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A Case Series of: Juvenile Angiofibroma

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Abstract: ***Juvenile nasopharyngeal angiofibroma is a rare benign tumour of vascular origin found in adolescent males, originating around the sphenopalatine foramen. Although the exact pathogenesis of the tumour is not yet known, natural history and growth patterns can be predicted. JNA progressively involves the nasopharynx, nasal cavity, paranasal sinuses, pterygopalatine fossa, infratemporal fossa and, in severe cases, an orbital or intracranial extension can be seen. Early diagnosis based on clinical examination and imaging is mandatory to ensure the best resectability of the tumour, as small to moderate tumours can be managed exclusively endoscopically. Preoperative angiography can reveal the vascular sources and allow embolization to prevent significant bleeding. We present a brief literature review followed by our case series of 5 juvenile nasopharyngeal angiofibromas.***

Keywords: Juvenile Angiofibroma, ENT, Otorrhinolaringology, facial swelling

1. Introduction

Juvenile Angiofibroma (JA) is an uncommon, benign and extremely vascular tumor. It develops exclusively in adolescent males and accounts for less than 0.5% of all head and neck tumors (1).

The tumor arises in the tissue within the sphenopalatine foramen, starts to grow in the submucosa of floor of nasopharynx, may grow anteriorly into nasal cavity, laterally towards the pterygomaxillary fossa and eventually may invade the infratemporal fossa and the middle cranial fossa.

Several staging classifications has been proposed. Following two are most practical and commonly used classifications.

2. Classification

Fisch Staging [(2)]

Stage	Details
1	Tumor limited to nasopharynx or to sphenopalatine foramen with negligible bone destruction.
2	Tumor invading the pterygopalatine fossa or the maxillary, ethmoid, sphenoid sinus with bone destruction.
3	Tumor invading infratemporal fossa or orbital region
	(a) Without intracranial involvement
	(b) With intracranial extradural involvement
4	Intracranial intradural tumor
	(a) Without infiltration of cavernous sinus, pituitary fossa or optic chiasm
	(b) With infiltration of cavernous sinus, pituitary fossa or optic chiasm

Radkowski Classification [(3)]

Stage	Details
IA	Limited to nose & nasopharyngeal area
IB	Extension into one or more sinuses
IIA	Minimal extension into pterygopalatine fossa
IIB	Occupation of pterygopalatine fossa without orbital erosion
IIC	Infratemporal fossa extension without cheek or pterygoid plate involvement
IIIA	Erosion of the skull base (middle cranial fossa or pterygoids)
IIIB	Erosion of the skull base with intracranial extension with or without cavernous sinus involvement

3. Material and Methods

This is a retrospective review of 5 patients with JA cases presented to our center from June 2017 to August 2018. The demographic data, clinical findings, imaging investigations and treatment modalities of these patients were collected and reviewed based on medical records.

All 5 patients were male. The mean age of diagnosis was 16 (range 11 - 19) years old. Three patients were presented with spontaneous, recurrent, painless epistaxis (60%) and two patients were presented with progressive nasal blockage (40%) with occasional scanty nasal bleeding. Other associated symptoms were headache (60%), reduction in sense of smell (60%), facial swelling (Frog face deformity) (20%).

Nasal endoscopy examination of nasal cavity revealed a vascular nasal mass occupying the nasopharynx in all the patients.

- All patients had preoperative imaging including contrast enhanced computed tomography (CECT scan) and angiography.
- The classifications were used to classify all tumors based on patients' CT scan reports according to the FISCH classification.
- The operations were performed under general anesthesia.
- The modality of treatment for three patients was endonasal endoscopic approach as they were fallen under stage 2 of FISCH classification and the rest of the two were opted for mid facial degloving approach as they were having stage 3a of JA.

4. Case Reports

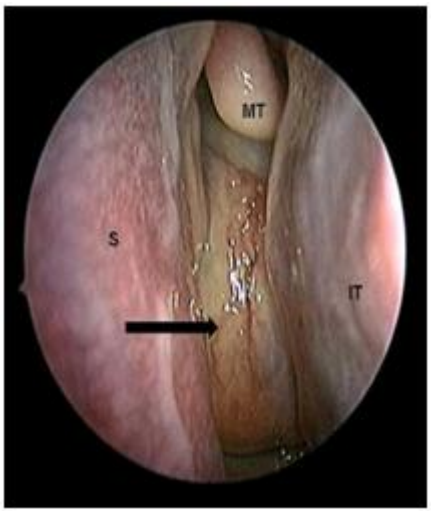

Figure 1: Nasal endoscopic findings showing irregular vascular mass lesion within left nasal cavity.

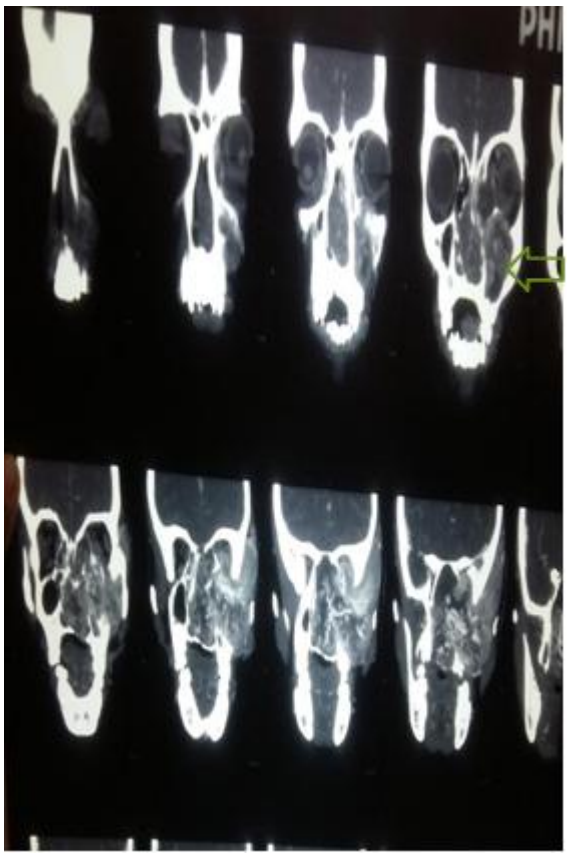

Figure 2: Axial CT scan of the same patient showing extensive nature of JA. The lesion was measured 70x48x52mm in size extending into pterygopalatine and infratemporal fossa and also invading infraorbital region.

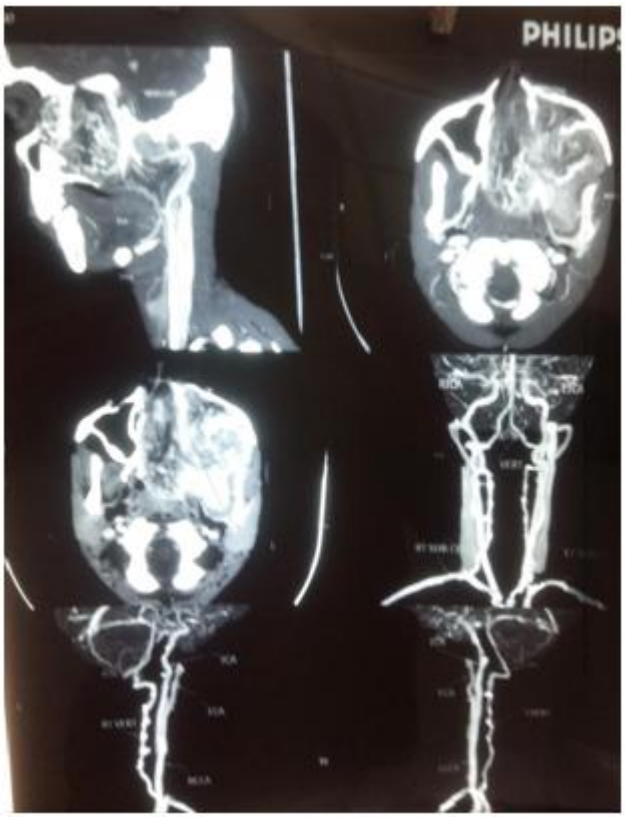

Figure 3: CT angiography image of the patient showing the "feeding vessel" - internal maxillary artery, branch of the external carotid artery.

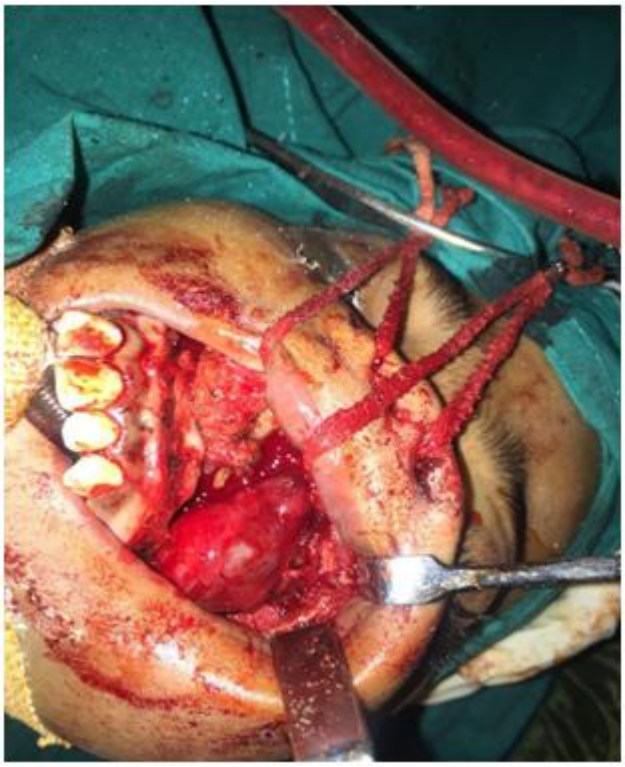

Figure 4: Intraoperative pictures of the same patient who was operated via midfacial degloving approach

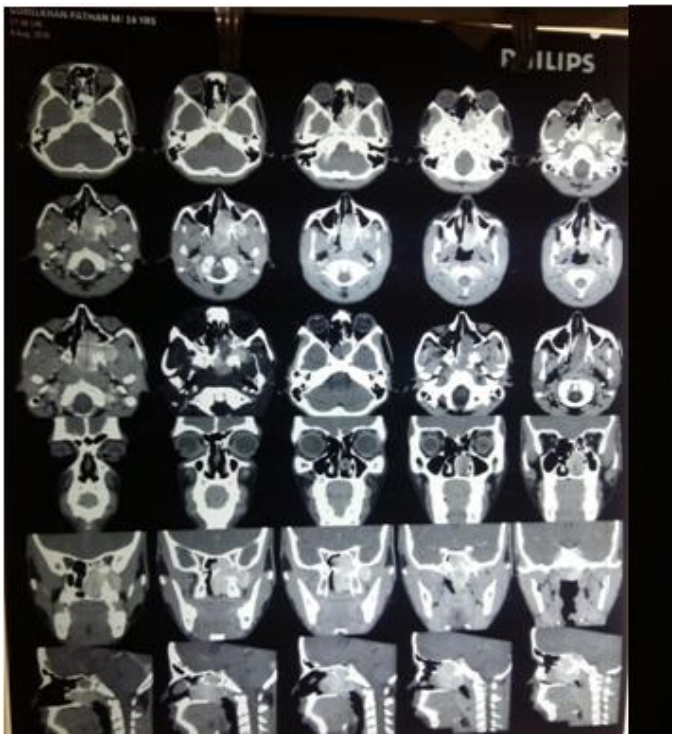

Figure 5: Above is the CT scan of another case of our study which was having stage 2 juvenile angiofibroma.

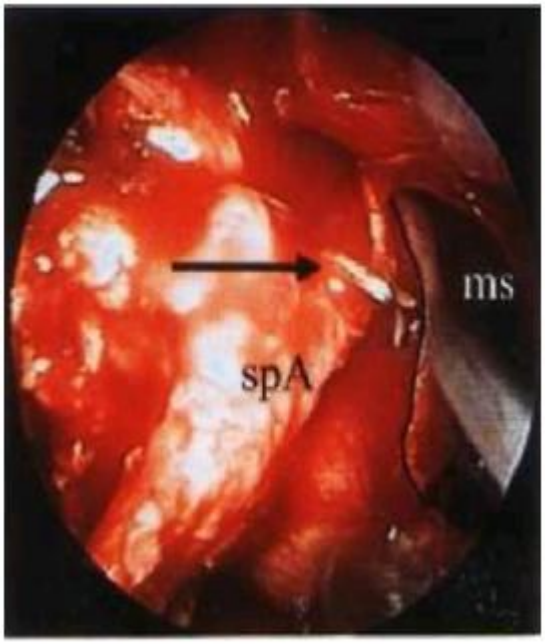

Figure 6: This is the picture of one of the case who was operated via endonasal endoscopic approach of JA excision showing the sphenopalatineartery, maxillary sinus opening and the mass lesion.

5. Result

All five patients underwent Contrast enhanced CT scan. FISCH Classification was used to stage the patient based on CT imaging result.

Three (60%) patients were at stage 2 and two (40%) patient were at stage 3a. Three (60%) patients were presented with JA arising from left side while two (40%) patients raised from right side. All patients underwent angiography imaging and showed all tumors received blood supply mainly (80 - 90%) from ipsilateral internal maxillary artery with few branches from ascending pharyngeal artery.

According the classification, the treatment modality was chosen. Three patients who were having stage 2, operated via endonasal endoscopic approach and the rest two patients who were having stage 3a were operated via midfacialdegloving approach.

External carotid artery was controlled at the time of surgery instead of preoperative embolization of feeding vessel.

In endonasal endoscopic excision, following steps were performed.

- After induction of anesthesia the nose was prepared with a vasoconstrictor solution
- The anterior end of middle turbinate and inferior aspect of inferior turbinate was resected.
- An ant ethmoidectomy together with removal of the medial wall of the maxillary sinus gave access to the post wall of the antrum
- This was then removed to achieve complete lateral exposure of the tumor.
- Posterior ethmoidectomy was done, sphenoid rostrum was reached.
- Then the tumor was dissected off with the help of bipolar cautery and suction cautery from adjacent structures; septum, sphenoid rostrum, lateral nasal wall, pterygopalatine fossa and nasopharynx
- Hemostasis was achieved and bilateral nasal cavities were packed.

In midfacial degloving approach for JA excision, following steps were performed.

- The procedure was started with complete transfixion incision, which is connected to bilateral intercartilagenous incisions and nasal floor incisions.
- Soft tissue was released from the dorsum of the nose.
- Sublabial incision was carried from first molar to molar across the midline.
- Wide subperiosteal dissection was carried out, adhesions between maxillary and nasal tunnels were released and the nasal - labial complex retracted over the nasal bridge.
- Anterior wall of maxilla drilled out.
- Medial maxillectomy performed, posterior wall of maxilla removed to reach up to the infratemporal fossa.
- Tumor identified up to its last limit and dissected out.
- Maxillary artery identified and ligated with ligaclips.
- Hemostasis achieved, maxillary sinus cavity and nasal cavity packing was done.

In all cases, the final histopathological examination revealed juvenile angiofibroma.

All patients were followed up and till date no recurrences were noted.

6. Discussion

Endoscopic resection of tumor has many advantages like bone resection is minimized, blood loss is reduced and the

normal growth of the adolescent facial skeleton is not altered significantly (4, 5).

By the endonasal access, the scar and the manipulation of the soft tissues of the face are avoided. The operative time and hospital stay are therefore reduced.

Another advantage of the endoscopic resection of JA is the improved, multiangled and magnified view of the tumor limits which facilitates tumor resection.

The limits for endoscopic resection are extensive extension beyond the boundaries of nasopharyngeal cavity. In these cases mid facial degloving approach can be the modality of choice.

Profuse bleeding, a hallmark of JA resection, can be controlled by the surgeon who has both hands free to use suction and diathermy compered to endoscopic technique.

It can avoid the external scar of a lateral rhinotomy approach and still provide desirable access to the tumor.

The morbidity of the external approach in the treatment of JA is well documented. Blood loss due to extensive osteotomies may necessitate blood transfusion and increase the operative time.

CSF leak, facial and infraorbital nerve damage, lacrimal dysfunction, facial deformities and dental malocclusion have been reported with transfacial approach (3).

The recurrence rate for endonasal endoscopic approach (10%) is lower than the values reported for external surgical approach (14.5%). Thus the endoscopic technique is at least as good as the external technique regarding the recurrence rate (5).

7. Conclusion

Preoperative detailed radioimaging techniques help in planning surgical approach. Endoscopic approach is an evolving treatment modality of JA, it permits less post - operative morbidity; improve visualization compared to traditional open methods and lower recurrence. In addition to avoiding external scar, midfacial degloving approach provides ample access to the tumor and space for surgical manipulation.

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