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ORIGINAL ARTICLE

APPRAISAL OF CLINICAL PROFILE AND MANAGEMENT OF JUVENILE NASOPHARYNGEAL ANGIOFIBROMA IN MALAYSIA

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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 embolisation (SSE) and surgical excision is the treatment of choice. The out patient clinic of ORL-HNS hospital of University Science Malaysia received 25 referrals, all male, majority between 9-13 years of age and few adolescents. Clinically the patients were consistent with symptoms of recurrent epistaxis and nasal blockage. They reported from October 1998 to October 2001 from with in the state of Kelantan and the nearby states of Pahang, Kedah and Terenganu. Diagnosis was mostly made on typical radiological findings and the tumours were classified accordingly into four stages. SSE and surgical excision was carried out in all cases. Regular follow-up helped us to identify early recurrences which were treated with salvage surgery or radiotherapy in one case with extensive intracranial extension. A retrospective review of presenting features, diagnostic difficulties, surgical approaches and its outcome is presented. Maxillary swing procedure performed in three cases as a new surgical option in the management of JNA is also discussed.

Key words : Juvenile nasopharyngeal angiofibroma, clinical profile, diagnosis, surgical options, maxillary swing.

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Introduction

JNA is an uncommon clinical entity comprising of 0.5% of all Head and Neck tumours[1]. 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[2]. 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[3].

Diagnosis is based on clinical evaluation in conformity with typical CT scan findings rather than tissue biopsy. Gold standard investigation is Angiography which shows typical tumour blush[4]. Surgery is the primary treatment modality employed in most centres. Radiotherapy is an alternative treatment reserved for intracranial extension[5] and inoperable advance diseases[6]. 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[7]. 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[8].

A retrospective review of JNA cases operated in Hospital Universiti Sains Malaysia is presented. The maxillary swing approach, which is practiced for nasopharyngectomy in selected cases of recurrent nasopharyngeal carcinoma in this centre, was also employed in three cases of angiofibroma in children. The procedure is critically evaluated for feasibility and adequacy of approach and its complication in children.

Patients and Methods

Medical records of 25 patients who were operated from October 1998 to October 2004 were analysed retrospectively. The cases were reviewed for their demographic data, clinical presentations, diagnostic investigations, treatment modalities and its outcome and recurrence. The complications of maxillary swing approach utilised in three cases in recurrent JNA were critically evaluated.

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. The cases were staged according to Chandler et. al. (1984).[9]

Pre operative tissue diagnosis were look for if present. The feeding vessels of the tumour in each case were noted in angiography. Final histopathological examination is considered as ultimate diagnostic criteria. Immunohistochemistry utilising various markers were employed in those cases where problem of diagnosis existed. Diagnosis other than JNA is excluded from 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

Demographic study revealed total number of cases is 25 exclusively male with majority of patients (17 cases) in their per-pubertal age of 9-13 years. Eight cases are above 13 years old and the oldest patient in this series was 24 year old. There were 13 cases referred from other hospitals in east Coast of Malaysia and Alor Star Hospital, Kedah. Presenting signs and symptoms are presented in table I.

There were no preoperative tissue diagnoses except in two cases because it is contraindicated. The two cases were mistaken for nasopharyngeal carcinoma and pyogenic granuloma respectively and biopsy was performed in other centres prior to their referrals.

Apart from classical sign of anterior bowing of posterior antral wall (fig.1), 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 (fig.2). Infratemporal fossa invasion has clinical sign of cheek swelling in two cases and proptosis invariably indicates orbital cavity invasion (figure 3 and 4).

Tumour was staged base on radiological findings. Majority of cases presented in stage II and III although two stage I and few stage IV cases were also reported (Table II). Extensions of tumours beyond ptygopalatine fossa are presented in Table III.

All surgical procedures were carried out after angiography and super selective embolisation (SSE) except in two cases. One case in stage I was operated through transpalatal approach without an embolisation due to unavailability of this facility in our centre prior to 1998. In the other case all attempts to embolise the feeding artery virtually failed and only angiography could be performed. Internal maxillary artery was the main feeding vessel in all cases where as branches from facial artery, ascending pharyngeal artery, superficial temporal artery, opposite internal maxillary artery minor supply from internal carotid artery were the other feeding vessels in some cases (Table IV).

In all cases which embolisation 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. In this series no major complication was noted.

Apart from surgical excisions, adjunct surgical procedures were also carried out. External carotid ligation for a failed embolisation, internal

maxillary ligation due to inadequate embolisation, reconstruction of eroded orbital floor utilizing rib graft, gastrostomy far absolute dysphagia and tracheotomy to provides patent airway in extensive approaches taken in some cases.

Criteria for selection of surgical approaches were decided based on the size and extend of tumour, intracranial extension and expertise available. In total there were 25 subjects with 30 surgical procedures including recurrent cases. In most of the cases two window approaches were employed to facilitate adequate exposure and convenience of dissection. Combined external cervical and neurosurgical approach were decided in cases of intracranial extension with dura, sellar and cavernous sinus encroachment. Besides the conventional surgical procedures, a sublabial transverse maxillary dislocation of palate, temporary anterolateral wall maxillectomy followed by subsequent plating and the maxillary swing procedure were the other surgical approaches carried out depending upon the extension of tumour into adjacent areas. Except the initial few cases and the maxillary swing approach which were done by Weber Ferguson incision, mostly the sublabial midfacial degloving incisions were employed to avoid any obvious scar or deformity. Endoscopic assisted together with transpalatal approach were performed in two cases. Distribution of incisions and approaches are shown in Table V. Blood loss during operative procedure was from 300 cc to 1000 cc except in two cases which blood loss more than 1 litre.

Size of the tumours removed were varied from 2 to 10 cm. Histopathological examination confirmed JNA in all cases which were shown to have variable thickness of smooth muscles around abundance of blood channels (Figure 5). These vessels were lined by endothelial cells with a rich fibrous stroma. Immunohistochemistry technique was utilized in some bizarre histopathological picture observed in some cases.

Post operative complications were trismus, epiphora, facial asymmetry, obvious scar, transpalatal fistula. Trismus and fistula were corrected by conservative spatula enforcement method and by secondary repair respectively.

Recurrences noticed in six cases. Two cases were picked up early in follow up whereas the remaing four were lost to follow up and later reported with extensive intracranial extension. All recurrence cases underwent revision surgery except one massive intracranial extension which was subjected to radiotherapy, a good control of lesion was achieved and no further recurrence until the last follow-up was observed. Histopathological examinations of removed specimen reconfirm the diagnosis of JNA.

Discussion

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 where as the ultimate histopathological diagnosis comes from surgical specimen. Recurrent epistaxis and bleeding during surgery is hazardous at time. 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. Nevertheless biopsy was obtained in two patients referred from different hospital may be due to inexperience doctors. In our experience, there were four cases which presented similar to JNA but later turn out to be nasopharyngeal carcinoma (11 yrs. and 14 yrs.), pyogenic granuloma (13 yrs) and chordoma (18 yrs).

Angiography is a gold standard investigation which usually essential in diagnosis. Furthermore pre-operative embolisation is considered mandatory for this procedure as it significantly reduces blood during surgery[7]. However in our practice we do not perform the diagnostic angiography to avoid a repetition of angiography in a short span of time. We only advocate single angiogram together with Super Selective Embolisation (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. Each method has its own limitations. We prefer two windows approach, one to provide adequate room for dissection and the other to allow sufficient pulling of the tumour which most of the time is unexpectedly large enough to let dissection and delivery through the same window.

Tumour staging has been helpful in understanding the comparative value of different surgical procedures[10]. For a very large tumour with intracranial extensions, maxillary swing approach with or without combined neurosurgical approach have been employed successfully in this series. Stage I and Stage II tumours were successfully treated with transpalatal and lateral rhinotomy approaches. However mid facial degloving incision with transnasal, transantral and trans-maxillary approaches were the main surgical procedure employed in nasal and postnasal space tumours. Weber Ferguson incision with transmaxillary approach in few cases and temporary anterolateral wall maxillectomy with subsequent plating in a couple of patients were employed earlier. Weber Ferguson Longmire incision was utilized in three cases of maxillary swing procedure.

Few authors are of the opinion that the endoscopic tumour resection with proper patient selection, endoscopic resection of juvenile nasopharyngeal angiofibroma is feasible and may be preferable to traditional open approaches. Results suggest that after endonasal resection, disease recurrence is low. Most larger lesions, especially those with intracranial spread, continue to require open approaches for complete resection[11]. Our experiences in two cases make us believe endoscopic resection should be reserved for early tumour in stage I or stage II. Excessive bleeding and inappropriate instrument in an occult anatomy of nasopharynx makes endoscopic surgery difficult [12].

Failure of surgical procedures resulting in early recurrences was encountered with almost all approaches. However maxillary swing approach was found to be a convenient procedure with adequate exposure, complete and *in toto* resection of tumour with acceptable morbidity in three selected cases of extensive tumour recurrences. Further recurrences which were not seen in any of these three cases done by maxillary swing approach do not reflect the advantage of this procedure. We value this approach for a wide exposure of tumour to facilitate dissection and mobilisation in extensive JNA encroach central skull base. This approach was combined with an anterior craniotomy in one case to release the intracranial subdural extension of tumour in temporal lobe. Resulting morbidity was low in all these three cases and there was no facial growth retardation on long follow up of three to five years in these cases. Therefore we recommend this approach for advance stage III and IV tumours.

Although surgery is considered to be essential choice of treatment for these tumours, radiotherapy was employed in one case of an advance intracranial extension implicating seller, temporal lobe of brain and the cavernous sinus. Repeat CT scan at 3 months interval revealed substantial reduction in tumour size and virtually no tumour nine months after the irradiation.

Incidences of complications in this series were linked to tumour extension, operative procedure and the radiotherapy. Tumour complications were reported as hazardous epistaxis, cheek swelling (a definite indication of infratemporal fossa involvement), proptosis, ptosis, blindness and conductive hearing loss. Complications related to surgery were bleeding, epiphora, trismus, palatal fistula and facial asymmetry. Post-radiation complication noticed in the only irradiated patient was osteotitis followed by necrosis of anteromedial wall of maxillary antrum and the adjacent nasal bone resulted in obvious facial asymmetry. Surgical excisions in recurrent cases were difficult due to associated extensive adhesion. This resulted in difficult tumour dissection and mobilization and therefore more bleeding.

In our experience exposure with inadequate dissection, excessive intraoperative bleeding and extension into skull base, orbit infratemporal fossa, sphenoid and ethmoid sinuses rendered the patient more vulnerable to recurrences. However pre-operative tumour delineation, effective embolisation and adequate surgical exposure were the main factors preventing the recurrence. In any case one must accept the fact that the biological behaviour of this tumour renders it highly unpredictable for recurrence and extension into adjacent sites, whatsoever modality of treatment or method of surgical excision is employed. It also noted previously that the extensions of the tumor seem to be independent, each one with distinct behavior[13]. Most of the recurrences will appear with in the first 12 months and a frequent follow-up during this period is essential for in time revision surgery with minimal morbidity.

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