Juvenile Nasopharyngeal Angiofibroma: A Rise in Incidence

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Received: 27 April 2015 / Accepted: 8 September 2015 / Published online: 16 September 2015
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Abstract Juvenile nasopharyngeal angiofibroma is a very rare, highly vascular, uncapsulated and locally invasive tumor. Recently a changing trend has been seen in its epidemiology, pathogenesis, diagnosis, medical management, pre-operative care, surgical management and post-operative care including radiotherapy. Study the changing trend basically in the north-eastern region of south-east Asia and its prospects. Retrospective study of patients presenting with JNA between the study period of 6 months (October 2014–March 2015) in the, Department of Otolaryngology and Head and Neck Surgery, Assam Medical College, Dibrugarh, Assam, India. Incidence in comparison to previous studies has increased. Eight patients were treated in the study period. Different approaches were used. Mean age of presentation was 18 years and more than 75 % of them were either Stage III or Stage IV. Different approaches like transpalatine, transpalatine along with transnasal, lateral rhinotomy and endoscopic surgery were used. The recurrence rate was 12.5 %. Surgery by transpalatine approach should be the choice of treatment. Endoscopic resection should be only used for Stage I or Stage II disease.

Keywords Juvenile nasopharyngeal angiofibroma · Incidence · Current trend and management · Anaesthetic management in juvenile nasopharyngeal angiofibroma

Introduction

Juvenile nasopharyngeal angiofibroma is a very rare disease. It is highly vascular, uncapsulated and locally invasive tumor. Its incidence has been cited to be 0.05 % of all head and neck neoplasms as per many study reports [1]. In recent times there has been a major change in its epidemiology, pathogenesis, diagnosis, medical management, pre-operative care, surgical management and post-operative care including radiotherapy. The exact site of origin is still unclear with current opinion being the postero-superior margin of the sphenopalatine foramen or from the contents of the distal vidian canal and the trending concept being a vascular malformation resulting from incomplete regression of the first branchial arch artery [2]. There has been an upsurge in the number of cases reporting to our department, Department of Otolaryngology and Head and Neck Surgery, Assam Medical College, Dibrugarh, Assam, India. Since the department caters to the need of most of the people residing in the north-eastern part of south-east Asia mainly India, it can be well said that a changing trend has been seen in the incidence of the disease in recent times. Most of patients present with nasal obstruction and bleeding from nose which can be life-threatening also. The most common presenting symptoms are severe, recurrent epistaxis with persistent nasal obstruction. As the disease progresses, facial deformities, proptosis, blindness, and cranial nerve palsies may occur. The diagnosis of JNA is essentially based on a careful history and nasal endoscopic examination, supplemented by imaging studies using computed tomogram (CT) and Magnetic Resonance Imaging (MRI). MRI is more accurate than CT in assessing intracranial extension [3]. Biopsies to establish histological diagnosis are contraindicated and definitive diagnosis is established by angiography which also serves as therapy

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since embolization of the tumor feeding vessels may be performed at the same sitting. Surgery is the main modality for this tumor. The treatment for the nasopharyngeal angiofibroma can present formidable problems for the surgeon. These most notably are concerned with the management of bleeding during extirpation of the lesion and because of these problems, the subject has stimulated a great deal of interest among Otolaryngologists and Head and neck surgeons [4].

Materials and Methods

The study is a retrospective as well as a comparative study. The main objective being to highlight the changing trend in the epidemiology, pathogenesis, diagnosis, medical management, pre-operative care, surgical management and post-operative care including radiotherapy. Although the study period is short but it is important and a note should be taken about the actual scenario that is trending. In a span of 6 months (October 14'–March 15) there has been alarming reporting of eight cases of which one recurred thrice even after endoscopic resection.

Out of eight patients six were of age group 12–19 years and two were of age group 20–30 years. On elaborating the history of later group it was found that the later two had a slow growth and less recurrent symptoms. The tumors were staged according to the Fisch staging and 75 % were of either Stage III or Stage IV.

Different approaches like transpalatine, transpalatine along with transnasal, lateral rhinotomy and endoscopic surgery were used.

The recurrence rate was 12.5 %. The patients are on continuous follow up.

Results

It was a study of eight patients in a period of 7 months. All the patients were of the age group 12–30 years. Dibrugarh and Tinsukia reported 37.5 % cases each and Sivasagar and Karbi Anglong reported 12.5 % each.

Region Wise Distibution

Dibrugarh	3 nos. of cases (37.5 %)
Tinsukia	3 nos. of cases (37.5 %)
Sivasagar	1 no. of cases (12.5 %)
Karbi Anglong	1 no. of cases (12.5 %)

Clinical Features

The clinical features can be broadly classified into [5].

Symptoms		Signs	
Nasal obstruction	100 %	Nasopharyngeal mass	100 %
Recurrent epistaxis	75 %	Nasal cavity and nasopharyngeal mass	75 %
Headache	75 %	Palatal displacement	25 %
Speech defects	50 %	Cheek swelling	75 %
Nasal discharge	37.5 %	Proptosis	37.5 %
Snoring	37.5 %	Retracted ear	25 %
Deafness	37.5 %	Conductive deafness	25 %
Post orbital pain	37.5 %	Reduced visual acuity	12.5 %

Staging

Fisch Staging for JNA.

Stage	Description of Tumour Involvement
I	Limited to nasopharynx, bone destruction negligible or limited to sphenopalatine foramen
II	Invades pterygopalatine fossa or maxillary, ethmoid, or sphenoid sinus with bone destruction
III	Invades infratemporal fossa or orbital region
IIIA	No intracranial involvement
IIIB	Extradural, parasellar involvement
IV	Invades dura
IVA	Without cavernous sinus, pituitary, or optic chiasm involvement
IVB	With the above

Stage Distribution

The patients in the study were distributed into the following stages.

I	0 %
II	12.5 %
III	75 %
IV	12.5 %

Treatment Modalities

All cases were treated by surgical modalities without pre-operative embolisation and adjuvant radiotherapy.

Pre-Operative Care

All patients were properly planned for operative procedures and pre-operative investigations included all routine investigations, liver function test, renal function test, chest x-ray, ecg, serology for infectious diseases etc. Apart from these tests a CTscan, MRI, and CT Angiogram were performed. In Fig. 1a, b we can see that there is an ill defined lobulated heterogeneously enhancing soft tissue density lesion measuring 4.1 cm (ap) $\times$ 5.33 cm (t) $\times$ 3.53 cm (cc), completely filling the left nasal cavity, causing widening of the sphenopalatine foramen and pterygopalatine fissure and extending into pterygopalatine and infratemporal fossae on right side and also extending to the right orbital apex. Posteriorly the lesion is projecting into the nasopharynx and causing pressure effect on right Eustachian tube opening. Superiorly the lesion is eroding the basisphenoid and extending into right sphenoid sinus. The lesion is noted eroding the lateral wall of the right sphenoid sinus and extending to the middle cranial fossa. The lesion is typically causing anterior bowing of the posterior wall of right maxillary sinus. Medially the lesion is causing deviation of the nasal septum to the left side. In Fig. 2 we can see that there is expansile and enhancing soft tissue mass filling the

sphenoid sinus with expansion and destruction of the sphenoid sinus walls and bone around foramen ovale on right side. The base of medial as well as lateral pterygoid plates are eroded with presence of soft tissue. There is minimal soft tissue in right pterygopalatine, medial temporal region as well. In Fig. 3 we can see that there is an well defined heterogeneously enhancing soft tissue density lesion measuring 2.7 cm (ap) $\times$ 2.8 cm (cc) $\times$ 1.4 cm (t) in the posterior aspect of the right nasal cavity in the region of choana. The lesion is extending into the nasopharynx and reaching into the posterior nasopharyngeal wall with compression of fat plane between the lesion and posterior pharyngeal wall. Superiorly the lesion is eroding the basisphenoid and extending into right sphenoid sinus. The lesion is noted to cause erosion of the medial portion of the body of right pterygoid. Right pterygomaxillary fissure is widened. Figure 4 shows that the mass is removed transpalatal. Figure 5 shows that the mass is removed transnasally. Figure 6 shows angiofibrotic tissue hold by the corresponding author. Five patients received pre-operative blood transfusion and the rest three received intraoperatively. None of the patient received adjuvant radiotherapy or hormonal therapy in the preoperative period. Airway assessment with Mallampatti Grading was I and II on average.

Fig. 1 CT scan of nose and PNS and base of skull

Fig. 2 CT scan of nose and PNS and faciomaxillary

Intraoperative and Anaesthetic Care

A haemodynamically stable patient, during and after surgery, is the main goal of anaesthetists. The prime anaesthetic concern in the resection of JNAs is prevention of the aspiration of blood, minimization of blood loss and prevention of airway obstruction in the postoperative period owing to surgical manipulation. In this study, after securing the cuffed endotracheal tube, a throat pack was inserted as an additional measure to prevent aspiration. Throat packs should be removed at the end of surgery before extubation to prevent airway obstruction [6].

The patients showed the following average findings.

	Blood pressure	Pulse rate	SPO ₂
Baseline	118/72 mmHg	98	100 %
After induction	110/72 mmHg	88	100 %
After intubation	118/74 mmHg	92	100 %
Intra-operative	98/60 mmHg	72	100 %
Premedication			Ondansetron Glycopyrolate Tramadol
Induction			Propofol
Muscle relaxant for maintenance			Vecuronium

Fig. 3 CT scan of nose and PNS and base of skull

Fig. 4 Transpalatine incision for surgery

Fig. 6 Angiofibrotic tissue

Fig. 5 Transnasal removal of mass

Reverse and extubate with Neostigmine and Glycopyrrolate when patient was awake and responding to verbal commands.

Surgical Approaches

Transpalatal	4 Cases	50 %
Transpalatal and transnasal	2 Cases	25 %
Lateral rhinotomy	1 Case	12.5 %
Endoscopic excision	1 Case	12.5 %

Post-Operative Care

All patients were given post nasal pack and the pack was removed under strict control of the whole procedure in the O.T. Post pack removal bleeding was not seen in any case. However five patients received blood transfusion as a preventive measure.

Outcome

All the patients presenting to the department were male of age group 12–30. The patients received utmost care and needful treatment. All patients received surgical treatment.

Only one case was operated endoscopically and he was in Stage II. One case was operated by lateral rhinotomy and he was in Stage IV. The rest of the patients were in stage III and were operated by transpalatine and both transpalatine and transnasal approach. No recurrence was found in seven cases. One of the patient was operated thrice after twin recurrence. He was operated by endoscopic procedure for the first episode of recurrence. However it again recurred.

There were no complications during the whole period. No patient developed palatal fistula or cerebrospinal leakage. There is no recorded death of any patient during the period.

It was seen from the study that JNA is more common than thought and the proper treatment depends on the exact extent of the tumour.

Discussion

Angiofibroma is a relatively rare tumour and the age of onset is in the second decade. The reported incidence ranges from 1/5000 to 1/50,000 of otolaryngological patients in different countries. Over a 30-year period (1949–1979), the Head and Neck Service of the Sloan-Kettering Memorial Cancer Centre in New York reported only 31 male patients between the ages of 11 and 21 (median age 14 years) [7].

Angiofibromata often grow and extend along natural foramina and fissures, displacing and distorting the adjacent structures. Larger tumours, however, may erode bone. As they expand, collateral blood supplies develop. The tumours spread laterally from the sphenopalatine foramen to the pterygopalatine fossa through the pterygomaxillary fissure. From this narrow fossa they eventually expand into the infratemporal fossa and the cheek. 25 They can also extend along the inferior orbital fissure, across the apex of the orbit into the superior orbital fissure. Continued tumour expansion causes pressure erosion of the base of the pterygoid plate and greater wing of the sphenoid. This brings the tumour against the dura of the middle cranial fossa. Medially, the tumour fills the nasopharynx and distorts the nasal septum, turbinates and the soft palate. It may erode into the posterior ethmoidal and sphenoidal sinuses, allowing direct extension of tumour into the orbit, cavernous sinus and the parasellar region [7].

Surgical resection is the recommended modality of treatment for this highly vascular benign tumor of the nasopharynx. The availability of different modalities for pre-op reduction in the vascularity of tumors have reduced intraoperative bleeding. However, management of JNA still poses a great challenge to ENT surgeons worldwide. In our facility where pre-op embolization is not available, adequate

exposure in transpalatine approach gives good results with minimum morbidity and mortality. Even tumors extending to pterygopalatine fossa and limited extension to infratemporal fossa can be managed through the transpalatine approach in experienced hands [8]. Endoscopic surgery is becoming a promising approach for early stage growth mainly I and II. The main advantage of endoscopic surgery is the possibility of obtaining a broad view of the lesion and its anatomic relationship with adjacent structures, promoting more accurate, complete dissection and better control of bleeding [9–11]. Other advantages include less surgical time, hospitalization, absence of visible scars, avoids complication such as epiphora, dysesthesia, trismus, and craniofacial deformities [10, 12–15].

External beam radiation therapy represents an effective mode of treatment for patients with advanced JNA. Although the latency period may be long, we think the likelihood of potentially fatal complications developing at the radiation dosages we recommend is less than the risk of significant morbidity and mortality associated with surgical intervention in these cases [16].

Image-guided endoscopic coblation may be a safe and effective method for resection of intranasal portions of advance JNA tumors. Currently proposed methods are limited by the complex instrumentation for adequate debulking and prolonged concern for frequent and copious bleeding during the procedure. Our proposed method of resection using the Coblator II System in many cases may limit the need for multiple instruments and improve control of intraoperative hemorrhage. Additional studies are necessary to comprehensively evaluate further applications for coblator-assisted resection of JNA [17].

The study previously done in various parts of the world have shown that juvenile angiofibroma is rare and its incidence is 0.05 %. However in this study it was seen that there should be a rise in the incidence as within a span of 7 months and just in a small region of the developing country there has been upsurge in the number of cases.

Conclusion

Surgery remains the mainstay of therapy. Newer advances in technology like endoscopic resection and co-ablation can be an effective tool in the coming days. However, it should be limited to earlier stages of the disease at present. Efforts should be made to found out the exact scenario of the disease condition and changing trend in its epidemiology, pathogenesis, diagnosis, medical management, pre-operative care, surgical management and post-operative care including radiotherapy.

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