CASE REPORT

Atypical Site of Juvenile Nasopharyngeal Angiofibroma: A Rare Case Report And Review of Literature

Pradip Kumar Tiwari¹, H.P. Saikia², Nabajyoti Saikia³, Jyotishmoy Bora⁴

ABSTRACT

Introduction: Juvenile nasopharyngeal angiofibroma is a very rare, highly vascular, unencapsulated 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. Atypical sites of juvenile nasopharyngeal angiofibroma are very rare. Management of atypical presentation of juvenile nasopharyngeal angiofibroma is complex and needs detailed evaluation and interdisciplinary approach.

Case report: Case report with complete review of other studies along with comparative findings and different management approaches during the study periods was taken in account. The presentation and procedures to differentiate the late stages of atypical juvenile nasopharyngeal angiofibroma from other tumors was done. A single case of atypical juvenile nasopharyngeal angiofibroma was evaluated and treated. Age of presentation was 26 years and Stage IV of the disease. Transpalatine along with transnasal approach was used. No recurrence till date has been seen. Patient is on continuous follow up.

Conclusion: 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, radiotherapy, Endoscopic resection

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INTRODUCTION

Juvenile nasopharyngeal angiofibroma is a very rare disease. It is highly vascular, unencapsulated 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 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 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 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 & neck surgeons. Reports of primary atypical juvenile nasopharyngeal angiofibroma, especially those originating in the nasal cavity structures have appeared exceptionally in the literature. The focus of our interest was to assess the important diagnostic modalities to differentiate the lesion and its exact line of treatment. We hereby tried to present a case of primary atypical juvenile nasopharyngeal angiofibroma and its detailed diagnostic evaluation and treatment.

CASE REPORT

A 26 year old male presented to the emergency department, Department of Otolaryngology and Head and Neck Surgery, Assam Medical College, Dibrugarh, Assam, India, with a chief complaint of bleeding from the nose. The bleeding was profuse and with proper conservative medication and surgical dressing it stopped. The patient also complained of difficulty of respiration. His blood pressure was 98/60 mm of Hg. Laboratory data were as follows: Hb 7.6, white blood counts 9600per mm$^3$, ESR 100 mm AEFH. Differential count $\text{N}_{70}\text{L}_{12}\text{M}_{1}\text{B}_{0}$ Es, random blood sugar 108 mg/dl, serum Na$^+$ 150.8 mmol/lt, K$^+$ 4.13 mmol/lt, PT 11.3, INR 0.97, urea 38mg/dl, creatinine 1.1 mg/dl, ALT 23, AST 31 and platelet count 1.68 lac. In Fig-1 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. There is erosion and widening of the sphenoid sinus walls. The soft tissue has anteriorly extended to the ethmoid sinuses bilaterally and partially filled the nasal cavity both the maxillary antrae. There is destruction of floor of sphenoid sinus with thickening of the nasopharyngeal roof. 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. There is minimal soft tissue in right pterygomaxillary fissure. There is remodelling with increased thickening of the orbital floor.

Clinical Features

The clinical features were as follows- symptoms included mild nasal obstruction, recurrent epistaxis, headache, speech defects, nasal discharge, snoring and post orbital pain whereas signs included proptosis and nasal resonance.

Staging: Fisch Staging for JNA

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

Stage Distribution: The patient in the study was in stage IV

TREATMENT MODALITIES

Pre-Operative Care

The patient was 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 CT scan, MRI, and CT Angiogram were performed. In Fig-1 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. There is destruction of floor of sphenoid sinus with thickening of the nasopharyngeal roof. 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. There is minimal soft tissue in right pterygomaxillary fissure. There is remodelling with increased thickening of the orbital floor. Airway assessment with Mallampatti Grading was II.
**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 report, 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.

The patients showed the following findings:

<table>
<thead>
<tr>
<th></th>
<th>Blood Pressure</th>
<th>Pulse rate</th>
<th>SPO₂</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baseline</td>
<td>118/72 mm Hg</td>
<td>98</td>
<td>100%</td>
</tr>
<tr>
<td>After Induction</td>
<td>110/72 mm Hg</td>
<td>88</td>
<td>100%</td>
</tr>
<tr>
<td>After Intubation</td>
<td>118/74 mm Hg</td>
<td>92</td>
<td>100%</td>
</tr>
<tr>
<td>Intra-operative</td>
<td>98/60 mm Hg</td>
<td>72</td>
<td>100%</td>
</tr>
</tbody>
</table>

Premedication: Ondansetron, glycopyrolate, tramadol; Induction: Propofol
Muscle relaxant for maintenance: Vecuronium
Reverse and extubate with Neostigmine and Glycopyrolate when patient was awake and responding to verbal commands

**Surgical Approaches: Transpalatal**

**Post-Operative Care**

The patient was 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. However the patient received blood transfusion as a preventive measure.
Outcome: The patient presenting to the department was male of age 26. The patient received utmost care and needful treatment. The patient received surgical treatment. No recurrence was found. There were no complications during the whole period.

**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. 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. 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 paranasal region.7

Surgical resection is the recommended modality of treatment for this highly vascular benign tumor of the nasopharynx. 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.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. The 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. Angiofibroma of the larynx is diagnosed by direct laryngoscopy and biopsy.

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 find 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. Whatever be the cause of angiofibroma an atypical site of angiofibroma should always be dealt with surgical care soon.

REFERENCES


