Introduction

Juvenile Nasopharyngeal Angiofibromas (JNA) are rare benign tumors of unknown etiology that occur in young males and are characterized by their high vascularity. Surgeons should select a surgical approach that provides adequate exposure of the tumor and permits the surgeon to control hemorrhage from the supplying vessels with minimal surgical morbidity. Preoperative assessment with computed tomography (CT), magnetic resonance imaging (MRI) and angiography is critical to evaluate the patterns of tumor growth and the vascular supply from branches of the internal and external carotid artery (ECA). Preoperative embolization is frequently utilized to minimize the extent of intraoperative blood loss. Recurrence rates following surgical resection exceed 20% in numerous published case series. Alternatively, low-dose radiotherapy of 30-36 Gy has a documented control rate of greater than 90%. This case report reviews the endoscopic-assisted surgical resection of a patient with a recurrent JNA and the unique management considerations when there is significant supply from the ICA.

Abstract

Objectives: To describe the management considerations of aggressive JNAs with atypical vascular contributions.

Study Design: Case report

Methods: This report describes a JNA that initially recurred following surgical resection and subsequently recurred after radiotherapy. Radiographically, the JNA extended from the pterygopalatine fossa into the nasopharynx. At the time of preoperative embolization, approximately 33% of its vascular supply was noted to arise from the internal carotid artery (ICA). The patient developed profuse epistaxis immediately following successful embolization of the external carotid supply to the JNA, which required tamponade via intranasal balloon placement until the time of surgery. An endoscopic-assisted resection of the JNA was successfully achieved following open medial maxillectomy that removed the lateral nasal wall and provided optimal access to the nasopharynx and pterygopalatine fossa. Profuse hemorrhage from the region of the Vidian Canal was endoscopically controlled with monopolar cautery.

Results: The JNA was resected en bloc without complication. The estimated blood loss was 2100 cubic centimeters, and four units of packed red blood cells were transfused intraoperatively. The patient’s postoperative course was uneventful. Subsequent magnetic resonance imaging surveillance documented the absence of recurrent disease.

Conclusions: Preoperative identification of atypical vascular contributions to JNAs permits the surgeon to employ an operative technique that will maximize exposure for uneventful resection and control of hemorrhage.

Case Report

Twenty-six year old male: Left-sided JNA

- 2001: Open surgical resection via lateral rhinotomy approach
- 2003: Recurrent JNA -- Radiotherapy with 3600 cGy in 180 cGy fractions once a day
- 2007: Recurrent epistaxis with radiographic recurrence—Referred to first author (JWW) for evaluation
- MRI findings: JNA extending from mid-pterygopalatine fossa into nasopharynx

Preoperative Embolization

- Super-selective angiography and embolization of the left ascending pharyngeal, sphenopalatine, and middle meningeal arteries with the Onyx ™ Liquid Embolization System: 67% of vascular supply embolized
- Remaining 33% of the vascular supply arises from inferolateral trunk of cavernous portion of left ICA (Vidian artery, perforators): not embolized
- Severe epistaxis developed following embolization, requiring balloon tamponade

Surgical Resection

- Lateral rhinotomy-medial maxillectomy with orbital floor preservation, creating a common nasoantral cavity
- Endoscopic sphenoidotomy and endoscopic-assisted removal of posterior maxillary sinus wall, gaining entry into pterygopalatine fossa
- Image-guided endoscopic cautery of Vidian Artery and perforators followed by en bloc endoscopic resection of JNA
- Dacrocystorhinostomy and closure of lateral rhinotomy
- EBL: 2100 cc, no surgical complications

Twelve Month Follow-up

- Asymptomatic
- Surveillance MRI negative
- Planned follow-up MRI and examination every six months

Conclusions

Thoughtful preoperative review of CT, MRI, and angiographic findings permits the surgeon to customize the surgical approach so that adequate surgical exposure is achieved for tumor resection and vascular control. An awareness of aberrant patterns of tumor growth and vascular supply during the preoperative planning phase minimizes the risk of adverse outcomes and excessive blood loss. Endoscopic-assisted resection of JNAs with image guidance is a valuable technique that is associated with minimal post-surgical morbidity.

References