Case Report

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Surgical Prognostic Factors for Juvenile Nasopharyngeal Angiofibroma

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Abstract

Background: Juvenile Nasopharyngeal Angiofibroma (JNA) are benign, rare and highly vascular tumors of nose and para nasal sinuses seen only in post pubertal males.

Aim: To study surgical prognostic factors for JNA.

Objectives: To study the effect of presenting features namely blood supply, blood loss, tumor grading, steroid therapy and age of presentation on surgical prognosis.

Material: Presenting features and surgical outcomes of all 4 operated JNA patients were compared from the Departmental records.

Result: Unilateral blood supply, Lesser pre-operative blood loss, lesser grade tumor, longer steroid therapy and more age at presentation have better surgical outcome.

Conclusion: Although benign and rare to encounter, their diagnosis and management is a challenge owing to their highly vascular nature and their blood supply directly from or from main branches of carotid arteries. Here, we present four cases of successful management of JNA with detailed discussion of surgical prognostic factors of the rare tumor.

Keywords: Juvenile nasopharyngeal angiofibroma JNA vascular tumor surgery prognosis.

INTRODUCTION AND REVIEW OF LITERATURE

JNA is a non-encapsulated highly vascular benign tumor found in post pubertal male adolescents presenting predominantly with aggressive nasal and oral bleeds. It is formed by abnormal and excessive proliferation of vessel sinusoids which lack the vessel wall and hence non-responsive to vasoconstriction for hemostasis. It is a rare tumor accounting for only 0.05% of head & neck tumors.[1]

Coordinating interprofessional care is required in the evaluation and treatment to improve patient outcomes. It needs integrated coordinated care by a team of Anesthetist and Critical Care Experts, pediatricians, ENT surgeons, neurosurgeons, pathologists and family medicine with shared responsibilities.[1,2,3] JNA are benign growth derived

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from the vessel sinusoids but they are known for aggressive growth into various compartments of skull base through narrow openings. Few cases have shown bony erosions of walls of nasal cavity, sinuses and pterygoid plates.[4]

JNA is derived from large vessel sinusoids and hence get direct blood supply from carotids and its branches. The most common tributary is Internal Maxillary Artery (IMA).[5] There may be n tributaries with direct contribution from carotid trunk. Ascending Pharyngeal Artery (APA) is another key player. Any branches of both the carotid arteries or the carotid itself may form its vascular pedicle. Opthalmic Artery and Vidian Artery are commonly contributing branches of Carotid Artery.[6]

The etiology of JNA is multifactorial. One of the factors is an arteriovenous malformation during embryogenesis which grows later in life in presence of high testosterone levels. The other factor is persistence of first branchial arch remanent in post-natal life.[2] This remanent theory also explains the anatomical formation and enlargement of JNA. The other factors are release of vascular endothelial growth factor-2. The tumor growth is supported by absence of female sex hormone Estrogen.

Grossly, JNA is a mass of sinusoids held together in fibrous tissue lacking a true capsule. The forming sinusoids are of diverse shape, size and caliber (*Figure 1.1*). Sinusoids are the thin-walled vascular spaces lined by a single layer of endothelium. So, it is a friable tumor.[7] Biopsy is contra-indicated for similar reasons. These masses bleed massively even without provocation and bleed more on any manipulation.[1]

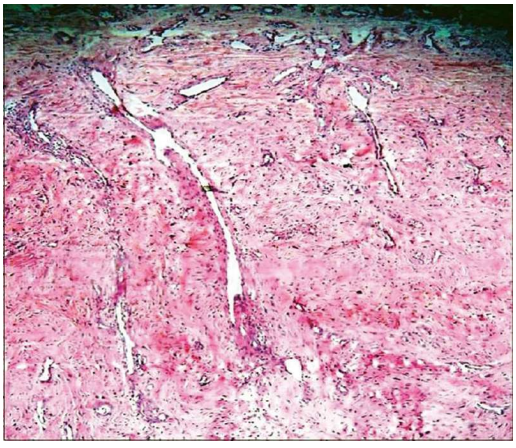


Fig. 1: H&E staining and observation under 100X microscope shows large sinusoids varying in size, ranging from slit-like to ectatic with a staghorn configuration, poorly developed myoid-type cells surround the endothelial-lined vascular channels with absence of elastic lamina

JNA is a rare tumour found usually in post pubertal male adolescents. Very rarely, these are reported after the age of 40 years in males. All JNA patients present with massive, recurrent and spontaneous nasal bleeds. 60-90% of patient complaint of nasal obstruction (bilateral>unilateral) which may be associated with de-nasal speech, mouth breathing and snoring. 10-70% of patients have conductive hearing loss and otitis media with effusion (OME), 0-70% of patients have broadening of nasal bridge, proptosis and swelling of cheek (frog face deformity) and 0-40% of patients have reduced vision or blindness due to involvement of optic nerve/optic tract and optic chiasma, reduced eye movements and ptosis due to involvement of 3rd,4th and 6th cranial nerves, loss of corneal reflex due to involvement of 5th cranial nerve. 10-70% of patients have conductive hearing loss and OME. 0-70% of patients have broadening of nasal bridge, proptosis and swelling of cheek (frog face deformity). 0-40% of patients have reduced vision or blindness due to involvement of optic nerve/optic tract and optic chiasma, reduced eye movements and ptosis due to involvement of 3rd,4th and 6th cranial nerves, loss of corneal reflex due to involvement of 5th cranial nerve.10-70% of patients have conductive hearing loss and OME. 0-70% of patients have broadening of nasal bridge, proptosis and swelling of cheek (frog face deformity). 0-40% of patients have reduced vision or blindness due to involvement of optic nerve/optic tract and optic chiasma, reduced eye movements and ptosis due to involvement of 3rd,4th and 6th cranial nerves, loss of corneal reflex due to involvement of 5th cranial nerve.

AIM

To study surgical prognostic factors for JNA.

OBJECTIVES

To study the effect of presenting features namely blood supply, blood loss, tumour grading, steroid therapy and age of presentation.

CASE PRESENTATION

Here we present four cases of JNA. All presented in outpatient department. All were male patients in age group 12-27 years. Excessive repeated nasal bleeding was the presenting complaint in all. Three patients complained of nasal obstruction also. One patient also complained about nasal discharge and

nasal twang in voice. One patient also complained of snoring.

Table 1: Clinical presentation and radiological staging of patients presenting with JNA

S. No.	Age/Sex	Steroid Therapy Duration (Days)	Andrews-Fisch Stage
1	27 Male	0	3a
2	18 Male	14	3a
3	12 Male	7	2
4	15 Male	14	3a

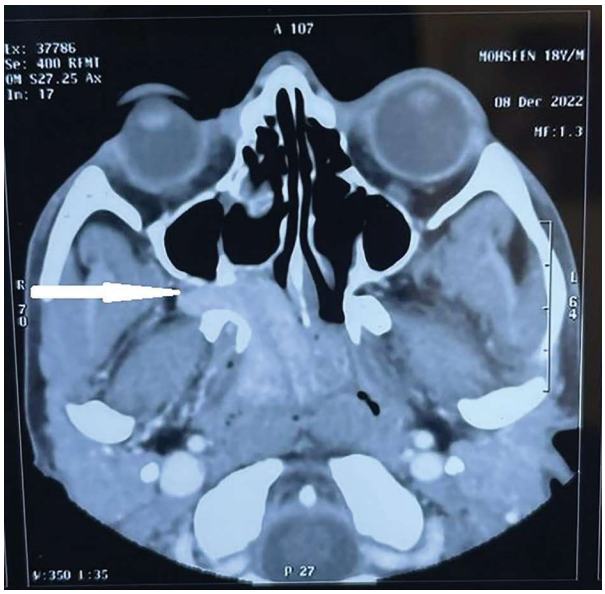


Fig. 2: Computerised tomography image showing juvenile nasopharyngeal angiofibroma (white arrow) completely blocking the nasopharynx and extending throughright pterygo- maxillary fissure into the right infratemporal fossa

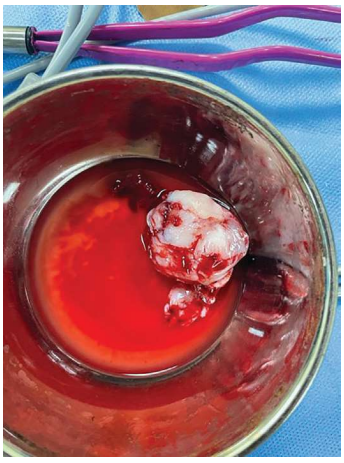


Fig. 3: Excised juvenile nasopharyngeal angiofibroma revealing its fibro-sinusoidal architecture, extensions into various skull base compartments and vascular pedicle

RESULTS

The factors favouring better surgical outcome are

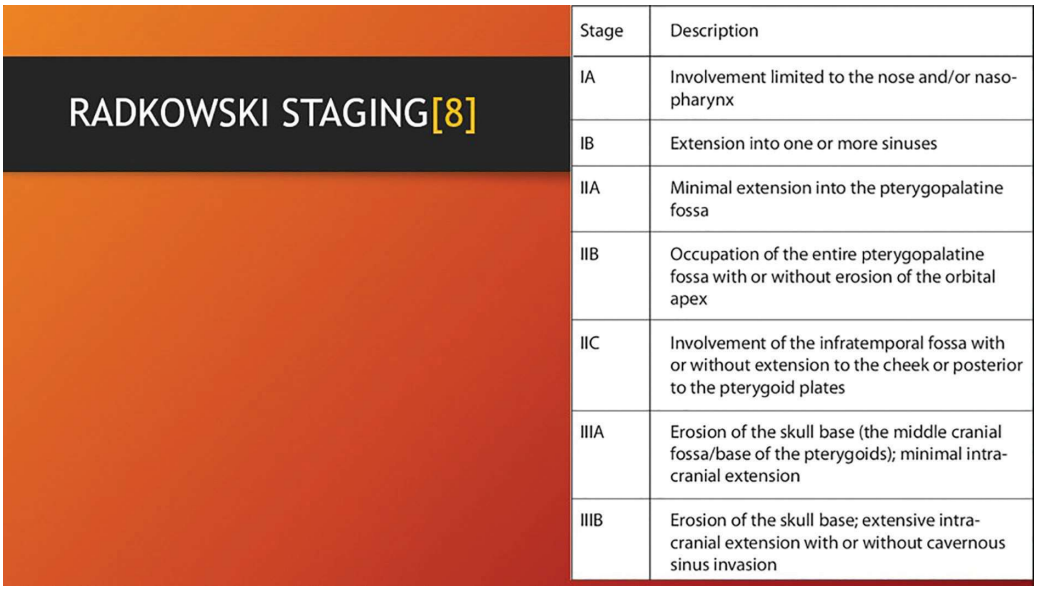
1. Unilateral blood supply
2. Lesser pre-operative blood loss
3. Lesser grade tumour
4. Longer steroid therapy
5. More age at presentation

DUSCUSSION

Various differentials of nasopharyngeal mass include

- Adenoid hypertrophy
- Antrochoanal polyp
- Squamous papilloma
- Thornwaldt's cyst
- Pleomorphic adenoma
- Craniopharyngioma
- Paraglioma
- Hamartoma
- Nasopharyngeal carcinoma
- Lymphoma
- Rhabdomyosarcoma
- Chordoma
- Plasmacytoma
- Hemangiopericytoma
- Malignant salivary gland tumours
- Melanoma

History and Examination play most important role in diagnosis.Direct nasal endoscopy is the mainstay of diagnosis. Slightest touching and manipulation of tumour is avoided.CECT Nose and paranasal sinuses is the investigation of choice. MRI is complementary to see soft tissue extensions and to rule intra-cranial, orbital and infra-temporal extension.X ray may show anterior bowing of posterior wall of maxillary sinus in X ray water's view and CT scans if there is infra-temporal extension (Holmen Miller's sign). Digital subtraction carotid angiography provides opportunity for super-selective embolization of the tumour.Tumor grading systems are very helpful in surgical planning, prognostication and treatment monitoring. Here, we have discussed two grading systems which are widely used by Rhinologists all over the world.[5]



Stage	Description
IA	Involvement limited to the nose and/or naso-pharynx
IB	Extension into one or more sinuses
IIA	Minimal extension into the pterygopalatine fossa
IIB	Occupation of the entire pterygopalatine fossa with or without erosion of the orbital apex
IIC	Involvement of the infratemporal fossa with or without extension to the cheek or posterior to the pterygoid plates
IIIA	Erosion of the skull base (the middle cranial fossa/base of the pterygoids); minimal intra-cranial extension
IIIB	Erosion of the skull base; extensive intra-cranial extension with or without cavernous sinus invasion

Fig. 4: Radkowski staging is a very popular staging system of JNA based on computerised tomography

Type	Description
I	Tumor limited to nasopharynx and nasal cavity; bone destruction negligible or limited to the sphenopalatine foramen
II	Tumor invading pterygopalatine fossa or maxillary, ethmoid, or sphenoid sinus with bone destruction
IIIa	Tumor invading infratemporal fossa or orbital region without intracranial involvement
IIIb	Tumor invading infratemporal fossa or orbit with intracranial extradural (parasellar) involvement
IVa	Intracranial intradural tumor without infiltration of cavernous sinus, pituitary fossa, or optic chiasm
IVb	Intracranial intradural tumor with infiltration of cavernous sinus, pituitary fossa, or optic chiasm

From Andrews JC, Fisch U, Valavanis A, Aeppli U, Makek MS: The surgical management of extensive nasopharyngeal angiofibromas with the infratemporal fossa approach. Laryngoscope 99:429-37, 1989.

Fig. 5: Andrew Fisch[9,10] Classification is another popular staging system of JNA based on computerised tomography. We have also used the same system for staging in present paper

Surgical excision is treatment of choice. Where facilities are available, Interventional Radiologist should carry pre-operative super-selective embolization of major feeding vessels while avoiding any collateral damage under live digital subtraction angiography (DSA) and imaging. It reduces the intra operative blood loss and hemorrhagic complications. It identifies the feeding vessels and provide a pavement for the segmental removal of tumor. Seldinger technique may be used for femoral assess to carotids. DSA is performed with a 5F catheter of the carotid system. The major supplying branches are further identified, sub-selected and injected with 3F catheter. Micro coils, sponge, gelatin and Hydroxyapatite crystals may be injected for this purpose.Non target embolization of ICA is avoided Target embolization also offers branch selectivity. If ICA and/or Opthalmic Artery is embolized, vision loss may be there. Cranial injuries may be seen with non-selective embolization of viable branches of carotid system. Side-effects of embolization include facial swelling, pain or abnormal sensation, headache, or nausea/ vomiting. JNA have variable composition of sinusoids and fibrous tissue which is difficult to assess correctly pre-operatively even after digital subtraction angiography because blood enter sinusoids by multiple mechanisms including direct, indirect and reverse blood flow, active and passive mechanisms. More the proportion of vascular tissue, more is the risk of bleeding.[11,12] Some centers specialized for JNA Excision use cell savers. Cell savers are special devices through which suctioned blood is passed to restore red blood cells, leucocytes and platelets which are continuously re-transfused into circulation throughout the surgery.[11]

Surgical approaches for JNA excision

- Endoscopic
- Trans Palate
- Trans-Nasal
- Trans-Palate + Trans-Nasal
- Trans- Palate + Sublabial (Sardana's)
- Lateral rhinotomy
- Medial maxillectomy
- Facial degloving
- Danker's approach
- Trans-Maxillary (Le Fort's)
- Maxillary swing
- Facial translocation (Wei's)
- Infra-temporal
- Intracranial-extracranial

The most dreaded complication of JNA is blood loss and its sequalae. Less than 1% of JNA have bilateral direct carotid involvement. In such cases, even if pre-operative embolization is done only limited embolization is possible as embolization of multiple vessels particularly internal carotid artery increases risk of stroke, multiple cranial nerve palsies, paralysis and death. Even external carotid artery ligation is not much useful in these cases. One-third tumors of Radkowski grade 3 are unresectable. Even those are resected, one-third of them shows recurrence.[1]

CONCLUSION

Higher stage tumours in younger patients indicate rapid rate of growth and carry high risk of morbidity and mortality. More blood loss in pre operative period signifies high vascularity of tumour and carries more risk of morbidity and mortality.A high-grade tumour may not comply with complete resection or may require a staged approach and has a higher recurrence rate. A pre-operative 14 days course of steroids reduces vascularity of tumor and decreases intra-operative blood loss, morbidity and mortality. Carotid digital subtraction angiography should be done pre-operatively in all patients to rule out bilateral blood supply of tumor irrespective of the plan of embolization.

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Ethics declaration: NIL

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