Nasopharyngeal Angiofibroma: Paradigm Shift in Management

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Received: 30 March 2016 / Accepted: 31 December 2016 / Published online: 4 January 2017
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Abstract To report on a series of patients of nasopharyngeal angiofibroma of varied ages with different stages and management algorithm which reduced morbidity associated with this tumour. Retrospective. We report a series of ten patients who presented to a tertiary care institution and were diagnosed to have NA from 2012 to 2014. Patients were categorized by Radkowski staging and data was collected to document differences in terms of presentation, operative technique, and postoperative course. All patients underwent preoperative embolization. Stage I and selected stage II lesions were approached endoscopically while the remainder underwent open resection. In comparison with open procedures, endoscopic procedures had less intraoperative blood loss (350 vs. 630 cc), operative time (90 vs. 150 min) and the average hospital stay was one day less (3 vs. 4 days). Proper preoperative work up including nonsurgical intervention in the form of embolisation and selecting proper surgical approach is rewarding in case of angiofibromas of all stages which help to reduce morbidity associated with these benign tumours.

Keywords CT · MRI · DSA

Introduction

Nasopharyngeal angiofibroma (NA) is a rare tumor that comprises only 0.5% of neoplasms in the head and neck [1–3]. It is pathognomonically characterized as a benign vascular tumor located in the posterior nasopharynx of adolescent males. It is the most common benign neoplasm of the nasopharynx. There is a strong predilection for NA to affect teenage males, typically between 14 and 15 years of age, although the reported age range spans between 6 and 35 years of age. The most common presenting symptoms are severe, recurrent epistaxis with persistent nasal obstruction. Surgery remains the definitive treatment modality. The treatment for the nasopharyngeal angiofibroma can present formidable problems for the surgeon. Proper preoperative work up including nonsurgical intervention in the form of embolisation and selecting proper surgical approach is rewarding in case of angiofibromas of all stages which help to reduce morbidity associated with these benign tumours.

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Materials and Methods

The study was conducted in the Department of ENT & HNS Govt Medical College Srinagar & included 10 male patients who were diagnosed as NA from 2012 to 2014. All the patients were evaluated clinically & CECT nose/pns (axial/coronal/sagittal) was done in all. Patients with CECT showing intracranial extension underwent MR imaging to define intracranial findings [4]. All these findings were summed up to stage the tumour according to Radkowski staging system [5].

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Radkowski Staging System

Stage	Details
IA	Limited to nose and/or nasopharyngeal vault
IB	Same as IA with extension into one or more paranasal sinuses
IIA	Minimal extension through the sphenopalatine foramen, including a minimal part of the medial part of the pterygopalatine fossa
IIB	Full occupation of the pterygopalatine fossa, displacing the posterior wall of the maxilla forward; lateral or anterior displacement of the branches of the maxillary artery; superior extension may occur, eroding orbital bones
IIC	Extension through the pterygomaxillary fissure into the cheek and infratemporal fossa or posterior to the pterygoid plates
IIIA	Erosion of the skull base with minimal intracranial extension
IIIB	Erosion of the skull base with extensive intracranial extension with or without cavernous sinus involvement

Digital Subtraction Angiography (DSA) was done by the concerned unit to see the feeding vessels and angioarchitecture of the tumour. Embolization was then carried out by superselective catheterization with Tracker microcatheter system. Distal internal maxillary artery and if necessary ascending palatine branch of facial artery on one or both sides were embolized. For embolization polyvinyl alcohol foam (PVA), gelfoam fine shreds and small pieces were used. Balloon occlusion test was done in the selected cases with intracranial extension into parasellar region. These cases were then operated at the earliest and in no case surgery allowed to extend 4 days after embolization. The peroperative blood loss was measured from suction bottle and soaked gauze packs. Postoperative CT study was done at one month and one year interval (Figs. 1, 2).

Observation

The age limits among patients showed broad spectrum with youngest being 8 years and the eldest 30 years with the average being 18.6 years. Seven patients had stage II disease, two patients had stage III disease and one patient belonged to stage I. Epistaxis was mode of presentation in eight cases who also had associated history of nasal obstruction. One patient presented with rapidly growing cheek swelling and nasal obstruction. Another patient who was being evaluated for headache was diagnosed to have nasopharyngeal angiofibroma as an incidental finding on MRI. In no case any symptom suggestive of intracranial extension was present.

Fig. 1 A patient of nasopharyngeal angiofibroma with facial deformity as presenting feature

Symptomatology of JNA

Symptoms and signs	Present (%)	Absent (%)
Nasal obstruction	90	10
Epistaxis	80	20
Facial swelling	10	90
Headache	10	90
Diminution of vision	10	90

Computed Tomography showed pterygomaxillary fossa to be involved in nine cases. Tumour had extended here through sphenopalatine foramen in six cases and in remaining three cases extension was through the pterygomaxillary fissure. Tumour had eroded the lateral wall of sphenoid sinus and extended intracranially in two patients. Orbital extension via inferior orbital fissure was present in one of these patients. Endoscopic excision of JNA was done in three patients (one of stage I and two of stage II). Larger pterygomaxillary fossa and small infratemporal extensions were removed through a transantral window by removing anterolateral wall and posterior wall of maxillary. Tumor with intracranial extension was removed by combination of transmaxillary subcranial approaches. The defect in skull base was repaired by temporalis fascia and pericranium. The estimated blood loss varied from 300 to

Fig. 2 **a** Coronal CT image of stage III A, **b** coronal MRI image of stage III A, **c** coronal CT image of stage I B

1200 ml (average 630 ml). For endoscopic procedures average blood loss was 350 and 700 ml in open procedures. Average operative time calculated was 90 min in case of endoscopic procedures and 150 min for open procedures. Average hospital stay for endoscopic procedures was 3 and 4 days for open procedures. We followed the patients for 1 year with nasal endoscopy and check CT, recurrence was not seen in any of these ten cases (Figs. 3, 4, 5).

Discussion

Juvenile nasopharyngeal angiofibroma is documented since antiquity by Hippocrates (fifth century BC) [6]. The most common presenting symptoms include the triad of unilateral nasal congestion, nasopharyngeal mass and recurrent epistaxis. In advance stages, nasopharyngeal angiofibroma may present with facial deformity, proptosis, headache and deafness. CECT and MRI have immensely helped in

Fig. 3 Stage IIIA tumor specimen (open procedure)

diagnosing and selecting an appropriate management protocols [7]. The juxtaposition of this benign yet locally invasive mass in an adolescent population mandate aggressive surgical resection while limiting scarring and disruption of midfacial growth potential [8, 9]. NA management has undergone significant evolution over the past decade. There is a shift toward transmaxillary and endoscopic approach. As endoscopic techniques coupled to frameless stereotactic image guidance evolve the limits of transnasal resection continue to be challenged. Though initially utilized for tumors limited to the nasopharynx or paranasal sinuses, these approaches have recently been described for tumors involving the pterygopalatine fossa, infratemporal fossa, and even select lesions with limited intracranial involvement. Endoscopic exposure and

Fig. 4 Stage IA tumor specimen (endoscopic procedure)

excision of nasopharyngeal angiofibroma can be considered as a standard approach for limited lesions extending up to pterygopalatine fossa. Larger pterygomaxillary fossa and small infratemporal extensions were removed through a transantral window by removing anterolateral wall and posterior wall of maxillary antrum. The management of NA with intracranial extension remains controversial despite improvements in neurosurgical techniques. Skull base erosion with minimal intracranial extension does not necessitate craniofacial resection to achieve cure as it is not a malignant tumor and often allows for complete tumour removal from below without formal craniotomy. A good tumour exposure before its removal reduces the tumour removal time and hence the blood loss. In our study wide tumour exposure and incising periosteum around the tumour attachment on skullbase as far as possible and then elevating the healthy periosteum in continuity with the tumour helped in complete and easy removal of tumour. Similar observation was made by Maharaj and Fernandes [10]. In the present study complete tumour removal was achieved in all the operated cases. Preoperation embolization results in less intraoperative blood loss (<1000 ml) and lower subsequent requirement for blood transfusion [11–13]. In our study it was found very effective since the blood loss estimated was 300–1200 ml (average 630 ml). In another series with similar large tumours, where embolization was not done average blood loss was 3000 ml [14]. There are reports of blood loss exceeding 2000 ml even after embolization is also important since gelfoam particles tend to dissolve and disintegrate after 5–7 days, development of collateral circulation after embolization. Preoperative embolization not only reduces blood loss during the surgery but also shortens the operative time (90 min for endoscopic and 150 min for open

Fig. 5 **a** Post operative CT image of stage IIIA tumour, **b** post operative CT image of stage IA tumor

procedures in our study) by allowing the easy identification of surgically important structures, thereby facilitating the tumor removal. The average duration of stay in the hospital was 3 days in cases with exclusive endoscopic removal whereas it was 4 days for the cases who underwent open procedures. Tumor recurrence was not seen in any of our cases after 1 year of follow up. All these changing trends in management of NA has made it a very surgeon friendly tumor and there has been reduction in complications, operative time, length of hospital stay, loss of school/working days and recurrence rates. Tumor recurrence was not seen in any of our cases after 1 year of follow up. Proper preoperative work up including nonsurgical intervention in the form of embolisation and selecting proper surgical approach is rewarding in case of angiofibromas of all stages which help to reduce morbidity associated with these benign tumours.

Conclusion

Nasopharyngeal angiofibroma presents a treatment challenge to surgeons, owing to its high vascularity and high recurrence rates following surgery. Preoperative detailed radioimaging techniques help in planning surgical approach. There has been changing trends toward anterior transfacial and endoscopic approaches. Preoperative embolization reduces the operative blood loss and therefore help in complete tumour removal. Wide exposure of tumour and its removal with a rim of healthy periosteum in nasopharynx also helps in its complete removal and prevents recurrence.

Compliance with Ethical Standards

Conflict of interest None.

Ethical Standards Statement All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent Informed consent was obtained from all individual participants included in the study.

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