Original article:

**Study of role of pre-operative embolisation of Juvenile Nasopharyngeal Angiofibroma (JNA) tumours**

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Abstract

Juvenile nasopharyngeal Angiofibroma (JNA) is a benign but locally invasive tumour. Patients are usually in their adolescent age and present with epistaxis and nasal blockage. Diagnosis is based on clinical evaluation and the C.T. scan findings. Pre-operative superselective embolization (SSE) and surgical excision is the treatment of choice. Patients, mostly in their pre-pubertal adolescent age present with epistaxis, nasal blockage, headache and hyponasal voice. Advance tumours may present with facial asymmetry, palatal bulge, proptosis and blurring of vision depending upon local and intracranial extensions. In patients with intracranial extensions, other symptoms were nausea and vomiting. The site of origin of the tumour is usually superior aspect of sphenopalatine foramen. Abnormal growth of embryological chondrocartilage, nasopharyngeal fibrovascular stroma and testosterone acting on dislocated inferior turbinate are the pertinent pathogenesis. A number of classifications so far have been proposed but none is universally accepted.

Keyword - JNA, Super selective embolization and blood loss.

Introduction

Juvenile nasopharyngeal Angiofibroma (JNA) is a lobulated, firm non-capsulated tumour with numerous attachment and multiple feeding vessels. Diagnosis is relied upon clinical and radiological findings. Recurrent epistaxis and bleeding during surgery is hazardous at time.[1,2] Pre-operative superselective embolisation has made the dissection feasible for surgeons. Surgery is considered the mainstay of all available treatment modality in this tumour. Diagnosis of this entity will rarely pose a difficulty of diagnosis if a CT scans in coronal cuts at and around posterior choanae is carefully looked for erosion of pterygoid plates, widening of sphenopalatine foramen and presence of mass in the pterygopalatine fossa. These signs in our series were the diagnostic criteria for angiofibroma. Since biopsy is contraindicated in suspected case of JNA, dilemmas of diagnosis sometimes cause delay in the commencement of actual management.

Gold standard investigation is Angiography which shows typical tumour blush [3]. Surgery is the primary treatment modality employed in most centres. Radiotherapy is an alternative treatment reserved for intracranial extension[4] and inoperable advance diseases[5]. Alternative treatment such as chemotherapy and hormonal therapy has shown limited success and it is hardly practiced. Recurrences are common, however in such cases an early diagnosis and a salvage surgery under Super Selective Embolisation (SSE) results in better outcome [6]. A number of surgical approaches have been utilised with acceptable morbidity. Moreover endoscopic surgery has
gain popularity for tumour removal in selected cases in their early stages[7]. In investigation, C.T. scans with and without contrast were the mainstay of diagnostic tool employed. Several coronal cuts at 3 mm each at and around the posterior choanae were categorically requested in all patients. Finding of each cases were noted for special characteristic. MRI scans were carried out in cases with suspected intracranial extensions. MRA (magnetic resonance angiography) was performed in some cases imposing diagnostic difficulty.

Apart from classical sign of anterior bowing of posterior antral wall, CT scans were observed for three more signs which were invariably found in all cases. These were, erosion of medial plate or root of the pterygoid process, widening of sphenopalatine foramen and presence of mass in pterygopalatine fossa. Infratemporal fossa invasion has clinical sign of cheek swelling in two cases and proptosis invariably indicates orbital cavity invasion.

Material and methods

The present work was conducted in our department. The Intervention department received 21 referrals, all male, and majority between 16-23 years of age. They reported from 1989 to 2014 at LTMMC and LTMGH, Sion hospital, Mumbai. Super selective embolization of the tumour done to reduce intraoperative blood loss.

Observations and results

There is insignificant intraoperative blood loss ranging from the 80 to 1500 ml in our series.

A. Preoperative

B. Postoperative

Discussion

In our series there are 21 patient were examined. All of the presented with h/o epistaxis and headache. Additionally nasal blockage was present in 6 patients and 4 patents are also presented with the proptosis and
blurring of vision. On CT PNS 15 patients have mass in the nasopharynx and 16 patient have mass in nasal cavity. Extension of the mass in the pterygopalatine region has been observed in the 20 patients. Paranasal sinus involvement was also not uncommon. Paranasal sinus involvement is noted in the 7 patients in the sphenoid, 4 patients in the ethmoidal and 9 patients having maxillary sinus involvement. Out of 21 patient have also intraorbital component within and 4 patients have intracranial involvement. Out of these 2 patients also had carotico-cavernous fistula. Extensions of tumors beyond pterygopatine fossa are presented. The cases were staged according to Chandler et. al. (1984).[8] . 2 were Chandler type 1, 5 were type II, 10 were type III and 4 were type IV. The feeding vessels of the tumour in each case were noted in angiography. In all cases which embolization were performed, a minimum of 48 hours lapse were allowed post SSE to observe any complication of this procedure, the worst of which is blindness or stroke. Incidence of complication was more in as stage advances. In this series a major complication occurred – Central retinal artery occlusion which was managed by immediate ophthalmology reference followed by paracentesis. The patient achieved 50 % recovery of the vision. Diagnosis was mostly made on typical radiological findings. Angiography is a gold standard investigation which usually essential in diagnosis. Furthermore pre-operative embolization is considered mandatory for this procedure as it significantly reduces blood during surgery [6]. However in our practice we did not performed the diagnostic angiography to avoid a repetition of angiography in a short span of time. We only advocate single angiogram together with Super Selective Embolization (SSE) 48 hours prior to surgery to reduce the risk involved. The surgical technique varies from transpalatal approach to maxillary swing approach in this series. Surgical procedure for each case was noted. All patients were operated by the same surgeon with or without other surgeon from ORL team or neurosurgeon. Regular follow-up every six weeks for first year and every 3 months for the following 2 years were carried out. Patients who default follow up also included in this series. All suspected cases for residual or recurrent tumours were subjected to a repeat CT scan or MRI depending upon recurrence at the local site or intracranial. Results – there is significantly low blood loss intraoperatively ranging from 400 to 800 ml.

Conclusion
As JNA is highly vascular tumours , it is necessary to perform the preoperative embolization tumour to reduce intraoperative blood loss.

References
