Case Report

JUVENILE NASOPHARYNGEAL ANGIOFIBROMA: A CASE REPORT

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ABSTRACT:

Juvenile Nasopharyngeal Angiofibroma is a rare benign neoplasm almost exclusively seen in adolescent males, which proliferates under the influence of steroid hormones at puberty. It arises from the margin of sphenopalatine foramen and spreads widely along the skull base. Histologically, it comprises a hamartomatous tumor of vascular tissue lacking smooth muscle and elastic coats. Its propensity to bleed heavily and extensive spread along the skull base and cranial fossae pose significant risk of hemorrhage during surgical resection. Various techniques are employed before and during surgery to reduce the risk of per-operative hemorrhage. Advances in interventional radiology such as pre-operative embolization of feeder vessels enable safer resection. We present a case of a 15-year old male admitted with triad of nasal obstruction, recurrent spontaneous epistaxis and fleshy mass in right nasal cavity. HRCT paranasal sinuses revealed a large nasopharyngeal mass infiltrating the nasal cavity, the nasopharynx, sphenoid sinus, and bilateral posterior ethmoidal cells, extending up to the infratemporal fossa. Cross-sectional imaging enabled accurate staging and open surgical technique was opted. Pre-operative embolization of internal maxillary artery allowed successful complete surgical resection with minimal bleeding per-operatively. With accurate staging and correct choice of hemostatic technique, angiofibroma can be excised completely with relatively less morbidity

Key Words: Angiofibroma, Interventional Radiology, Epistaxis, Head and Neck Neoplasm

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NTRODUCTION

Juvenile Nasopharyngeal Angiofibroma (JNA) is a rare androgen-dependent locally aggressive benign tumor accounting for less than 1% of head and neck neoplasms. It is almost exclusively seen in males with mean age of onset between 13 and 22 years,[1] hypothesized to arise from the surge in steroid hormone at puberty.[2] JNA tumors have been postulated to originate from the margin of sphenopalatine foramen, spreading submucosally along the skull base and cranial fossae with 4-11% of cases being associated with intracranial invasion.[3] Histologically, it is a lobulated, non-encapsulated tumor arising from hamartomatous nidus of vascular tissue.[4] Over 10 staging systems having been proposed based on cross-sectional imaging. Coronal computed tomography shows extent of bony destruction whereas, magnetic resonance imaging highlights soft tissue involvement pre-operatively and residual or recurrent disease post-operatively. JNA is managed definitively by surgical resection involving endoscopic resection, open surgical resection, or a combination of the two. Radiotherapy is reserved for residual or inoperable cases such as those involving internal carotid artery.[5] Recent advances in interventional radiology have allowed pre-operative embolization of feeder vessels to reduce blood loss during resection. JNA remains a surgical challenge,

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especially in developing countries where embolization facilities are not available ubiquitously. Here we present a case of a 15-year old male having JNA with intracranial extension treated by pre-operative embolization and surgical resection at a tertiary care hospital of Lahore.

CASE DESCRIPTION

A 15-year-old male was admitted in Otorhinolaryngology Department of Jinnah Hospital, Lahore in September 2024 with chief complaint of nasal obstruction and recurrent per nasal bleed for 6 months. Nasal obstruction was gradual in onset, constant, slowly progressing from unilateral (right) to bilateral, not relieved by topical or oral medication. It was associated with occasional serous discharge and no air entry. For the preceding 3 months, it was associated with progressively increasing mouth breathing, snoring and development of nasal speech. The patient also complained of multiple episodes of profuse spontaneous epistaxis comprising fresh blood clots, relieved by topical medication and application of cold water. In the preceding one month, he developed progressive swelling of right cheek and right orbit. Systemic inquiry revealed occasional episodes of ipsilateral hearing loss, otalgia and cough. He also reported progressive post-nasal dripping, anosmia, broadening of nasal bridge and widening of sclera. However, he had no complaints of decreased visual acuity, diplopia or ophthalmoplegia. He had no noteworthy past medical history, family history or drug history. General physical examination revealed pallor and S-shaped external nasal deformity. Anterior rhinoscopy revealed mucoid discharge and a red fleshy mass filling the right nasal cavity. He also had swelling of right cheek with normal overlying skin. There was mild proptosis of right eye. However, visual acuity was 6/6 bilaterally and all eye movements were full in range. Oro-dental and neurological examination was unremarkable, with intact cranial nerve function. Routine hematological investigations were within normal range except decreased hemoglobin level (10.5 g/dl) (Table 1).

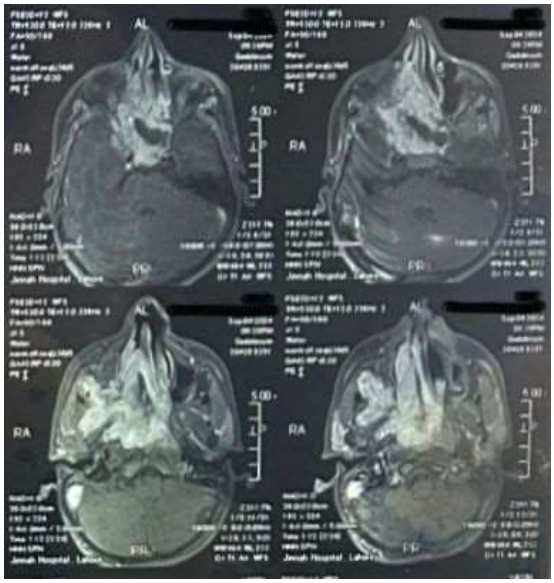
Table 1. Results of baseline investigations performed prior to open surgical resection under general anesthesia

Laboratory Parameter	Value
Hemoglobin (g/dl)	10.5
Total Leukocyte Count (x10^9/L	8.0
Platelet Count (x10^9/L)	303
Hematocrit (%)	33.4
Urea (mg/dl)	13.97
Creatinine (mg/dl)	0.54
Bilirubin total (mg/dl)	0.5
ALT (U/L)	24
AST (U/L)	29
Sodium (mmol/L)	132
Potassium (mmol/L)	4.78

HRCT paranasal sinuses revealed a large sinonasal mass lesion involving posterior portion of both nasal cavities and ethmoidal air cells, expanding the sinuses with a large nasopharyngeal component. There was compression of the right osteomeatal complex with fluid impaction in the right maxillary sinus. Left maxillary sinus and bilateral frontal sinuses were unremarkable. MRI face with contrast showed soft tissue mass lesion measuring 7.5 x 6.5 x 6.5 cm (craniocaudal anteroposterior transverse) centered in the right sphenopalatine foramen, infiltrating right nasal cavity, nasopharynx, sphenoid sinus, bilateral posterior ethmoidal cells, right pterygopalatine fossa and right infratemporal fossa (Figure 1). Right masticator space was also invaded. Mass involved the orbital apex closely abutting the right optic nerve, extending into the middle cranial fossa (intracranial, extra-axial),

contacting the cavernous sinus. Provisional diagnosis of nasopharyngeal angiofibroma (Andrew-Fisch Stage 3B) was made.

Figure 1. Cross-sectional imaging showing extent of sinonasal mass



Patient underwent digital subtraction angiography of feeder vessels and pre-surgical embolization under general anesthesia at Lahore General Hospital (Figure 2). Bilateral feeder vessels from both branches of internal maxillary artery were embolized with spongostan. Recovery was uneventful and patient was prepared for surgical resection within the following 24 hours.

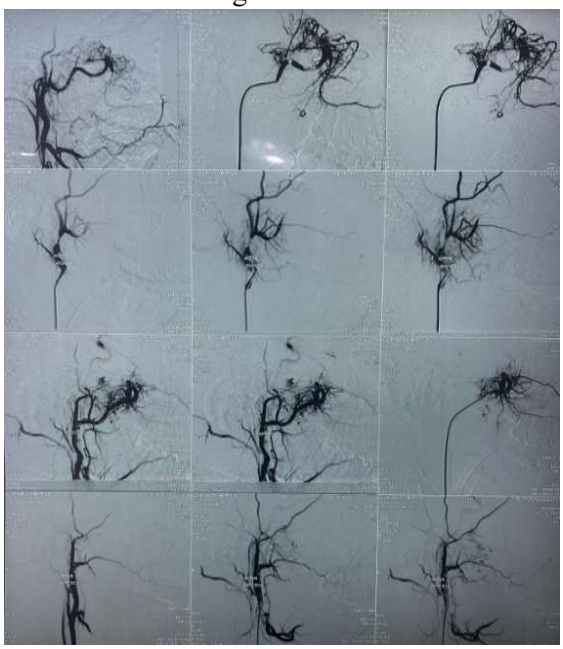


Figure 2. Pre-operative digital subtraction angiography performed at a tertiary care hospital of Lahore to identify feeder vessels for embolization prior to open surgical resection of angiofibroma

Open surgical resection was performed under general anesthesia in reverse Trendelenburg position with 15-degree head elevation and intra-oral delivery of endotracheal tube. Lateral rhinotomy incision was given and right medial maxillectomy was performed. Intra-operatively, the tumor was found to involve nasopharynx and sphenoid sinus, extending up to right infratemporal fossa. Posterior nasal plug was placed followed by anterior nasal packing. On 2nd post-operative day, the nasal pack was removed under general anesthesia.

Pre-operative and immediate post-operative hemoglobin levels were 10.8 and 9.5 g/dl, respectively. Two pints of blood were transfused post-operatively to achieve baseline hemoglobin levels within first two post-operative days. Recovery was unremarkable. Patient developed mild swelling of right cheek and right eye following surgery and was managed successfully on intravenous dexamethasone. Neurological function and visual acuity remained intact post-operatively. Histopathological examination of resected specimen revealed proliferating vessels in a background of fibrous and collagenized stroma, thus confirming the diagnosis of JNA. Patient was discharged on sixth post-operative day and follow-up nasal endoscopic examination revealed no residual disease.

DISCUSSION

Juvenile Nasopharyngeal Angiofibroma (JNA) is a benign locally aggressive tumor of the head and neck seen almost exclusively in adolescent males. It accounts for 0.05 to 0.5% of head and neck neoplasms[6]. Mean age of presentation ranges from 13 to 22 years.[1] Immunohistochemical staining has identified its receptor status as androgen positive thus suggesting androgen influence on the origin of this tumor.[7] Patient usually presents with a triad of nasal obstruction, recurrent spontaneous epistaxis and a mass in the nasal cavity. Erosion

of orbit and intracranial involvement may cause proptosis and cranial nerve palsies. Advanced stages result in anosmia, eye pain and Eustachian tube blockage leading to recurrent otitis media. Our case was a 15-year-old adolescent male who presented with typical signs and symptoms of JNA in accordance with previous literature. Our differential diagnoses included antrochoanal polyp, nasopharyngeal cyst, sinonasal malignancy and granulomatous diseases of nose.

JNA tumors arise from margin of sphenopalatine foramen. Acharya et Al. has identified its precise location of origin as trifurcation of the sphenoidal process of palatine bone, roof of pterygoid process and horizontal process of vomer[4]. It is known to spread along every axis; superiorly into sphenoid and cavernous sinuses, medially pushing the nasal septum, posteriorly invading the basisphenoid and laterally extending into the pterygomaxillary and infratemporal fossae. 10-20% of JNAs possess the potential to extend intracranial and into the orbit through the infraorbital fissure; however, dural invasion is rare.[8]

Cross-sectional imaging has a valuable rule in diagnosis and staging. Coronal computed tomography (CT) shows extent of bony destruction and anatomical location of the tumor. Lateral expansion of the tumor results in bowing of the posterior wall of ipsilateral maxillary sinus, which is known as the Holman Miller sign - pathognomonic for angiofibroma. However, magnetic resonance imaging (MRI) is superior to CT in delineating soft tissue and intracranial involvement. The size of this tumor as seen on cross-sectional imaging may not be an accurate estimate of the actual size. Acharya et al. has described the tumor as "only tip of the iceberg."[4] Over 10 staging systems have been devised based on cross-sectional imaging, of which Radkowski and Andrew-Fisch are commonly used (Table 2).

Table 2. Radkowski and Andrew staging systems in use for Juvenile Nasopharyngeal Angiofibroma

Stage	Radkowski[9]	Andrew[9]
1	**A=Limited to nose or nasopharyngeal vault B=Extension into one or more sinuses**	Limited to nose or nasopharyngeal vault
2	**A=Minimal extension into pterygopalatine fossa B=Involvement of entire pterygopalatine fossa C=Extension into infratemporal fossa or posterior to pterygoid plates**	Invasion of pterygopalatine fossa or any sinus
3	**A=Minimal involvement of middle cranial fossa or pterygoid plates B=Intracranial extension with or without cavernous sinus invasion**	A=Extension in infratemporal fossa or orbital invasion B=Intracranial extradural extension
4	-	Intra-dural extension

In this case, both HRCT paranasal sinuses and MRI face were done and extent of the tumor was delineated. The nasopharyngeal mass was large, infiltrating the nasal cavity, nasopharynx, sphenoid sinus, and bilateral posterior ethmoidal cells, extending up to the infratemporal fossa. Andrew-Fisch staging was used and the tumor was classified as stage 3b.
Treatment options include nasal endoscopic, endoscopic-assisted and open surgical resection. Open surgical approaches based on the anatomical location and extent of tumor include lateral rhinotomy, transpalatal, transmaxillary, mid facial degloving, Le Fort I-

III, Denker, infratemporal and various combinations of approaches. Radiotherapy is reserved for recurrent, residual or inoperable cases such as those having intracranial extension. Open surgical technique was thus preferred over endoscopic technique in our case to allow maximal resection.

Histologically, nasopharyngeal angiofibroma comprises a lobulated non-encapsulated mass of stellate and staghorn blood vessels in loose fibrous stroma[4]. These blood vessels lack elastic or smooth muscle coat, thus accounting for excessive bleeding during surgery. The propensity to bleed heavily and ability to spread along cranial fossae and base of skull make the tumor surgically challenging.[10] Biopsy of the resected specimen confirmed the diagnosis of our case. Network of blood vessels in fibrous stroma was identified on histopathology, in line with previous case report.[4]

Various techniques may be employed pre-operatively and intra-operatively to reduce the risk of hemorrhage during surgical resection, such as ligature of internal maxillary artery and pre-operative embolization of feeder vessels[11]. The tumor is usually supplied by branches of internal maxillary artery and occasionally by other branches of external carotid artery. Feeder vessels can be identified pre operatively via digital subtraction angiography. Pre-operative embolization of feeder vessels from external carotid allows surgical resection with relatively less bleeding. Blood loss in non-embolized patients is reduced to almost half in embolized patients[11]. JNA recurrence in embolized patients has also been reported to be lower than in non-embolized patients.[12] However, surgical resection must be done within 48-72 hours of embolization before revascularization from contralateral vessels occurs.

JNA is a highly vascular tumor. As surgical manipulation carries high risk of severe bleeding intra-operatively, multi-disciplinary approach was undertaken by otorhinolaryngologists and interventional radiologists for optimal management of this case. Pre-operative digital subtraction angiography and embolization of feeder vessels permitted surgical resection with minimal blood loss and patient's hemoglobin was optimized within 2 days following surgery. Nasal packing removed under general anesthesia on 2nd post-operative day revealed minimal bleeding.

Our case had certain limitations. Pre-operatively, the patient had to wait for embolization facilities to be available, which was also only possible at another tertiary care center of Lahore. The patient returned to out-patient department for follow-up only once and could not be followed any further.

CONCLUSION

Juvenile nasopharyngeal angiofibroma is a surgical challenge for otorhinolaryngologists, owing to its intense vascularity and complex anatomical location. However, cross-sectional imaging allows accurate staging of the tumor and hence, the correct choice of surgical technique. It is mandatory to employ techniques to reduce per-operative hemorrhage. Radiological advancements such as digital subtraction angiography and pre-operative embolization allow safer resection with better surgical outcome.

AUTHORS' CONTRIBUTION

NZ: Introduction, Case Description, Discussion, and Abstract
MT: Case Description, Review, and References

CONSENT FOR PUBLICATION

Written informed consent was taken from patient and his guardians regarding data collection and publication.

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CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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