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Case Report

Juvenile Nasopharyngeal Angiofibroma: A Case Report in Bangalore, South India

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Abstract

Juvenile nasopharyngeal angiofibroma is a tumor almost exclusively seen in adolescent males, constituting <1% of all neoplasms in head-and-neck. They are benign and locally spreading and are highly vascular in nature with life threating complications and thus prove to be a challenge for anesthesiologist to maintain controlled hypotension to decrease blood loss and to provide a clear field of vision for endoscopic surgery. Here, we present a case report of a 19-year-old male presenting with a mass in his left nasal cavity, with nasal obstruction and recurrent epistaxis. CECT of paranasal sinuses (PNS) paranasal sinuses contrast imaging revealed an intensely enhancing mass lesion in the nasopharynx extending into posterior aspect of the left nasal cavity, left maxillary sinus, sphenoid sinus, and infratemporal fossa with no evidence of intracranial extensions. The approach to its surgical management and perioperative and intraoperative management was a combined multi-disciplinary effort of otorhinolaryngology surgeons and anesthesiologists.

Key words: Angiofibroma, Benign, Vascular

INTRODUCTION

Juvenile nasopharyngeal angiofibroma is a tumour almost exclusively seen in adolescent males, constituting less than 1% of all neoplasms in head and neck. They are benign and locally spreading and are highly vascular in nature with life threating complications and thus prove to be a challenge for anaesthesiologist to maintain controlled hypotension, invasive monitoring, decrease blood loss and provide a clear field of vision for endoscopic surgery

CASE REPORT

A 19-year-old male presented with chief complaints of nasal obstruction which was unilateral (left) for a period of 6 months, followed by nasal bleeding that lasted for 5–10 min actively for 2–3 episodes/day, about a volume of 3–5 mL for 3 months. He was prescribed decongestants and got relieved intermittently over the period of time. Now, on presentation, he had 8–10 episodes of painless epistaxis, loosing approximately 60–70 mL of blood, for which he was admitted in our center he had no history of headache, vomiting, blurring of vision. Endoscopic examination of the nose revealed that the mass was congested, compressible, filling the left inferior, and middle meatus [Figure 1]. There was no tenderness on palpation and no external facial deformity. Probe test revealed no attachments in the nasal cavity. On posterior rhinoscopy, the mass could be seen extending into nasopharynx.

The orodental examination was normal. General examination revealed no significant abnormality. Routine hematological investigations were found to be within normal limits. Computed tomography (CT) revealed an ill-defined enhancing soft-tissue mass lesion involving the left maxillary sinus, posterior aspect of the left nasal cavity, anterior aspect of nasopharynx, and sphenoid sinus with bony changes possibly nasopharyngeal angiofibroma with sinus extension, minimal bilateral ethmoid and sphenoid sinusitis and deviation of nasal septum to the left side [Figure 2].

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CECT of paranasal sinuses shows intensely enhancing mass lesion in the nasopharynx measuring approximately 7.0 (AP) × 4.7 (ML) × 5.7 (CC) and extending into posterior aspect of the left nasal cavity, left maxillary sinus, sphenoid sinus, infratemporal fossa, no evidence of intracranial extensions, and bilateral ethmoid and sphenoid sinus noted.

Anesthesia Management

The diagnosis of juvenile nasopharyngeal angiofibroma (JNA) was made from the history and a CT scan finding pertaining to the nasal and PNS. Patient did not receive pre-operative blood transfusion, neither have co-morbidities nor predictable difficult airway (the modified Mallampati score was 2). Apart from standard routine laboratory tests (complete blood count, coagulation profile, and liver and renal function tests), CECT SCAN and angiogram were performed. He underwent embolization of the feeding artery (maxillary artery) 24 h before surgery to reduce the intraoperative bleeding.

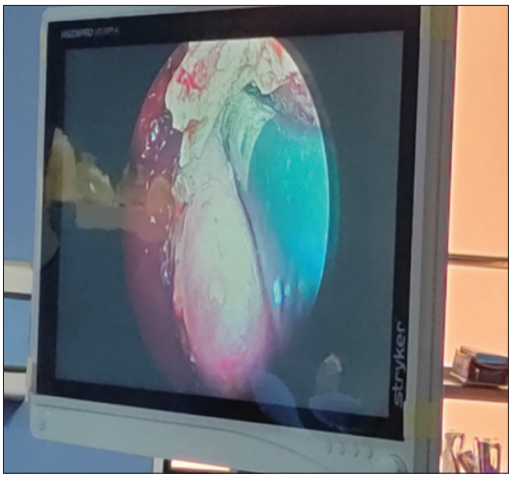

Figure 1: Endoscopic image of JNA

Figure 2: CECT PNS

After a thorough pre-anesthetic evaluation, written informed consent was taken and overnight fasting for 8 h, patient was taken into operation theatre. In the operation theatre, routine standard monitors (an electrocardiogram, oxygen saturation [SpO_2], and non-invasive blood pressure) were attached, and base line values were recorded. Two large bore IV cannula were secured with 16, 18 gauge on the upper limbs for peripheral lines. Patient was pre-medicated with intravenous glycopyrrolate 0.2 mg, intravenous ondansetron 4 mg, intravenous midazolam 1 mg, and with intravenous fentanyl (2 μg/kg). Anesthesia was induced with propofol (2 mg/kg) and neuromuscular blockade was achieved with vecuronium (0.1 mg/kg). After securing the oral cuffed flexometalic endotracheal tube of 8mm size, a throat pack was inserted. The patient was placed supine, in a 30°, reverse Trendelenburg position. The surgical approach was modified Denker's approach – medial maxillectomy.

For maintenance of anesthesia, patient was started on infusion dexmedetomidine 0.5 mcg/kg/h and vecuronium 4 mg/h infusion and N_2O: O_2:Isoflurane (50:50:>1). Hypotensive anesthesia was maintained intraoperatively throughout the surgery [Figure 3]. The tumour was excised intoto as shown [Figure 4] Blood loss was about 1800–2000 mL, patient was infused with crystalloids (intravenous [IV] ringer's lactate and IV normal saline) and there was 1-pint packed red blood cell intraoperative blood transfusion. Patient was hemodynamically stable throughout the procedure and was shifted to Intensive care unit and extubated next day.

Figure 3: Intraoperative endoscopic image showing clear surgical field & patient vitals

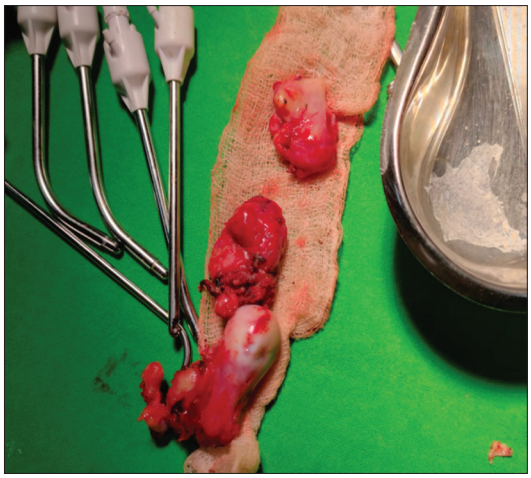

Figure 4: Picture showing excised JNA

Post-operative period was uneventful. The patient was shifted to ward on 2nd day and subsequently on 3rd day, no signs of bleeding noted and was discharged.

DISCUSSION

JNA is a rare tumor presenting mostly in adolescent males. Hippocrates described this tumor in the 5th century AD, but Friedberg first used the term angiofibroma in 1940.[1] The tumor is predominantly seen in adolescent males in the age group of 10–20 years, because hamartomatous nidus of vascular tissue in the nasopharynx activates to form angiofibroma when male sex hormones appear.[2] JNA originates close to the superior margin of sphenopalatine foramen, and further spreads locally into the pterygopalatine fossa, nasal cavity, and PNS.

It is made up of vascular and fibrous tissue. Mostly, the vessels are just endothelium-lined spaces with no elastic or muscular coat. This accounts for the severe bleeding as the vessels lose the ability to contract, so the bleeding cannot be controlled with application of adrenaline.

Patient most commonly presents with recurrent epistaxis, nasal obstruction, and mass in nasopharynx. They might also have conductive hearing loss, proptosis, and "frog-face deformity" swelling of cheek, infratemporal fossa, or involvement of IInd, IIIrd, IVth, and VIth cranial nerves will depend on the extent of tumor.

Diagnosis is based on clinical presentation. Biopsy of the tumor is attended with profuse bleeding and is, therefore, avoided. If it is essential to differentiate it from other tumors, biopsy can be done under general anesthesia with all arrangements to control bleeding and transfuse blood.

Radiological investigations and CT scan of the head with contrast-enhancement are the investigation of choice, it shows the extent of tumor, bony destruction, or displacements. Anterior bowing of the posterior wall of the maxillary sinus, often called "antral sign or Holman-miler sign," is pathognomic of angiofibroma. MRI is complementary to CT scans and shows any soft-tissue extensions present intracranially in the infratemporal fossa or in the orbit.

Recurrence has been suggested as a major complication in greater part of surgical literature,[5-8] despite the benign and unifocal origin of JNA. Tumor recurrence is primarily seen with high grade tumors with ICE, in part, due to failure to recognize skull base erosion.[2,9] Recurrence rate for JNA has been reported from 17% to 86%.[9]

Histologically, JNAs originate from myofibroblasts. The tumor usually spreads submucosally and is non-encapsulated. It is composed of intricate mixture of stellate and staghorn blood vessels with variable vessel wall thickness ranging from single layer of endothelium to variable smooth muscle coat, irregular fibrous stroma (loose edematous to dense and acellular) positive stains include c-Kit/CD117 and androgen receptor (75%).[8]

Surgery remains the main stay of treatment.[10] Endoscopic nasal microsurgery is a viable approach with prospectively lesser post-operative morbidity and intraoperative blood loss, but it is advocated only for limited lesions. However, for larger tumors, approaches such as trans palatal, lateral rhinotomy and midfacial degloving have been suggested. Pre-operative embolization of tumor is recommended with all of these approaches to limit the intraoperative blood loss.[3,4]

CONCLUSION

JNA, occurring exclusively in adolescent males, is a benign and vascular rare tumor of the nasopharynx. Diagnosed clinically with features of recurrent epistaxis, nasal obstruction, and mass in the nasal cavity.

A contrast CT is pathognomonic for the diagnosis of JNA and allows accurate staging of the tumor which is necessary for choosing the surgical technique, estimating prognosis, and reporting results. MRI and angiography are also preferred techniques for surgical planning.

Despite advances in surgical and anesthetic techniques, surgery for JNA remains a challenge for anesthesiologists' latter should be aware of advances in the management of these tumors, the latest drugs, monitoring modalities, and blood conservation strategies, as well as anesthetic techniques, which can help to provide optimal perioperative

care of these patients. Pre-operative embolization and controlled hypotension are major factors used to decrease blood loss during endoscopic resection of JNAs. Multimodal blood conservation techniques, in combination with hypotensive anesthesia, should be used to decrease blood loss and provide a drier field during endoscopic resection of a JNA.

Endoscopic surgery was preferred over open surgery under hypotensive general anesthesia as, in our case, to prevent intraoperative bleeding.

REFERENCES

1. Mehmet A, Orhan S, Suphi M. Juvenile nasopharyngeal angiofibroma: Radiological evaluation and pre-operative embolization. KBB Forum 2006;5:58-61.
2. Antonelli AR, Cappiello J, Di Lorenzo D, Donajo CA, Nicolai P, Olandi A. Diagnosis, staging, and treatment of juvenile nasopharyngeal angiofibroma (JNA). Laryngoscope 1987;97:1319-25.
3. Alves FR, Granato L, Soares M, Lambert E. Surgical approaches to juvenile nasopharyngeal angiofibroma-case report and literature review. Intl Arch Otorhinolaryngol 2006;2:162-6.
4. McDaniel RK, Houston GD. Juvenile nasopharyngeal angiofibroma with lateral extension into the cheek: Report of case. J Oral Maxillofac Surg 1995;53:473-6.
5. Harrison DF. The natural history, pathogenesis, and treatment of juvenile angiofibroma. Personal experience with 44 patients. Arch Otolaryngol Head Neck Surg 1987;113:936-42.
6. Lloyd G, Howard D, Phelps P, Cheeseman A, Diver J. Juvenile angiofibroma: Modern imaging and its influence on the surgical treatment of juvenile angiofibroma. J Laryngol Otol 1999;113:127-34.
7. Elhammady MS, Johnson JN, Peterson EC, Aziz-Sultan MA. Preoperative embolization of juvenile nasopharyngeal angiofibromas: Transarterial versus direct tumoural puncture. World Neurosurg 2011;76:328-34.
8. Oliveira JA, Tavares MG, Aguiar CV, Azevedo JF, Sousa JR, Almeida PC, *et al*. Comparison between endoscopic and open surgery in 37 patients with nasopharyngeal angiofibroma. Braz J Otorhinolaryngol 2012;78:75-80.
9. Radkowski D, McGill T, Healy GB, Ohlms L, Jones DT. Angiofibroma. Changes in staging and treatment. Arch Otolaryngol Head Neck Surg 1996;122:122-9.
10. Mair EA, Battiata A, Casler JD. Endoscopic laser assisted excision of juvenile nasopharyngeal angiofibroma. Arch Otolaryngol Head Neck Surg 2003;129:454-9.

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