Review Article

Juvenile Nasal Angiofibroma (JNA) - Review of Management with a Special Focus on Surgical Approach and Resection

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Abstract

Juvenile Nasal Angiofibroma is a condition which almost exclusively occurs in young males who present with features of nasal obstruction and epistaxis. Over the past few decades there has been a paradigm shift in how we manage this condition with changes in diagnostic criteria, imaging modalities, staging as well as surgical approaches. Endoscopic and Endoscopic Assisted resection techniques have been increasingly popular over the past two decades and have shown to decrease surgical time, post-operative stay as well as intraoperative blood loss.

It is our recommendation that future surgeons be adequately exposed and trained in endoscopic resection techniques of angiofibroma, as the authors believe, given the data at hand that this is the way forward.

Key words: Juvenile nasopharyngeal angiofibroma, Endoscopy, Surgery


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Introduction

Juvenile Nasal Angiofibroma (JNA) is a relatively uncommon lesion that mainly present in adolescent male children in the form of nasal obstruction and sudden epistaxis. Analysis of epidemiological data suggest that it accounts for around 0.5% of all head and neck tumours with general incidence of 1:150,000. Other clinical features at presentation include facial swelling, visual and neurological disturbances and even conductive hearing loss. Although transnasal biopsy is now relatively contraindicated investigations such as MRI (Magnetic resonance imaging) with MRA (Magnetic resonance angiography) and CT (Computer tomography) of the nose and paranasal sinus will enable you to make the diagnosis radiologically, stage it and plan surgical resection accordingly. The commonest staging system developed for this lesion was introduced by Radkowski et al which classifies it into 3 distinctive groups.

Surgical resection with or without embolization of the internal maxillary artery is the preferred method of treatment. The approach to resection largely depends on the staging of the lesion, with endoscopic approaches favouring early stage disease and more radical open approaches (lateral rhinotomy approach and maxillary swing approach) favouring more advanced stages. There have been reports of endoscopic resection of advanced tumours but, this is more the ‘exception’ than the rule. Along with endoscopic resection, there has been an increase in usage of Coblation technology to resect these lesions. Coblation gives the advantage of better visualization of the lesion during endoscopic resection and debulking. This has a knock on effect of reducing surgical time which itself is an added advantage since endoscopic resection shortens surgical time compared to an open approach in experienced hands.

Clinical features

Classically these patients present with nasal obstruction (commonly progressive and thus rarely noticed till it is clinically proven) and epistaxis in a young male. A great majority of the patients being young adolescents. With the clinical suspicion nasal endoscopy is performed and a mass in the nasal cavity particularly in the posterior aspect with characteristic appearance is almost diagnostic.

Investigations

Contrast enhanced computer tomography (CECT) seems to be the investigation of choice. This investigation was also helpful in staging the disease as well as planning the surgical approach and the post-operative evaluation of the patient. Other investigations used were magnetic resonance imaging (MRI) of the brain and postnasal space, magnetic resonance angiography (MRA), and contrast angiography. MRI was considered mostly when the tumour involved the skull base and contrast angiography was considered mostly when preoperative embolization of the tumour was chosen.

Biopsy of the tumour is now relatively contraindicated as this may lead to uncontrollable bleeding and the diagnosis is clinched on the trifecta of demography, clinical features, and radiological investigations.
Clinical Staging

Multiple staging systems are available. The commonest being used are the Andrews classification (Also known as the Modified Fisch Classification) and the Radkowski staging system. A new classification system called the UPMC Staging system (The University of Pittsburgh Medical Center staging system) is also available which apparently has a higher correlation with intraoperative blood loss and the need for recurrent surgeries. (Refer Table 1) According to this classification, stages four and five were associated with the blood loss of greater than 1 litre.

<table>
<thead>
<tr>
<th>Stage</th>
<th>The University of Pittsburgh Medical Center (UPMC) staging system</th>
</tr>
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</table>
| I     | Nasal cavity  
Medial pterygopalatine fossa  
No residual vascularity after preoperative embolization of external carotid blood supply |
| II    | Paranasal sinuses  
Lateral pterygopalatine fossa after preoperative embolization of external carotid blood supply  
No residual vascularity |
| III   | Skull base, orbit, Infratemporal fossa  
No residual vascularity after preoperative embolization of external carotid blood supply |
| IV    | Skull base, orbit, Infratemporal fossa  
With residual vascularity after preoperative embolization of external carotid blood supply |
| V     | Intracranial extension:  
medial (medial cavernous sinus) and lateral (middle fossa) routes of extension  
With residual vascularity after preoperative embolization of external carotid blood supply |

Table 1: The University of Pittsburgh Medical Center (UPMC) staging system

<table>
<thead>
<tr>
<th>Stage</th>
<th>The Andrews / Modified Fisch staging system</th>
</tr>
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<tbody>
<tr>
<td>I</td>
<td>Limited to the nasopharynx and nasal cavity. Bone destruction negligible or limited to the sphenopalatine foramen</td>
</tr>
<tr>
<td>II</td>
<td>Invading the pterygopalatine fossa or the maxillary, ethmoid, or sphenoid sinus with bone destruction</td>
</tr>
</tbody>
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| III   | Invading the infratemporal fossa or orbital region without intracranial involvement  
Invading the infratemporal fossa or orbit with intracranial extradural (parasellar) involvement |
| IV    | Intracranial intradural without infiltration of the cavernous sinus, pituitary fossa, or optic chiasm  
Intracranial intradural with infiltration of the cavernous sinus, pituitary fossa, or optic chiasm |

Table 2: The Andrews / Modified Fisch staging system
Treatment

Adjuncts prior to surgical resection

Preoperative embolization was not considered in certain studies claiming that it increases the risk of bleeding during surgery. It was also suggested that it increases the risk of leaving a residue during surgery but there is little evidence to support this. Usually this is done 24 to 48Hrs prior to resection surgery.

Usage of vessel loops around the external carotid artery to facilitate ligation of the vessel in case of emergency has also been advocated. However, the authors believe that this option should be carefully considered if a completely endoscopic resection is planned as it defeats the purpose of minimal access surgery in addition to causing a cosmetic defect. Further, the usefulness of the intervention in terms of reducing the amount of bleeding is also doubtful.

Anaesthesia

General anaesthesia was preferred with endotracheal intubation, throat packing and the patient positioning usually being a reversed Trendelenburg berg position with a 30° angulation. Fibre optic intubation techniques have been described when dealing with these cases as well. Rapid sequence induction was considered in actively bleeding cases. Non-invasive as well as invasive intraoperative active monitoring techniques were employed in most cases and patients were maintained at a central venous pressure (CVP) of 10 cm of water. Hypotensive anaesthesia was maintained targeting a mean arterial blood pressure of around 60 to 65 mmHg. Vasodilators, beta-blockers, and Inhalation anaesthetics were utilised to this regard. Depending on the length of surgery and the propensity for bleeding, delayed extubation in an intensive care setup with laryngoscope in guided oral suction has been described.

Surgical resection

Site preparation

Nasal cavity preparation was done with 1:100,000 adrenaline and xylometazoline solution and transnasal infiltration of 2% lignocaine and 1: 100,000 adrenaline was administered around the tumour site followed by warm saline irrigation intermittently during the surgery was mentioned in studies. The role of warm saline was to facilitate vasoconstriction.

General Technique

Some techniques describe blunt dissection around the tumour to identify tissue planes and establishing the origins of the tumour. Others describe a primary objective of locating the feeding artery and gaining ‘lateral’ control before starting the bulk of the dissection which is also very logical.
**Endoscopic Resection**

The approach depends on the staging of the tumour. With early stage tumours requiring a wide antrostomy and ethmoidectomy for initially approaching the tumour and later stages requiring a modified Denker’s approach\(^{14}\) in addition to this. Removal of the posterior wall of the maxillary sinus exposing the infratemporal fossa and the maxillary artery is then described. Advanced stage tumours require in addition to the above, removal of the antero-lateral wall of the sphenoid sinus exposing the vidian canal, quadrangular space and the internal carotid artery\(^2\). Posterior septotomy was also described to increase instrument access\(^2\). Some authors also describe performing a ‘Caldwell-luc’ surgery to increase access and gain lateral control \(^{10}\).

Along with the endoscope Coblation technology has also been used successfully in treatment of angiofibromas\(^8\). This technology relies on a molecular level of disintegration of tissue planes with minimal collateral damage. Studies have also indicated the use of navigation technology further improves the outcome when used in combination with the endoscope\(^{15}\).

Endoscopic resection is currently indicated for small and intermediate size juvenile angiofibromas and selective advanced lesions with the caveat that it be performed by experienced users \(^9\).

**Open resection**

Multiple approaches have been described including the Midfacial degloving approach, The Lateral rhinotomy approach, The Transpalatal approach, The Weber-Ferguson incision with maxillary swing approach and the transcranial approach combined with Diffenbach’s modification of Weber Ferguson incision\(^2\). However, the approaches described above are associated with a higher rate of morbidity and the theoretical risk of affecting facial growth in adolescent males in addition to significant scarring of the face, which itself is not ideal considering the fact that these lesions are essentially benign \(^9\).

These approaches are usually considered for large more advanced tumours and can be combined with endoscopic approaches depending on the user’s experience \(^9\).

**Post-operative care**

At the end of the surgery, postnasal packs as well as anterior nasal packs (endoscopic) were commonly employed to control residual oozing. These packs were generally removed in a theatre setting under anaesthesia depending on the stage and extent of surgery\(^{13}\).

**Radiation therapy**

Radiation therapy has been considered for tumours that were inoperable and in cases where there were recurrences (In both instances total resection was not possible) \(^2\). This is considered especially when critical structures are involved and can be damaged during surgical resection. It is reported to have a limited role in treatment of juvenile angiofibroma \(^9\).

**Follow-up**
The patient is followed up in the OPD setting with routine nasal endoscopy and MRI scanning done at six monthly intervals five years preferably clearly end of the adolescent period ideally. MRI has an added benefit of being sensitive in picking up residual tumour in a background of inflammation.

**Trend analysis**

All stages considered, there was a decrease in operation time and average hospital stay when the endoscope was involved. There was also a significant decrease in intraoperative blood loss across all stages as well. The recurrence rate however did not change significantly. Unsurprisingly the frequency of ‘exclusively’ open surgical approaches have seen a decline over the past 2 decades with an increasing favouritism to endoscopic approaches.

**Outcome Analysis**

There have been reports of spontaneous regression of residual tumour, however this should never be relied upon but is sometimes the basis for a wait and see approach when residual tumour is detected postoperatively.

Interestingly when we consider surgical approaches, it seems that endoscopic approaches are comparable to the open approaches with regard to tumour control with the latter showing higher morbidity. Indeed some early studies and systemic reviews indicated that intraoperative blood loss, length of hospital stay and rate of recurrence was in fact lower when endoscopic removal was considered.

Overall recurrence rates (regardless of approach used) varied from 7% to 45% with higher rates reported in higher stages of disease.

**Conclusions**

Endoscopic resection techniques are fast becoming the gold standard of management of these lesions especially when they detected at a lower stage and smaller size.

Open resection techniques should be known by all concerned and implemented for higher stage and larger lesions. They should be utilized only as a fall-back when dealing with lower stage smaller lesions.

We would like to recommend wider adoption of endoscopic techniques in approaching this condition. These approaches seem to have comparable clinical outcomes with lesser morbidity. Training in open techniques also should be done concomitantly as a fall back in a difficult situation.
References


