# INTRODUCTION

**J**uvenile angiofibroma is an uncommon, benign and extremely vascular tumour that arises in the tissues within the sphenopalatine foramen. Rarely, it is found at other sites in the nasal cavity and paranasal sinuses..

**J**uvenile angiofibroma accounts for less than 0.5 percent of all head and neck tumours. It develops almost **exclusively** in adolescent males,

The tumour extends into
the nasopharynx, paranasal sinuses, pterygopalatine and infratemporal fossa. Larger tumours can involve the orbit and cavernous sinus.

# PATHOGENESIS

Juvenile angiofibromas present as well-defined, lobulated tumours that are covered by nasopharyngeal mucosa. The tumour consists of proliferating, irregular vascular channels within a fibrous stroma.

Tumour blood vessels typically lack smooth muscle and elastic fibres, this feature contributing to its reputation for sustained bleeding. The stromal compartment is made up of plump cells that can be spindle or stellate in shape and give rise to varying amounts of collagen. It is this that makes some tumours very hard or firm, while others may be relatively soft.

As this tumour is almost exclusively found in adolescent boys, there has always been much speculation and indirect evidence that sex-hormone receptors play some part in its development.

Recent immunocytochemical techniques have been used to show that androgen receptors are present in at least 75 percent of tumours, these receptors being present in both the vascular and stromal elements. Amuch smaller proportion of tumours also have some
progesterone receptors. In contast, oestrogen receptors.
have not been demonstrated? Other factors also play their part in the development of this tumour.

The angiogenic growth factor (vascular endothelial growth factor (VEGF)) has been found localized on both endothelial and stromal cells, perhaps indicating that both cell types play a role in tumour development.

**Overexpression of insulin-like growth factor II (IGFII) has also been found in a large number of juvenile angiofibromas. The IGFII gene is situated on the short arm of chromosome 11 and at that site is the target of genomic**
**imprinting, expressing the paternal allele only. It is thought that overexpression of IGFII might be associated with a tendency to recurrence and poorer prognosis**.

**Juvenile angiofibromas have also been reported to develop 25 times more frequently in patients with familial adenomatous polyposis, a condition that is associated with mutations of the adenomatous polyposis coli (APC) gene. As a result, it has been suggested that germline mutations in the APC gene on chromosome 5q might also be involved in the pathogenesis of sporadic juvenile angiofibromas.**
**. Mutations of beta-catenin have been found in sporadic and recurrent juvenile angiofibromas.**

# PRESENTATION

1- Recurrent severe epistaxes accompanied by progressive nasal obstruction are the classical symptoms of juvenile angiofibromas at the time of presentation.

2-These tumours do not grow fast and so many months or even years may pass before it occurs to the patient or their parents that there is anything seriously amiss. In most, there is a delay of at least six or seven months between the onset of symptoms and presentation. By that time, have other signs and symptoms of tumour
growth and extension. These may include swelling of the cheek, trismus, hearing loss secondary to Eustachian tube obstruction, anosmia and a nasal intonation or plummy quality to the voice.

**3-** More extensive tumour growth with invasion of the orbit and cavernous sinus may cause proptosis, diplopia, visual loss, facial pain and headache

**4-**Anterior rhinoscopy is likely to confirm the presence of abundant mucopurulent secretions in the nasal cavity that usually obscure the tumour from vision, though a few patients have tumour prolapsing from the anterior nares.

**5-**The soft palate is often displaced inferiorly by the bulk of the tumour which can be seen clearly as a pink or reddish mass that fills the nasopharynx.

# ASSESSMENT

- In the past, the exact nature of these tumours was suggested by the plain lateral skull radiographic appearance that would show anterior bowing of the posterior wall of the maxillary sinus.

- Nowadays, the diagnosis is based on the CT and MR appearances that are sometimes confirmed by angiography.

- A trans-nasal biopsy is not necessary and can provoke brisk haemorrhage.

- The exact extent or stage of the tumour can only be determined by a combination of CT and MR imaging and this is vital when planning the surgical resection

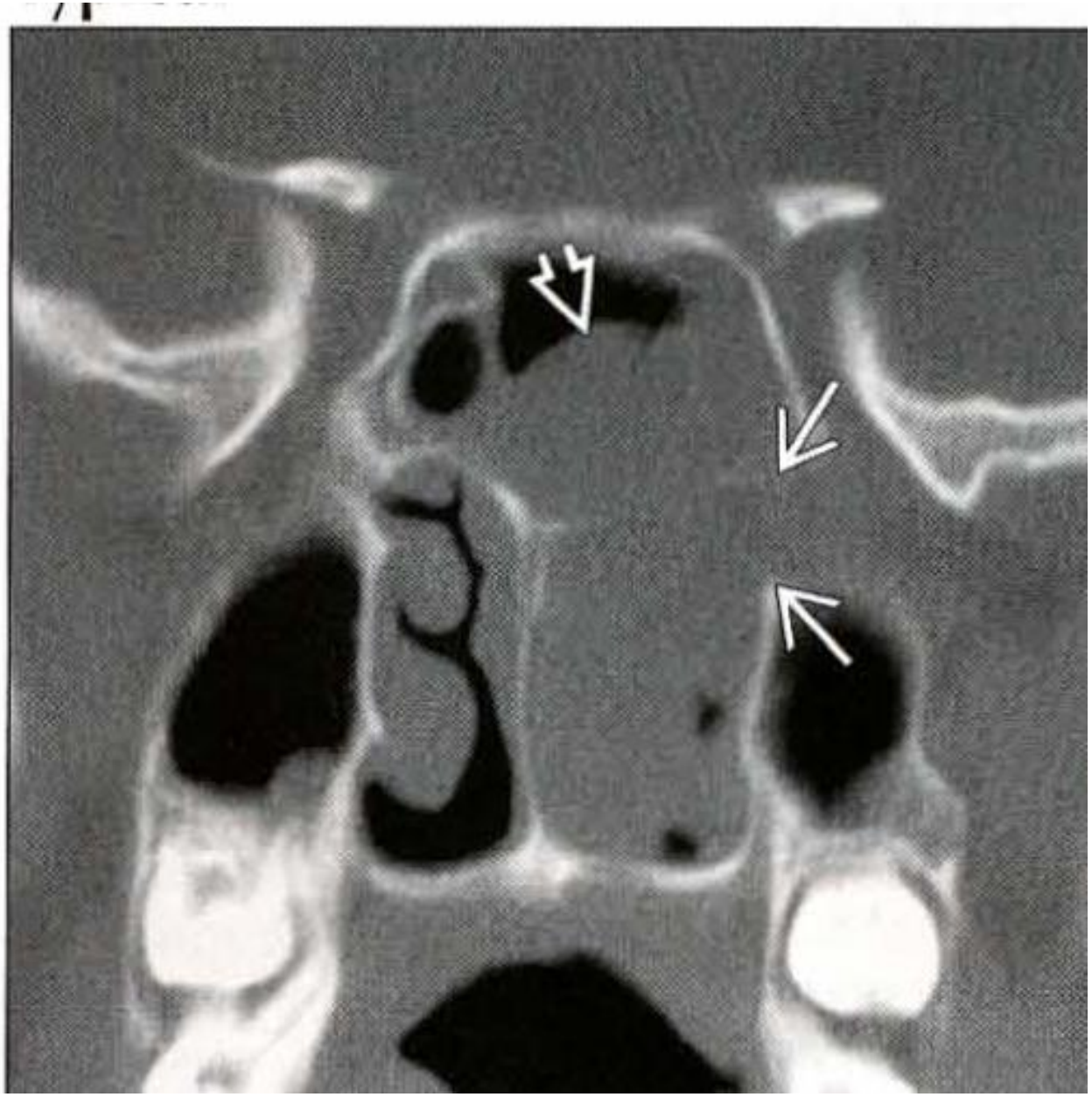
# Fisch staging system of juvenile angiofibromas.

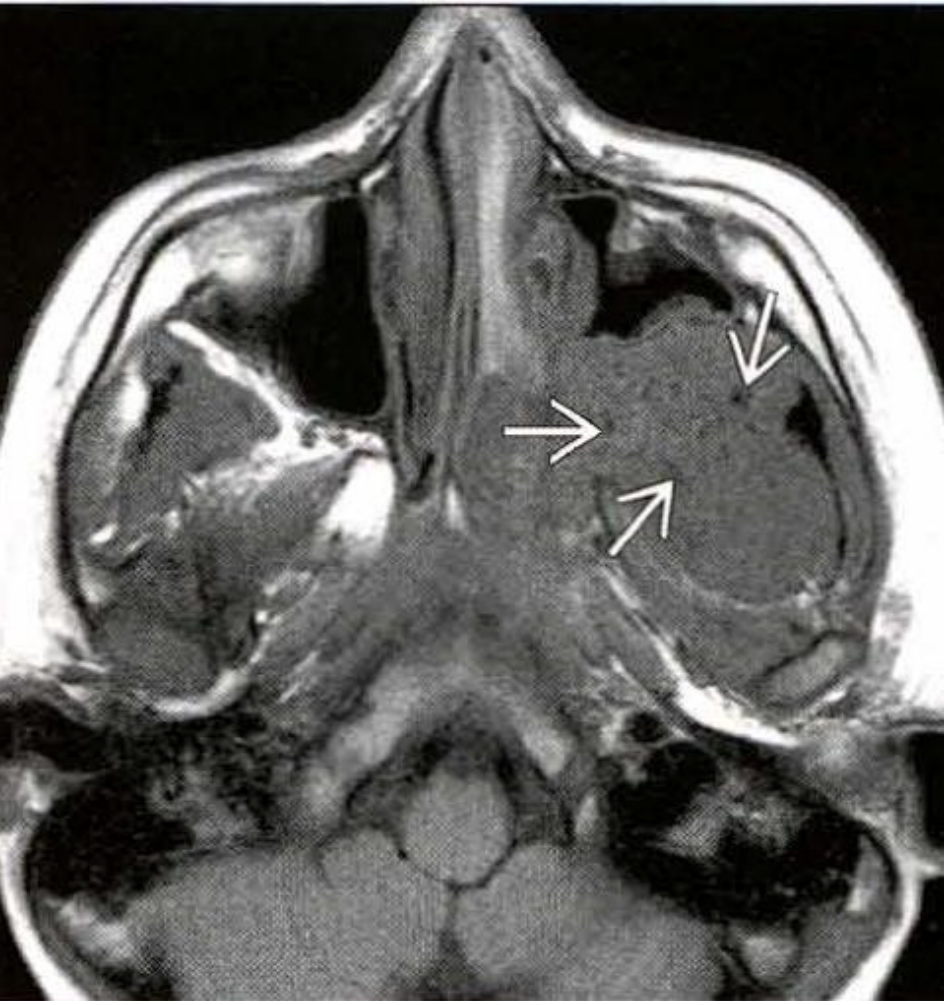
1 Tumour limited to the nasopharyngeal cavity; bone destruction negligible or limited to the sphenopalatine foramen

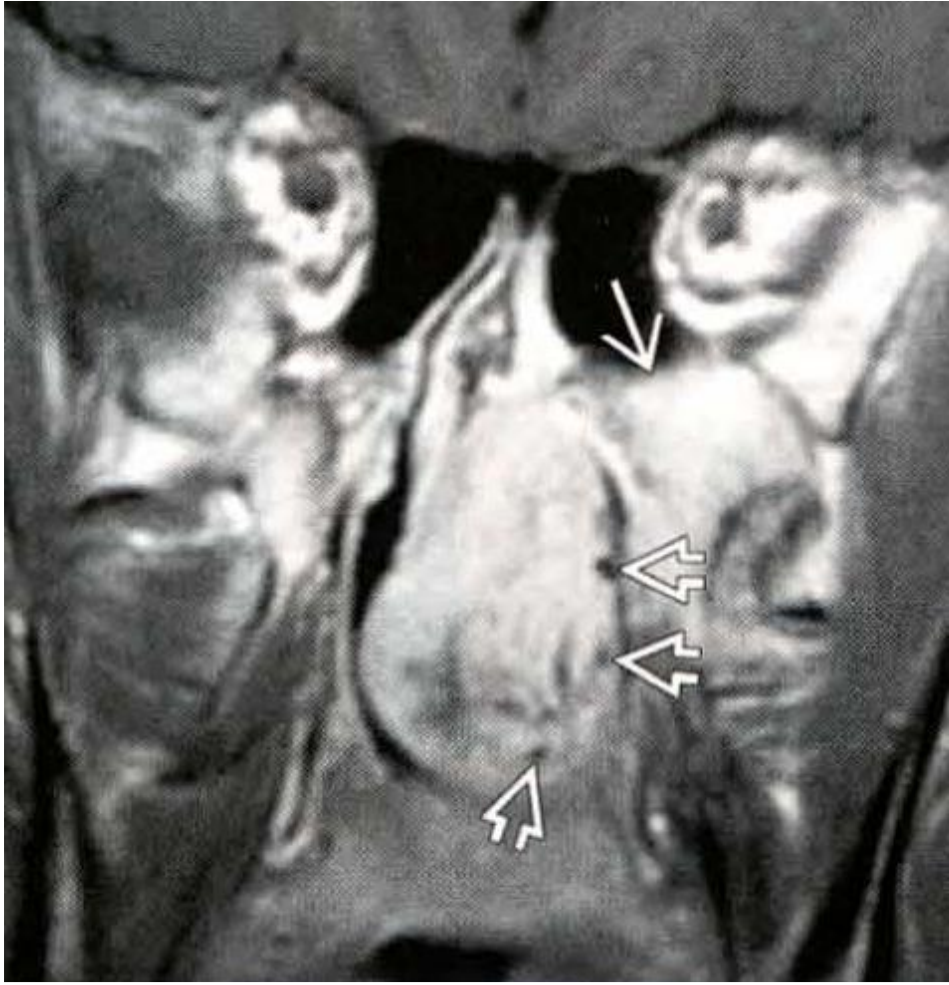
2 Tumour invading the pterygopalatine fossa or the maxillary, ethmoid or sphenoid sinus with bone destruction

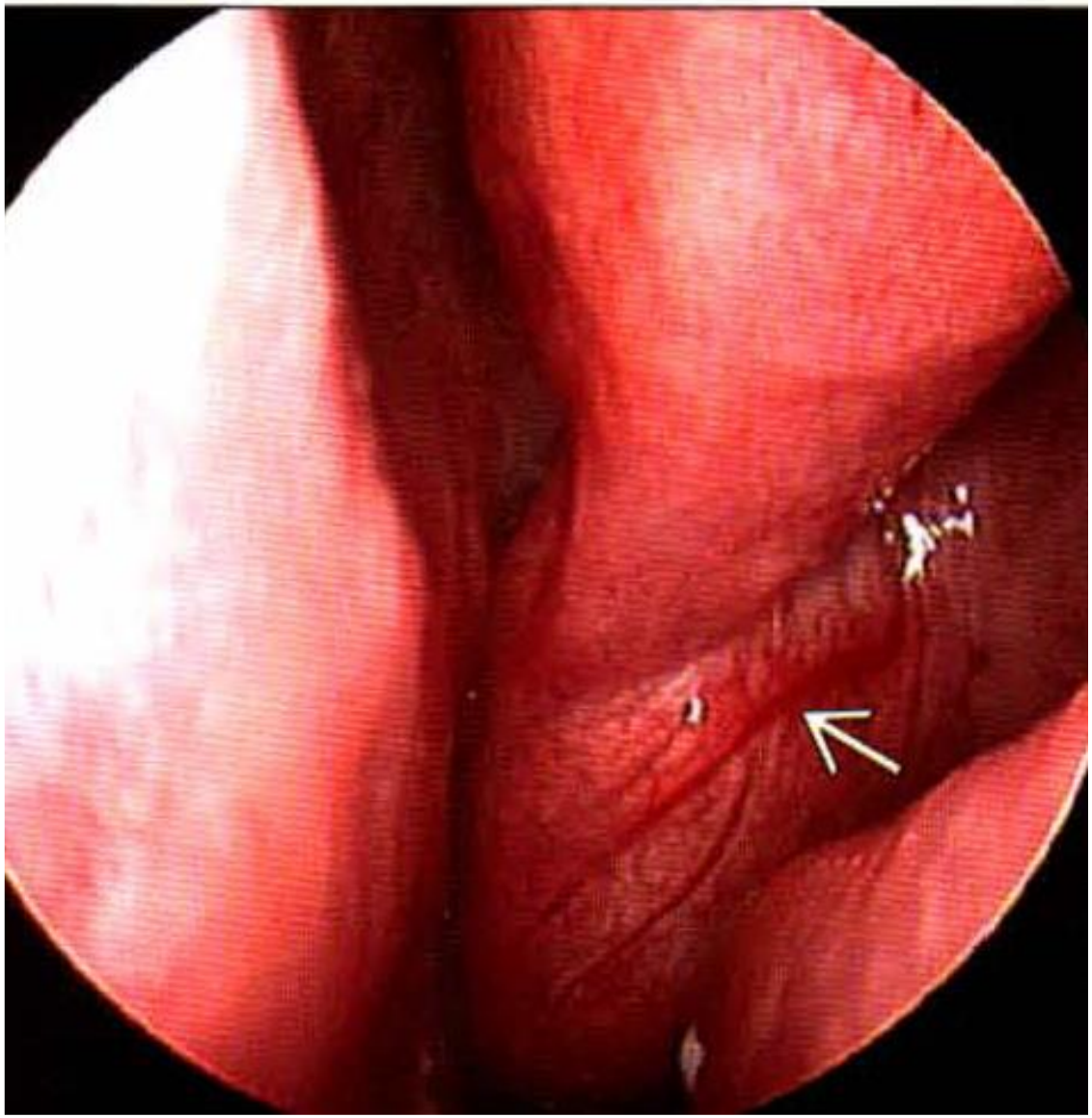
3 Tumour invading the infratemporal fossa or orbital region:
(a) without intracranial involvement
(b) with intracranial extradural (parasellar) involvement

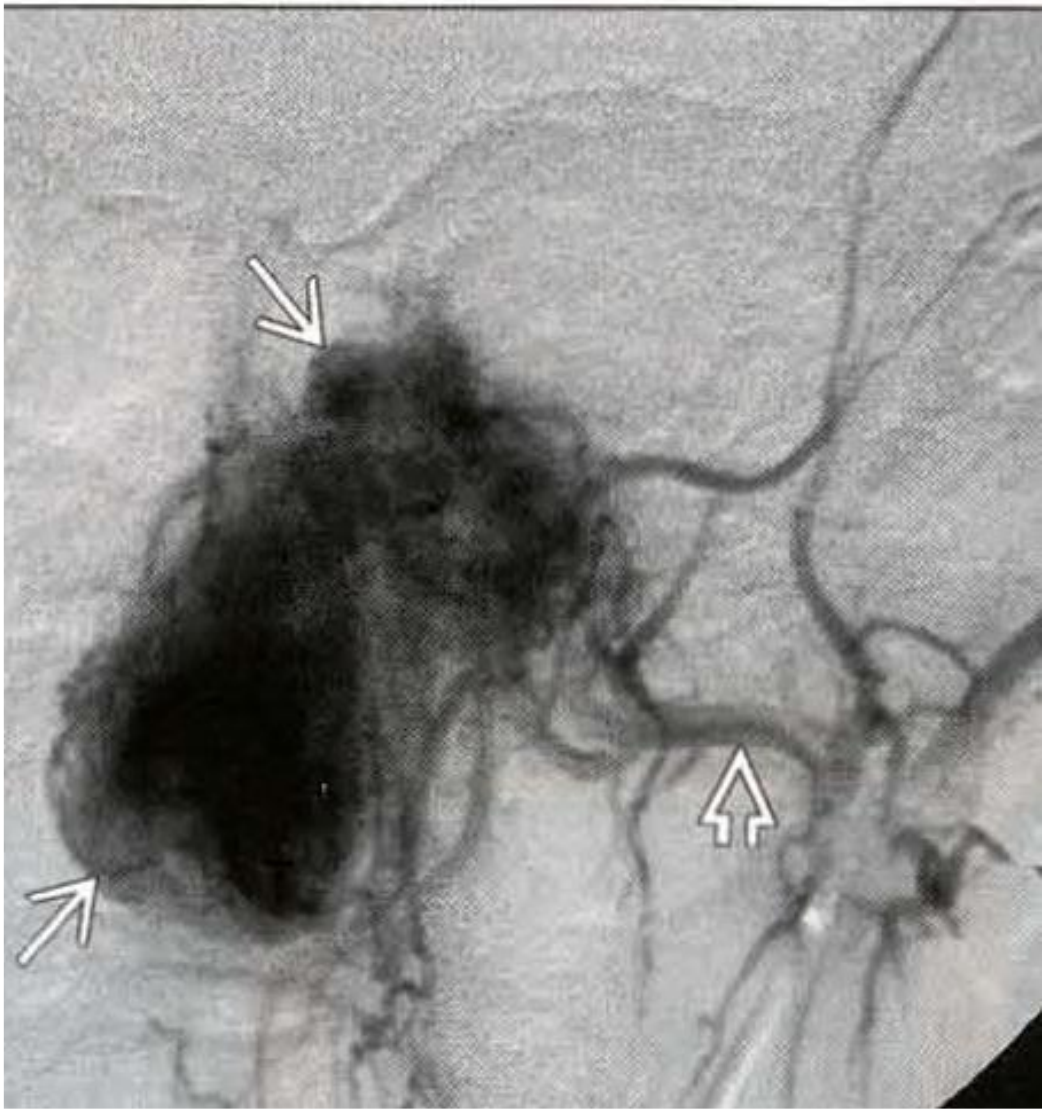
4 Intracranial intradural tumour:
(a) without infiltration of the cavernous sinus, pituitary fossa or optic chiasm
(b) with infiltration of the cavernous sinus, pituitary fossa or optic chiasm











# TREATMENT

## PREOPERATIVE EMBOLIZATION

The role of preoperative embolization in the surgical management of this tumour is controversial. the blood supply is predictable, usually the terminal branches of the internal maxillary artery, and these can be controlled easily at the time of surgery.

Embolization in such cases would seem to be unnecessary.

More extensive tumours acquire a blood supply from other vessels, branches of both the external and internal carotid circulations.

More extensive tumours acquire a blood supply from other vessels, branches of both the external and internal carotid circulations. Surgical resection of these tumours can be formidable and preoperative selective embolization, some days before surgery, is prudent at the very least.

# PREOPERATIVE CHEMOTHERAPY

Oestrogens have been reported to induce shrinkage in some but their effect is variable and not without complications. At the very least, oestrogen therapy delays surgery and the secondary feminizing effects are certainly unwanted by an adolescent boy. In a small series of patients given the nonsteroidal androgen receptor blocker, flutamide, tumour shrinkage of up to 44 percent was reported.

# SURGICAL RESECTION - TECHNIQUES AND APPROACHES

## APPROACHES

most small tumours were resected either through a transpalatal approach, lateral rhinotomy or mid-facial degloving approach.

Open approaches can be used for tumours of all stages and certainly were the only option before the application of endonasal endoscopic techniques became more widespread.

Nowadays, stage Fisch I, 2 and some type 3 tumours are suitable for endoscopic resection using one or two surgeon techniques.

There is much to be gained by endonasal endoscopic techniques, for example, reduced intraoperative blood loss, fewer postoperative complications and a reduced length of hospital stay.

## Endoscopic endonasal techniques

Preoperative embolization is usually undertaken,. The anterior end of the middle turbinate is resected at the outset of the procedure.

An anterior ethmoidectomy together with removal of the medial wall of the maxillary sinus gives access to the posterior wall of the antrum. This is then removed to achieve complete lateral exposure of the tumour .Dissection then continues into the sphenoid until its rostrwn is reached following which the tumour can be peeled inferiorly

Throughout this process it is necessary to use bipolar diathermy and ligaclips to control the feeding blood vessels. The use of a second surgeon

## Open approaches

Transpalatal and lateral rhinotomy approaches have largely, but not completely, given way to mid-facial degloving and infratemporal approaches that were popularized in the 1980s. Most surgeons now have adopted the technique of mid-facial degloving for the resection of juvenile angiofibromas. Using the exposure afforded by this approach, the anterior, medial, lateral and posterior walls of the maxillary antrum can be removed.. Extensions into the inferior part of the orbit and infratemporal fossa can also be removed .

Extensive juvenile angiofibromas are better resected through skull base approaches, preferably undertaken by a combined otorhinolaryngological and neurosurgical team.

The components of tumour in the cavernous sinus and any intradural disease demands adequate exposure and this can only be achieved with a subtemporal pre-auricular infratemporal fossa approach, usually combined with a modified middle fossa craniectomy.

# Radiotherapy

External beam radiation was delivered in several fractions to achieve a total tumour dose of 30-55 Gy.
All series report that regression of angiofibromas after radiotherapy is very slow indeed, often taking two to three years before radiological stabilization' is achieved.

Local control rates of 80-85 percent have been achieved as assessed by clinicalexamination..

Treatment failure was apparent, usually within the first two to three years, and surgical salvage was generally successful in all these patients. There are no reports on the efficacy of gamma-knife therapy as yet.