Rhinology, 28, 97-102, 1990

Recurrence in juvenile angiofibroma

Andrew McCombe, Valerie J. Lund and David J. Howard

Institute of Laryngology and Otology, London, United Kingdom

SUMMARY

A potential for recurrence of juvenile angiofibroma exists after all treatment modalities, both surgical and medical but the methods of defining recurrence and failure to cure varies considerably from series to series. To evaluate factors which might influence successful treatment, a series of 33 patients have been reviewed retrospectively. All patients were treated by simple or extended lateral rhinotomy as a primary or secondary procedure. The final long-term disease control rate was 97% but during the treatment period the overall symptomatic recurrence rate was 50%. However, amongst those treated primarily the recurrence rate was 34%. Of the factors examined, the strongest predictor of recurrence was preoperative embolisation. This group exhibited both early and multiple recurrence when compared with the non-embolised group and the possible reasons for this are examined.

INTRODUCTION

Juvenile angiofibroma is a benign tumour composed of variable proportions of vascular stroma within a fibrous framework and affects young males often presenting at the time of puberty. It arises from the region of the sphenopalatine foramen and medial pterygoid plate, and enlarges initially into the nasopharynx and later may expand to fill the sphenoid sinus, infratemporal fossa, posterior orbit and ultimately middle cranial fossa (Harrison, 1987). Tumours are staged according to the Chandler classification by their degree of local spread (Chandler et al., 1984). Many groups in the literature recommend an extensive pretreatment work-up including angiography and CT scanning to accurately stage these tumours. Two main treatment modalities are then available; most groups favour a primary surgical approach (Waldman et al., 1981; Witt et al., 1983; Bremer et al., 1986; Economou et al., 1988) (the chosen approach depending on the tumour staging), although good results have been reported from Toronto where radiotherapy is the preferred primary treatment (Cummings et al., 1984). Management is obviously fraught with problems; access to the tumour is often difficult, disfiguring surgery is unwelcome for essentially benign disease and radiotherapy at such a young age must pose questions for the future.

Table 1. Recurrence rates in literature

	recurrence rates			
author	treatment	patients	overall recurrence	recurrence after treatment at centre
Waldman, 1981	surgery	10	0%	0%
Bremer, 1986	surgery	30	30%	17%
Economou, 1988	surgery	82	34%	25%
Witt, 1983	surgery	31	55%	30%
Cummings, 1984	radiotherapy	55	N/A	20%

Despite the various treatments available, a marked potential for recurrence exists in juvenile angiofibroma. The literature quotes rates from 0–55% although patient numbers range from 10 to 82 (Table 1). It is apparent that recurrence in this condition is not infrequent and with this in mind a series of 38 patients were reviewed with emphasis on any factors which might predict postoperative recurrence.

METHOD

A series of 38 patients with a histological diagnosis of juvenile angiofibroma were analysed retrospectively over a 10 year period between 1978 and 1988.

Full information was available on 33 patients. All 33 were treated under the care of Professor D. F. N. Harrison either primarily or secondarily for recurrence after primary treatment elsewere.

Information recorded included age at presentation, duration of history, tumour size, staging and histology, preoperative interventions and surgical approach. Any other treatment modalities were recorded. The presence and time to recurrence were noted. Recurrence in this series was defined as the presence of disease after treatment regardless of the presence or absence of symptoms.

RESULTS

Thirty-three male patients were analysed, the average age at presentation being 14.5 years (range 8–28 years).

The preferred mode of treatment was surgical and a lateral rhinotomy or "extended" lateral rhinotomy (Weber-Fergusson) were the only surgical approaches used in 38 operations. Four patients referred from elsewhere had five previous transpalatal approaches. One patient had had a Caldwell-Luc approach as a secondary procedure. Seven of our patients were given radiotherapy for intracranial tumour extension or recurrent disease following several surgical procedures.

Recurrent disease, that is the presence of disease after primary treatment, occurred in 17 cases. Of these 17 cases, six were referred from other centres with recur-

rence after primary treatment. Thus the overall disease recurrence rate was 53% but recurrence after primary treatment at the Institute occured in 11 cases (33%). The average time to develop symptoms of recurrence was 19 months and five patients developed more than one recurrence.

In considering preoperative factors which may contribute to recurrence, it was found that preoperative embolisation had been performed in six patients and all six patients developed recurrence of tumour. Five of the six developed symptoms of recurrence and did so, on average, within 10 months of treatment. This compares with an average time to symptoms in the rest of the recurrent group of 24 months. This difference is statistically significant ($p < 0.05$).

Recurrence was also more likely if the tumour was large (stage III or IV) than if it was small (stage I or II) at the time of treatment (Table 2).

Table 2. Relationship of recurrence to staging of angiofibroma by Chandler classification

tumour staging and recurrence		
tumour	stageno. of cases	no. of recurrences
I	7	0
II	7	3
III	10	6
IV	3	2

Despite this there was no greater predominance of large tumours in the embolised subgroup.

The average age of the recurrent group was 14 years and of the embolised subgroup was 13.8 years and there was no difference in the histological features of the tumours in the various groups.

Five patients had more than one recurrence of whom three had had preoperative embolisation. One patient had had two previous transpalatal approaches and one had had a primary lateral rhinotomy and a secondary Caldwell-Luc. Only one patient had three recurrences and he had received initial preoperative embolisation.

One patient is asymptomatic with known static residual disease and another patient is currently having radiotherapy for a massive intracranial recurrence. Thirty-one patients are symptom-free and are not known to have recurrent or residual disease. Thus the overall control rate is 97% (32/33) with follow-up times ranging from 5 months to 11 years (average 5 years).

DISCUSSION

Although thorough preoperative investigation is the rule in these patients, not unnaturally postoperative investigations are reserved for those who develop

symptoms of recurrence. As it is well-established that patients with disease may be asymptomatic, the diagnosis of recurrence must be inaccurate.

In addition to the relatively uncommon nature of this condition, treatment is often concentrated in "specialist centres", and referral to these centres after the failure of primary treatment. Analysis of the results presented by the "specialist centres" shows that quoted recurrence rates are those following treatment at the centre and do not take account of the initial recurrence with which the patient was referred (Table 2). This obviously presents misleadingly low recurrence rates (Waldman et al., 1981; Witt et al., 1983; Bremer et al., 1986; Economou et al., 1988).

It would seem unlikely that if a benign neoplastic tumour is completely excised, one should get a de novo recurrence at the same site. It would seem much more plausible that what is currently called recurrence is in fact regrowth of residual disease. This series clearly shows that recurrence with this disease is common. All studies have shown that recurrence is more likely with a larger tumour which is no surprise in view of the previous hypothesis; with a large tumour in this difficult area it is more likely that there will be residual tumour left at operation (Waldman et al., 1981; Witt et al., 1983; Bremer et al., 1986; Economou et al., 1988).

In this series preoperative embolisation is associated with both rapid and frequent recurrence. In the absence of any other differences between the two subgroups of recurrent disease, it is not unreasonable to think that they may be causally related. If one accepts that residual disease is the cause of recurrence, it is easy to postulate that embolisation shrinks the tumour but in doing so makes complete removal more difficult and the amount of residual tissue is larger than if embolisation is not used.

The natural history of this uncommon condition is far from clear. Spontaneous regression after puberty has been suggested but the evidence for this is scanty; to date only one documented case exists in the English literature (Jacobson et al., 1989). It would seem that if after treatment a small enough volume of disease remains, further growth ceases or is minimal such that the patient remains asymptomatic and a "cure" is achieved (Stansbie and Phelps, 1986). Ninety-seven per cent of our patients have reached such a symptom-free state regardless of the number of treatments required, with follow-up times ranging from 5 months to 11 years.

However, experience of one patient who developed symptoms of recurrence only after the tumour had invaded the middle cranial fossa and the knowledge that recurrence may be asymptomatic for a considerable time has led to the conclusion that regular CT scanning is justified. The demonstration of residual disease is not a sine qua non for surgery but would alert the clinician to the possibility of aggressive disease thereby enabling closer monitoring to pre-empt massive

intracranial spread and would also provide a greater understanding of the natural history of this disease. Only by such methods will the true incidence of recurrence become apparent.

It has recently been demonstrated that magnetic resonance tomography is the imaging modality of choice in these patients (Lund et al., 1989). In treating this condition it is important to know both the extend and vascularity of these tumours. Both of these factors are demonstrated very elegantly by MRT and this is done in one procedure, without the attendant morbidity of angiography or exposure to ionising radiation. Consequently angiography is redundant as a diagnostic and therapeutic modality.

At the present time and with our current knowledge of this condition, one can argue that the procedure which allows the greatest access to effect a thorough and controlled surgical excision is the safest and most appropriate treatment to avoid frequent and rapid recurrence. These objectives combined with good control rates have been achieved by the lateral rhinotomy approach alone.

It is therefore recommended that preoperative embolisation be abandoned in the light of the frequent and rapid recurrence found in our patients and that the lateral rhinotomy be used to provide optimal access to this difficult disease site.

ACKNOWLEDGEMENTS

We would like to thank Professor D. F. N. Harrison for allowing us to discuss his patients.

REFERENCES

1. Bremer JW, Neel HB, DeSanto LW, Jones GC. Angiofibroma: treatment trends in 150 patients during 40 years. Laryngoscope 1986; 96: 1321–1329.
2. Chandler JR, Goulding R, Moskowitz L, Quencer L. Nasopharyngeal angiofibroma: staging and management. Ann Otol Rhinol Laryngol 1984; 93: 322–329.
3. Cummings BJ, Bland R, Keane T, et al. Primary radiation therapy for juvenile nasopharyngeal angiofibroma. Laryngoscope 1984; 94: 1599–1605.
4. Economou TS, Abemayor E, Ward P. Juvenile nasopharyngeal angiofibroma: an update of the UCLA experience, 1650–1985. Laryngoscope 1988; 98: 170–175.
5. Harrison DFN. The natural history, pathogenesis and treatment of juvenile angiofibroma. Arch Otolaryngol Head Neck Surg 1987; 113: 936–942.
6. Jacobsson M, Petruson B, Ruth M, Svendson P. Involution of juvenile nasopharyngeal angiofibroma with intracranial extension. Arch Otolaryngol Head Neck Surg 1989; 115: 238–239.
7. Lund VJ, Lloyd GAS, Howard DJ. Juvenile angiofibroma: imaging techniques in diagnosis. Rhinology 1989; 27: 179–185.
8. Stansbie JM, Phelps PD. Involution of residual juvenile nasopharyngeal angiofibroma. A case report. J Laryngol Otol 1986; 100: 599–603.
9. Waldman SR, Levine HL, Astor F, Wood BG, Weinstein M, Tucker HM. Surgical experience with nasopharyngeal angiofibroma. Arch Otolaryngol 1981; 107: 677–682.

10. Witt TR, Shah JP, Sternberg SS. Juvenile nasopharyngeal angiofibroma: A 30 year clinical review. Am J Surg 1983; 146: 521–525.

Valerie J. Lund, M.S., F.R.C.S.
Professorial Unit
Institute of Laryngology and Otology
330 Gray's Inn Road
London WC1X 8DA
United Kingdom