

Removal of Angiofibroma with Maxillary and Mandibular Swing: A Clinical Report

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ABSTRACT

Juvenile nasopharyngeal angiofibroma is vascular tumor found in adolescent males. With development of endoscopic fraternity, tumor can be addressed successfully with endoscopic approach; but one needs maximum exposure for large angiofibroma with local infiltration around. Maxillary and mandibular swing technique for removal of the tumor gives excellent exposure and good control on vascularity of the tumor.

Postoperative follow-up for 1 year has shown minimal visible scar, cosmetic deformity in the patient.

Keywords: Juvenile nasopharyngeal angiofibroma, Mandibular swing, Maxillary swing, Weber-Fergusson incision.

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INTRODUCTION

Though most common of all benign nasopharyngeal tumors, juvenile nasopharyngeal angiofibroma (JNA) is rare and less than 0.5% of all head and neck tumors. JNA is highly vascular tumor occurring exclusively in adolescent males. The lesion is locally expansile in pushing manner with some capacity to infiltrate local structures. These tumors have the potential to cause life-threatening complications secondary to bleeding and intracranial extension.²

EPIDEMIOLOGY AND ETIOLOGY

The tumor arises at the junction of sphenoidal process of palatine bone and pterygoid process of sphenoid bone, just superior to sphenopalatine foramen.

Ipsilateral internal maxillary artery is main blood supply via distal branches sphenopalatine or vidian artery. There can be contribution from ICA via mandibular and inferolateral trunk in 50% cases.

Sternberg¹⁶ and Hubbard¹⁷ suggested angiofibromas as vascular neoplasms similar to hemangioma. Maurice and Millard interpreted angiofibromas as hamartomas resulting from misplaced genital tissue. Beham concluded them as vascular malformations.

APUD cell theory proponents thought that these tumors originate from non-chromaffin paraganglionic cells present at the terminal end of the maxillary artery.¹⁸

Immunohistochemical studies have shown transforming growth factor B1, IGF-II might be growth regulators of the tumor.⁶ Platelet derived growth factor (PDGF-B) contribute partly to neovascularization and fibrosis.¹⁹

Recent studies have suggested angiofibroma is extra-colonic manifestation of familial adenomatous polyposis.²⁰ Immunohistochemical studies also suggest that stromal cells rather than endothelial cells are neoplastic cells of JNA.¹

Extensive research to discover etiology of the tumor has not yet concluded and no theory has gained widespread acceptance.¹

PRESENTATION

Nasal obstruction with recurrent epistaxis is common presentation. On lateral growth, neoplasm erodes into pterygoid plates and extends into infratemporal fossa via pterygopalatine fissure. In 20 to 35% cases intracranial extension is observed in middle cranial fossa, pituitary fossa and anterior cranial fossa.²

HISTOLOGY

JNA is unencapsulated neoplasm consisting of endothelially lined vascular spaces and fibrous connective tissue. The nasopharyngeal surface is lined by mucosa. The tumor has actively proliferating part and nonactive part with transition zone in between. The actively proliferating part has hyperplastic capillaries and venous sinuses that lack muscular components and in periphery of the tumor.

In central part, tumor has more fibrous component. This suggests that JNA is reactive process or defective development rather than neoplastic process.

RADIOLOGICAL EVALUATION

Contrast-enhanced CT scan and magnetic resonance imaging (MRI) are useful for evaluation. CT scan will show bony erosion and orbital fissure enlargement.

MRI will show pterygomaxillary, infratemporal space or intracranial extension with soft tissue differentiation. It can determine whether intracranial extension is intradural or extradural.

Angiography can be done preoperatively to evaluate the source of blood supply and to selectively embolize feeding vessels.

DIFFERENTIAL DIAGNOSIS

Nasopharyngeal carcinoma, rhabdomyosarcoma, Kaposi sarcoma, neovascularized inflammatory polyps, teratoma, hemangioma and lymphoproliferative disorder.²

Staging

*Fisch Staging System*⁴

- Stage I: Tumor limited to nasopharyngeal cavity, bone destruction negligible or limited to the sphenopalatine foramen.
- Stage II: Tumor invading the pterygopalatine fossa or the maxillary, ethmoid or sphenoid sinus with bone destruction.
- Stage III: Tumor invading infratemporal fossa or orbital region:
- (A) Without intracranial involvement.
 - (B) With intracranial extradural (parasellar) involvement.
- Stage IV: Intracranial intradural tumor:
- (A) Without infiltration of the cavernous sinus, pituitary fossa or optic chiasm.
 - (B) With infiltration of the cavernous sinus, pituitary fossa or optic chiasm.

*Radkowski et al System*⁵

- Stage I: (A) Tumor limited to nose or nasopharyngeal vault.
- (B) Extension into sinuses.
- Stage II: (A) Minimal extension into pterygomaxillary fossa.
- (B) Full occupation of pterygomaxillary fossa with or without erosion of orbital bones
- (C) Posterior to pterygoid plates.
- Stage III: (A) Erosion of skull base (middle cranial fossa or pterygoids).
- (C) Erosion of skull base with intracranial extension with or without cavernous sinus.

DISCUSSION

Treatment Options

Surgery remains primary treatment with various external approaches including lateral rhinotomy, transpalatal, transantral and midfacial degloving, with Le Forte I osteotomy or medial maxillectomy.^{10,14} Endoscopic approach reserved for lower stage JNA, preferably with preoperative embolization by some authors.^{1,7,11} Some authors doubt utility of embolization in small and medium sized tumors.^{3,8} For large tumors (stage II B onward), open

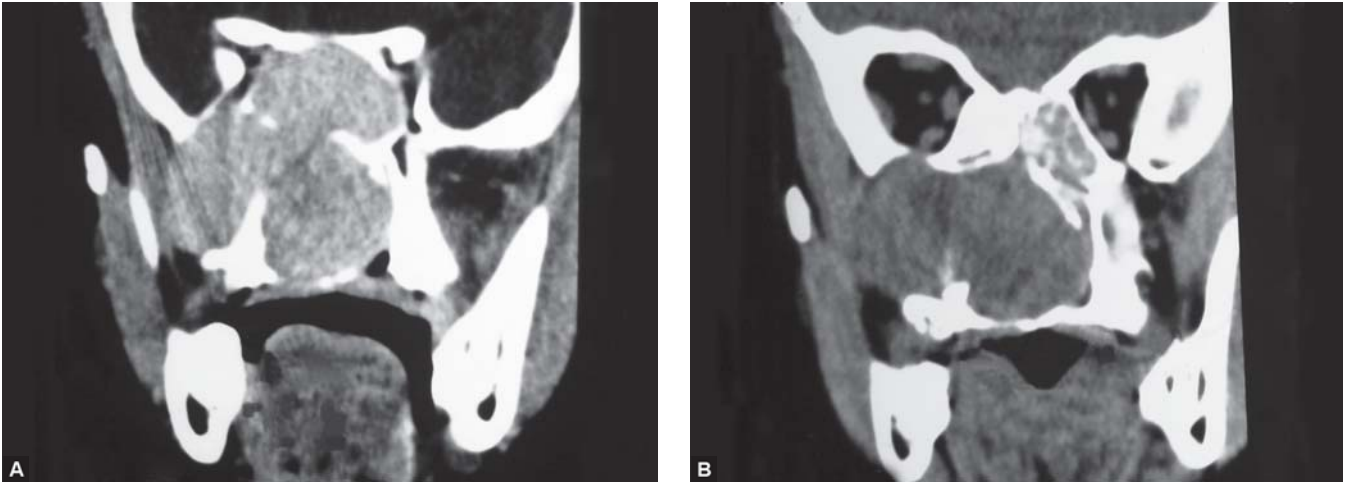
approach is safe alternative to endoscopic removal.¹⁵ As open approach gives wider exposure, chances of recurrence are least and using endoscope in open approach enhances utility of both the concepts.^{13,15} Risk of recurrence increases with invasion into vidian canal and sphenoid bone. Drilling sphenoid, pterygoid plates and the clivus to minimize recurrence treats invasion of tumor into cancellous bone of sphenoid.^{11,12} First surgery for excision of tumor is best considering difficulty in excision of recurrent tumor owing to loss of planes.^{1,12}

Radiotherapy is reserved for tumors with intracranial extension, unresectable tumor due to proximity to vital structures.⁹

Case Details

A 13 years old boy came to OPD with chief complaints of snoring since 3 months, epistaxis from right nostril and open mouth during sleep since 2 months. Snoring started in insidious manner, was progressive and with 2 to 3 apnoeic episodes. He had 10 to 15 episodes of spontaneous epistaxis per right nostril, lasted for 30 minutes with around 50 to 60 cc blood loss in 2 to 3 episodes and stopped spontaneously. There is no h/o swelling of the cheek, trismus, hearing loss secondary to Eustachian tube obstruction, anosmia, trauma to nose, change in voice, diminished visual acuity, diplopia, proptosis, facial pain, headache, spontaneous bleeding from other body sites, loss of consciousness. There was h/o grade I consanguineous marriage. His sleep was disturbed, appetite reduced, bowel and bladder habits were normal. He lost 2 kg weight in 3 months. His pulse, blood pressure, along with systemic examination, especially all cranial nerves examination was normal.

On OPD examination, oral cavity, throat, ear examination was normal and on anterior rhinoscopy, abundant mucopurulent secretions seen along the floor of right nostril. Nasal endoscopy with rigid Hopkins rod showed vascular white soft to firm, compressible mass in right nasal cavity along the floor, and occupying almost whole posterior choana as seen via left nostril. The lesion was submucosal and bled on probing. Patient was subjected to CT scan with contrast which showed that large lobulated intensely enhancing mass involving right maxillary sinus extending medially to right nasal cavity with destruction of medial wall of maxillary sinus and pushing nasal septum on opposite side (Fig. 1A). Posterosuperiorly it destroyed the inferior wall of right orbit, ethmoidal cells, floor of sphenoid sinus and floor of pituitary fossa. Posterolaterally it was extending to nasopharynx. It was extending to right pterygopalatine fossa with mild destruction of base of the plates. The hard palate was destroyed posterolaterally on



Figs 1A and B: Extent of the JNA

right side. There was mild protrusion of mass in inferior part of suprasellar cistern. Posteriorly mass was touching to the prevertebral soft tissue. The intraocular soft tissue planes with extraocular muscles and optic nerve were normal (Fig. 1B).

Intraoperative blood loss in after embolization is certainly less. In view of nonavailability of interventional radiology and doubtful efficacy of embolization in medium size tumors, we decided to proceed without preoperative embolization.

Patient was posted for removal of JNA with open approach with maxillary and mandibular swing considering Stage III (A) Radkowski tumor. Avoidance of recurrence was as important as wide exposure for total removal.

Surgery

Procedure was planned under general anesthesia with oral endotracheal intubation. Elective tracheostomy was done to replace endotracheal tube (ET) with 6.5 number portex cuffed tracheostomy tube in view of maxillary and mandibular swing.

Prophylactic IV antibiotics started 24 hours prior to surgery. The patient placed in supine position with neck extended with table inclined in a head-up position so as to reduce venous pressure in head and neck region.

Weber-Fergusson incision on right face with transcervical approach with medial mandibulotomy was planned to approach the tumor.

Surgery was off the mark by incision in neck at the level of hyoid bone from midline to anterior border of sternomastoid, then curving superiorly to right mastoid tip. The incision continued through subcutaneous tissue and platysma to the superficial layer of deep cervical fascia.

Dissection continued in subplatysmal layer exposing the anterior belly, intermediate tendon and posterior belly of

digastric muscle along with anterior border of sternomastoid. Anterior facial vein was divided and ligated. Anterior facial vein and overlying superficial layer of deep cervical fascia are carefully elevated over submandibular gland to reflect mandibular branch of facial nerve away from the operative field. The facial artery was identified and ligated. Lingual artery was ligated. Submandibular gland was then mobilized and retracted anteriorly and superiorly with presevation of hypoglossal nerve, marginal mandibular nerve. Tail of parotid gland was then mobilized and freed from sternomastoid and digastric muscle. Carotid sheath was then exposed by retracting sternomastoid posteriorly.

The transcervical incision was then carried up the midline of the neck to the level of the mandible, curved round the contour of the chin and back to the midline of the lower lip. The lip was transected in the midline. On completion of the skin incision the periosteum overlying mandible incised and elevated. Prior to osteotomy drill holes were made for plating the mandible. Mandibulotomy was achieved through right paramedian incision between lateral mandibular incisor and first premolar tooth. Gum papillae were raised on either side of mandibular teeth to achieve strong floor of mouth postoperatively so that chances of salivary fistula can be minimized. Then right half of mandible swung laterally after dividing mylohyoid muscle. Submandibular gland retracted superiorly and ECA ligated at the level of facial artery (Fig. 2).

For maxillary swing, Weber-Fergusson incision was modified. It started 2 cm lateral to outer canthus of right eye to expose zygomatic arch. The incision curved in region of medial canthus in obtuse manner to allow ample blood perfusion. The upper lip was transected in midline. Incision then passed between central incisors and continued in midline of hard palate up to the junction with soft palate, turning laterally toward tuberosity of maxilla. Maxilla was left attached to anterior cheek flap. Before osteotomy, holes

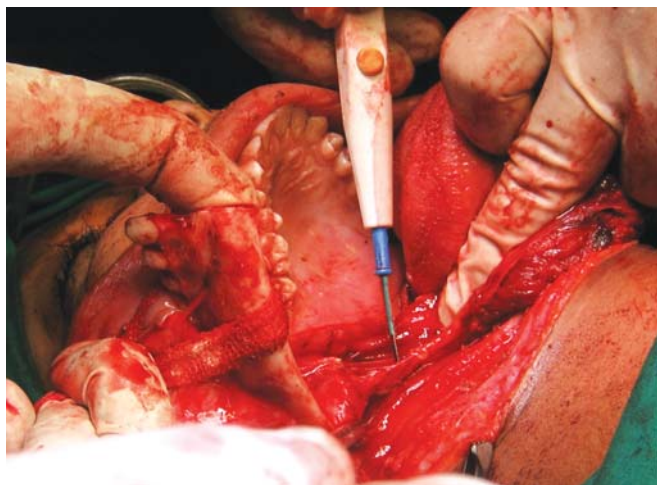


Fig. 2: Mandibular swing

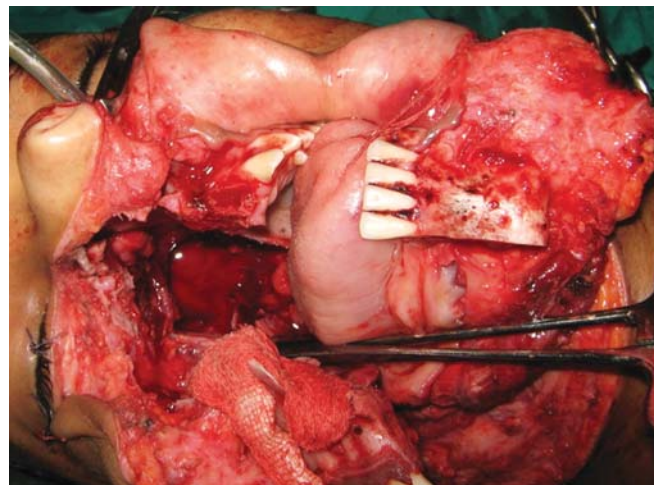


Fig. 3: Mandibular and maxillary swing with tumor removal



Fig. 4: Angiofibroma removed

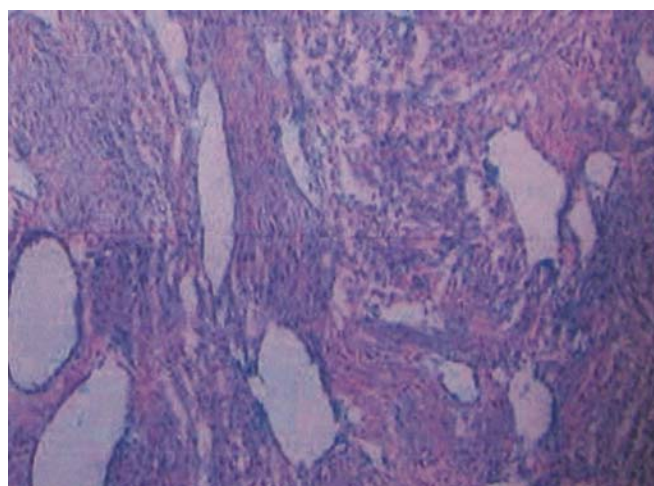


Fig. 5: Microscopic view

are drilled for the miniplates over zygomatic arch, under nasal spine and superomedial aspect of anterior wall of the maxilla. Right maxilla was then swung laterally to expose nasopharyngeal and paranasopharyngeal area (Fig. 3). There was tumor occupying nasopharynx and extending to right pterygopalatine fossa, presellar region. Tumor was removed *en masse* due to wide exposure and easy differentiation from surrounding structures on palpation (Fig. 4). Hemostasis achieved with packing. Swung maxilla and mandible realigned to their natural position and fixed with titanium plates (Fig. 6). Suturing was done in layers. Merocele was placed in both nostrils.

Patient stood procedure well and shifted to ICU for intensive monitoring. He was in ICU for 4 days, and then shifted to ward. Drain output became serous, considerably reduced and hence removed on POD 10. Patient had lower gingivobuccal sulcus infection with small soft palate suture dehiscence. Timely treatment cured both the complications with secondary healing. Patient was discharged on POD 20, with successful weaning of tracheostomy tube. He received 4 points of whole blood and 1 point of fresh frozen plasma

in first 4 POD. Surgery was accomplished by team approach with head and neck surgeon, maxillofacial surgeon and anesthetists.

Follow-up and Outcome

Histopathological examination of excised mass was confirmatory of angiofibroma. On gross examination it was soft to firm nodular mass $4.8 \times 4.0 \times 3.5$ cm grayish white homogenous mass (Fig. 4). Histological examination showed arborization of dilated thin-walled vascular channels separated by fibrous tissue. No evidence of degenerative changes or malignancy in any sections studied (Fig. 5).

Patient was followed up in OPD for 1 year with nasal endoscopy during each visit. Even though having scar on the face, there is no recurrence of the tumor till date (Fig. 7). Follow-up after 1 year with faint facial scar.

Even though endoscopic surgery has become preferred approach in JNA patients, open approach is safe alternative in comfortable *en masse* removal of large tumors under direct vision, it gives acceptable facial scar without bony



Fig. 6: After plating and suturing of the incision



Fig. 7: Follow-up after 1 year with faint facial scar

defect, keeps us abreast with open approach as any endoscopic surgery might need to be converted in open incision. It is of great advantage for periphery hospitals with no availability of interventional radiology facility. We found total maxillary swing to be a relatively safe alternate approach for the resection of NPA. We removed the entire tumor in a wider surgical field, with reduced blood loss and minimal complications or chance of recurrence.

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