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Surgical approaches to juvenile nasopharyngeal angiofibroma

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SUMMARY. Introduction: **Juvenile nasopharyngeal angiofibromas are highly vascular, non-encapsulated tumours affecting predominantly young males. These lesions are benign histologically but they may become life-threatening with excessive bleeding or intracranial extension.** Material and methods: **The surgical approaches to 22 male patients with nasopharyngeal angiofibromas are reviewed. A modification of midfacial degloving performed without rhinoplasty incisions and lateral osteotomies is described.** Results: **The patients' ages ranged between 9 and 26 years (mean 14.9). Three stage I tumours, 8 stage II tumours, 6 stage III tumours and 5 stage IV tumours were included into this study. All stage I lesions and one stage II lesion were treated via transnasal endoscopic approach. A modified midfacial degloving approach was used for the removal of seven other stage II lesions, all six stage III lesions, and three stage IV lesions. A combined midfacial degloving-infratemporal fossa Fisch C-transcranial approach was the route chosen for the remaining two stage IV lesions. The complications encountered in the postoperative course include temporary facial palsy in one patient (following a Fisch C infratemporal resection), mild crusting in the nasal cavity in 8 patients, and facial paraesthesia in 6 patients whose tumours were resected via midfacial degloving, and rupture of the subpetrous part of the internal carotid artery in one patient.** Conclusion: **The suggested treatment of juvenile nasopharyngeal angiofibroma consists of an endoscopic transnasal approach for early stage lesions, and a modified midfacial degloving for almost all of the advanced lesions. The latter approach is very useful considering surgical exposure, duration of surgery, cosmetic outcome, and morbidity. It can be combined with an infratemporal approach or craniotomy if necessary.** © **2005 European Association for Cranio-Maxillofacial Surgery**

Keywords: **juvenile nasopharyngeal angiofibroma; surgical approach; modified midfacial degloving**

INTRODUCTION

Juvenile nasopharyngeal angiofibromas are highly vascular, non-encapsulated tumours affecting predominantly young males, most commonly between the ages of 14 and 25 years (*Fagan* et al., 1997). They originate from vascular structures in the basisphenoid region, particularly in the region of the sphenopalatine foramen and have a tendency to local recurrence when resected incompletely (*Jacobsson* et al., 1988; *Mishra* et al., 1989). Even though these lesions are benign histologically, they are very locally invasive, spread submucosally and thus may easily become life-threatening due to excessive bleeding or by intracranial extension (*Fagan* et al., 1997). Spontaneous regression is reported only for residual angiofibromas (*Neel* et al., 1973; *Stansbie* and *Phelps*, 1986). Neverthless, the tendency to bleed diminishes due to an increasing proportion of fibrous elements (*Girgis* and *Fahmy*, 1973).

The most common presenting symptoms are usually painless, unilateral nasal obstruction and epistaxis (*Fagan* et al., 1997). The treatment options consist of surgery, radiotherapy, electrocoagulation, oestrogen therapy, embolization, injection of sclerosing agents and cryotherapy (*Jacobsson* et al., 1988; *Standefer* et al., 1983). The negative effects of radiotherapy on craniofacial growth and its potential carcinogenic effects limit its use, thus surgery is the mainstay of treatment. Location and extension of the tumour and the experience of the surgical team are important factors when choosing the appropriate surgical approach. The most frequently used surgical routes include transpalatal, Le Fort I maxillotomy, medial maxillotomy, facial translocation, infratemporal, intranasal endoscopic approaches and midfacial degloving (*Fagan* et al., 1997; *Howard* and *Lund*, 1992).

PATIENTS AND METHODS

Twenty-two patients diagnosed with juvenile nasopharyngeal angiofibroma and operated on in this department by two surgeons between 1988 and 2004 were reviewed. They were all male and the mean age at the time of the diagnosis was 14.9 (range: 9–26 years).

The patients were initially evaluated with case history, general examination and a complete otorhinolaryngological examination including nasal endoscopy. Computed tomography (CT) scans were taken for each patient, and some also underwent magnetic

resonance imaging (MRI). Superselective digital subtraction angiography and preoperative embolization were performed 24–72 h before surgery in 13 cases. The tumours were classified according to *Fisch* (1983), stage I tumours being limited to the nasal cavity and nasopharynx, with no bone destruction. Stage II tumours invade the pterygomaxillary fossa and/or the paranasal sinuses with bone destruction. Stage III tumours invade the infratemporal fossa, orbit and/or the parasellar region, but remain lateral to the cavernous sinus. Stage IV tumours invade the cavernous sinus, optic chiasma region, and/or the pituitary fossa.

All patients were treated solely with surgery, radiotherapy was not given. All stage I lesions were treated via a transnasal endoscopic approach. This approach was also used for the removal of one stage II lesion. Seven other stage II lesions were operated on using the modified midfacial degloving (MMFD) approach. All six stage III lesions were also operated on with MMFD. One of these cases had previously been operated in another institution with a lateral rhinotomy. Three stage IV tumours (Fig. 1) were operated with MMFD, among whom one case had previously been operated on using a lateral rhinotomy approach in another hospital. Combined MMFD-infratemporal fossa Fisch C-transcranial (frontotemporal) approach was the route chosen for the remaining two stage IV lesions (Table 1).

RESULTS

Table 2 lists the preoperative and postoperative courses of all 22 patients. Three stage I tumours, 8 stage II tumours, 6 stage III tumours and 5 tumours with intracranial extension (stage IV) were included in this study.

All patients underwent a CT scan while 9 also had MRI. Revisional surgery had to be performed in four cases for a recurrence. Two of these had been operated on previously in another institution while the other two patients' first operations were performed in this hospital. One other patient had undergone radiotherapy previously in another institution before his admission to this hospital. Follow-up ranged from 5 months to 10 years.

The complications encountered in the postoperative course were temporary facial palsy in one patient (following a Fisch C infratemporal resection), mild crusting in the nasal cavity (which disappeared within 3 months) in 8 patients whose tumours were resected via midfacial degloving. Facial paraesthesia was seen in 6 patients also operated on using MMFD. In one patient, the subpetrous part of the internal carotid artery was ruptured after removal of the tumour. This complication was managed by an interventional neuroradiologist with an intravascular covered stent (Fig. 2). There was no other complication.

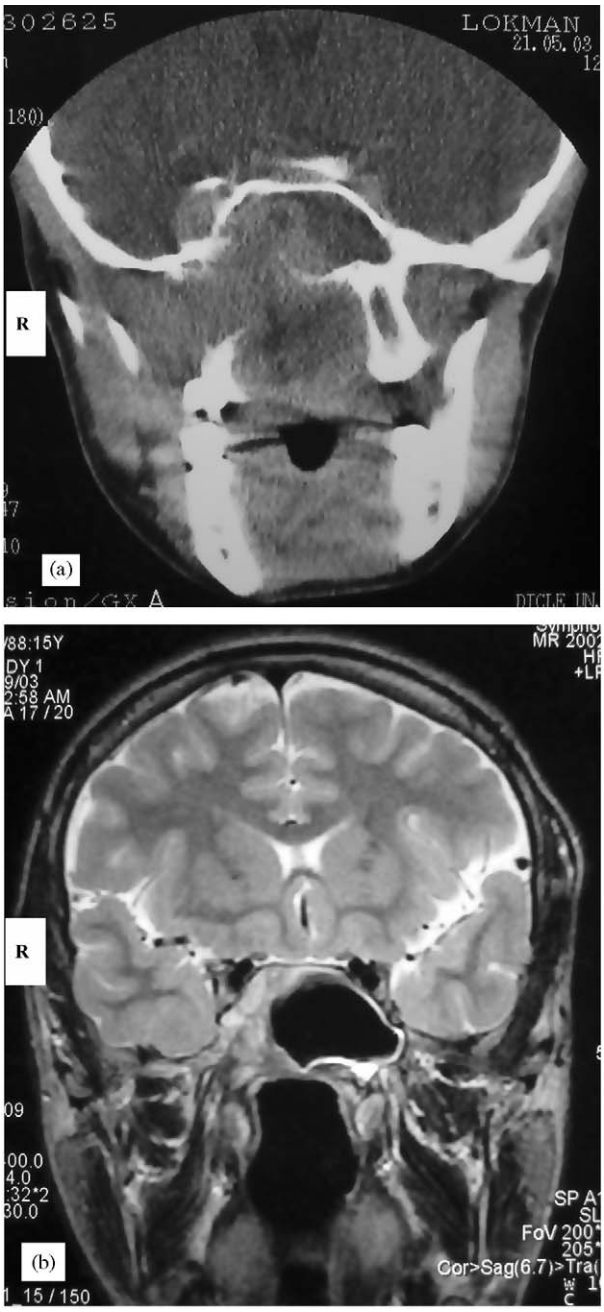

Fig. 1 – (a) Enhanced CT scan (coronal plane) demonstrating a bulky nasopharyngeal (hypervascular) mass extending into the sphenoid sinus, cavernous sinus and right infratemporal fossa. (Patient 2). (b) Coronal T2 weighted MRI of the patient after surgical excision via modified midfacial degloving.

DISCUSSION

Juvenile nasopharyngeal angiofibromas are benign, affecting especially adolescents and young adult males, and comprise about 0.5% of all head and neck tumours (*Jacobsson* et al., 1988). They originate from the vascular structures in the basisphenoid region, particularly the sphenopalatine foramen, and the roof of the anterior nasopharynx or from the posterior nasal fossa (*Jacobsson* et al., 1988; *Mishra* et al., 1989; *Standefer* et al., 1983). They have a