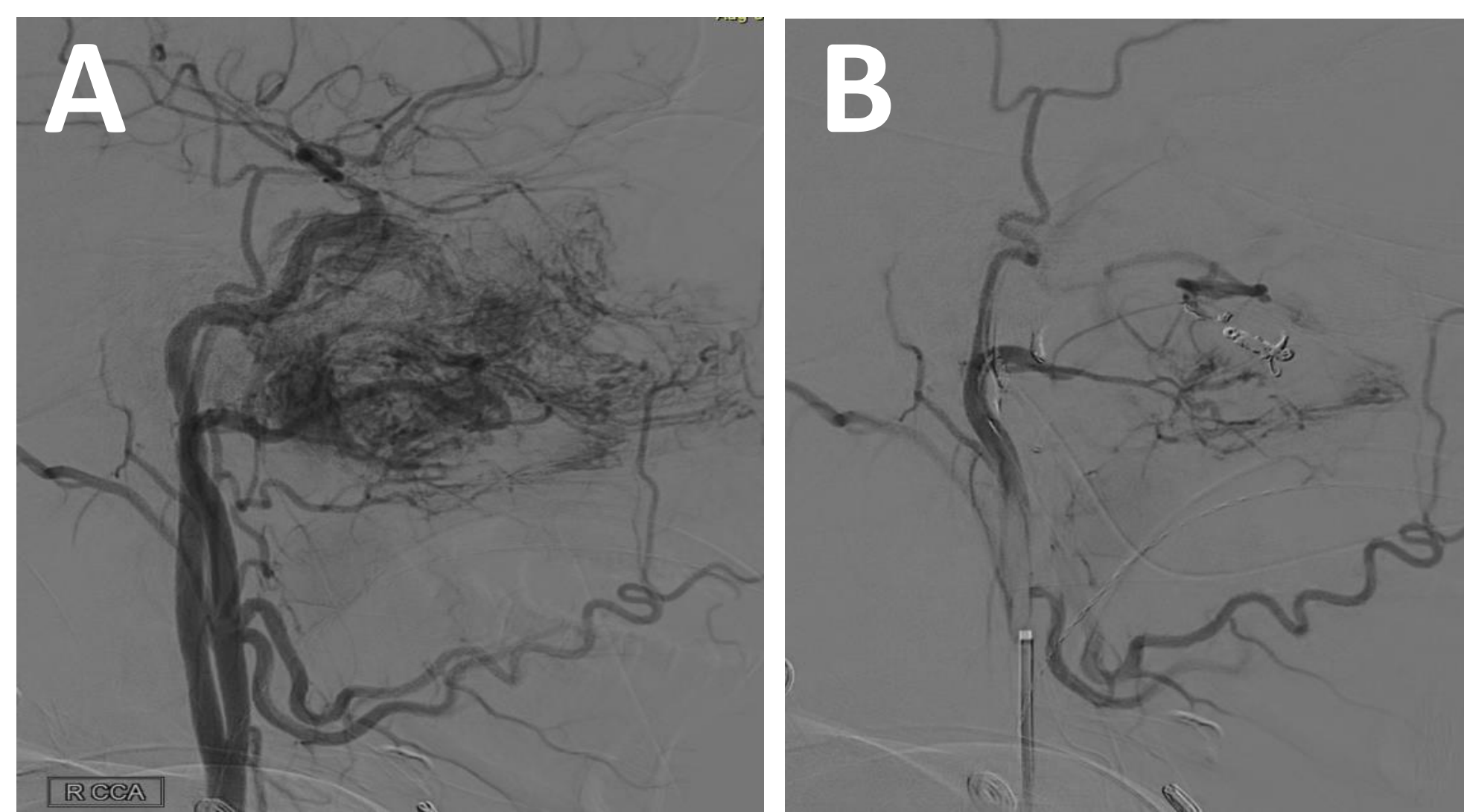


Figure 4. Endoscopic intraoperative views. (A) Large vascular tumor filling the nasal cavity. (B) Resection of the tumor was performed using a 2-surgeon technique. (C) Sinonasal cavity after the tumor resection.

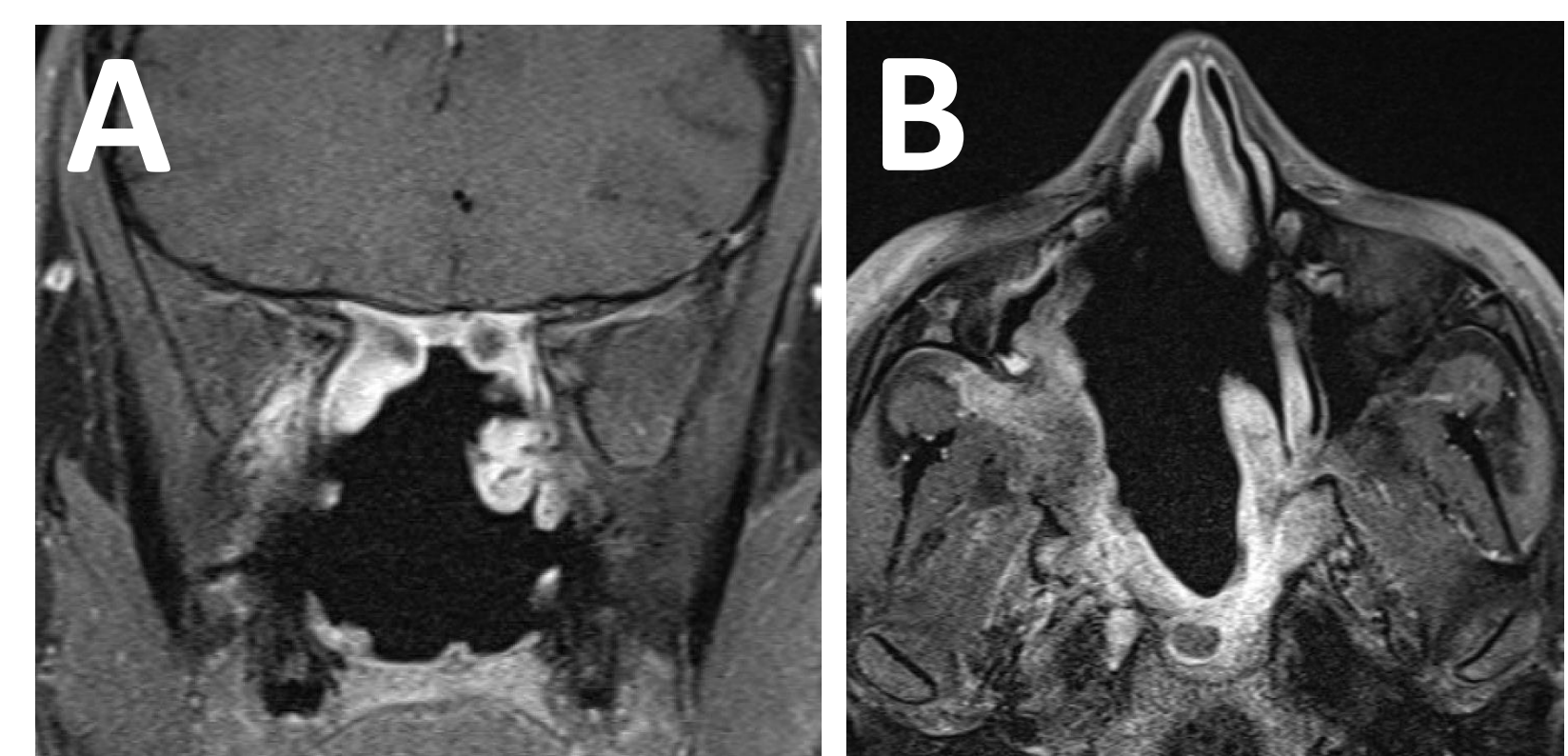


Figure 5. (A) Coronal and (B) axial cuts of a T1 post-contrast MRI face that was obtained 4 months postoperatively demonstrates near total resection of the tumor. Given the benign nature of JNAs, a small amount of tumor was left in the PPF, ITF, and cavernous sinus.

Conclusions

- Pre-operative embolization is important in the management of JNAs, but blood loss remains an important concern and surgeons should have blood products readily available for these cases.
- During preoperative embolization, both external carotid arteries should be evaluated, as bilateral vascularity may be present.
- Complete exposure of the tumor prior to resection is key to controlling hemorrhage and a more effective tumor resection. Debulking of the tumor may be necessary in large tumors prior to obtaining complete surgical exposure.

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