Juvenile Angiofibroma : Review of a Case Series and Role of Embolization

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Here 15 cases of advanced Juvenile angiofibroma are presented. Advantages of wide tumour exposure and preoperative embolization are discussed.

INTRODUCTION

Juvenile Angiofibroma is now a very well known entity. It has a very special place in E.N.T. surgery on account of being highly vascular. All along E.N.T. surgeons efforts have been directed towards reducing the vascularity of the tumour to reduce blood loss during surgery thereby enabling its complete removal. Hormones are one of the effective methods of reducing the tumour vascularity, however the tumour embolization is the best method of doing it in modern E.N.T. practice. Another important development has been the availability of better imaging techniques viz. Computerized Tomography (CT), Magnetic Resonance Imaging (MRI), Digital Subtraction Angiography (DSA) which provide better understanding of skullbase anatomy, its surgical approaches and tumour vascularity.

MATERIAL AND METHODS

We had 15 cases of advanced juvenile angiofibroma referred to us in last 3 years. These cases after clinical examination were first subjected to plain and contrast CT study (axial and coronal cuts) to assess the extent of the lesion and then tumour was staged according to Chandler et al (1984) classification, Stage I – Tumour confined to nasopharynx, Stage II – Tumour extending into nasal cavity and/sphenoid sinus, Stage III – Tumour extending into one or more of the following : maxillary antrum, ethmoid

sinuses, pterygomaxillary and infratemporal fossae, orbit and/or cheek and Stage IV – Tumour extending into cranial cavity.

DSA was done by Digital Subtraction Angiography Unit (DAR 1200 Shimadzu) to see the feeding vessels and angioarchitecture of the tumour. Embolization was then carried out by superselective catheterization with Tracker 18 microcatheter system. Distal internal maxillary artery and if necessary ascending palatine branch of facial artery on one or both sides were embolized (Fig. 6 & 7). For embolization polyvinyl alcohol foam (PVA), gelfoam fine shreds and small pieces were used. These cases were then operated at the earliest and in no case surgery allowed to extend 4 days after embolization. Cases of stage II, III and one case

Fig.1

Coronal contrast enhanced C.T. of sphenoidal region shows homogeneously enhancing mass lesion occupying left choana with deviation of nasal septum. Bone destruction to sphenoid sinus and intracranially in medial basi-temporal region is seen

of stage IV where intracranial extension was small were operated in a single stage even though more than one surgical approach was needed. However two stage procedure was done wherever intracranial extension was large. One patient (Case No. 6) was not operated as he had a very large intracranial tumour and most of the blood supply was from internal carotid system. He was subjected to radiotherapy by Cobalt-60. The peroperative blood loss was measured from suction bottle and soaked gauze packs. Postoperative CT study was done at one month and one year interval.

OBSERVATION

We had 15 cases of Juvenile Angiofibroma in last 3 years. One patient was of stage II, 8 were of stage III and 6 were of stage IV. Most of them (i.e. 13 cases) had been operated one to four times before they came to us. Fourteen of these cases were referred to us after diagnosis was established by other E.N.T. surgeons.

Epistaxis was mode of presentation in all cases, 12 patients complained of nasal obstruction, two had cheek swelling, one had diplopia and visual

After CT study pterygomaxillary fossa was found to be involved in 14 cases. Tumour had extended here through sphenopalatine foramen in four cases and in remaining ten cases extension was through the pterygomaxillary fissure. Intracranial extension was present in 6 cases, in 4 of them tumour had eroded the lateral wall of sphenoid sinus and extended intracranially, in another two cases tumour had extended intracranially, through the foramen lacerum (Fig. No. 3). In both cases with orbital involvement,

Fig.3

Coronal contrast C.T. scan of sphenoidal region showing homogeneously enhancing mass lesion occupying nasopharynx, sphenoid sinus and middle cranial fossa through a large defect in its floor

Fig.2

Post-operative Coronal C.T. shows complete removal of the mass lesion and normal pneumatization of sphenoid sinus

impairment was present in one case. In no case any symptom suggestive of intracranial extension was present.

orbital extension had occurred through inferior orbital fissure, in one of them tumour was also extending into superior orbital fissure. Here middle cranial fossa extension had grown anteriorly medial to optic nerve filling up the superior orbital fissure (Case No. 15). In one patient (Case No. 1) intracranial extension was removed through transpalatal approach (Fig No. 1 & 2). Other cases (2, 13, 14 & 15) were operated in two stages, in stage one intracranial part of tumour was removed by combination of transpalatal, retromolar and transmaxillary approaches. Then after a period of 1-2 weeks modified pterional craniotomy in the form of frontotemporal craniotomy and orbitotomy was

Fig. 4

C.T. scan of the same patient (Fig. No. 3) after transpalatal removal of the nasopharyngeal part.

done to remove intracranial extension. In our series preoperative blood loss ranged from 500 ml. to 2500 ml. (average 1450 ml.) Tumour removal was complete in 13 cases. Only one patient showed small tumour in cavernous sinus area on CT scan one year after surgery (Case No. 4).

DISCUSSION

Preoperative radiological evaluation was very helpful to us in tumour assessment and in planning surgery. All of our cases needed more than one surgical approach since they had large tumours involving more than one region. This has been experience of others also (Standefer et al 1983). We have found transpalatal approach with retromolar extension very useful for removing nasopharyngeal and pterygomaxillary fossa extensions. We could remove even small intracranial extensions through this approach (Fig. Nos. 1 & 2). Larger pterygomaxillary fossa and small infratemporal extensions were removed through a transantral window by removing anterolateral wall and posterior wall of maxillary antrum in continuity through a separate sublabial incision.

Tumours with large intracranial extension were first operated for extracranial part and then after

a period of about 1-2 weeks intracranial portion of tumour was removed through a unilateral frontotemporal craniotomy. A bicoronal skin incision was made behind the hair line starting from 1 cm. anterior to tragus below the root of the zygoma to opposite temporal line. Flap was then elevated and reflected anteriorly in the areolar plane. After dissecting the superficial

Fig. 5

C.T. scan of the same patient (Fig. No. 3 & 4) after removal of mass from sphenoid sinus and intracranial part through a pterional craniotomy. A part of sphenoid sinus floor was removed for post-operative drainage

temporal fascia, the deep temporal fascia was incised along the zygoma and the orbital rim and temporal muscle was reflected down. A unilateral frontotemporal craniotomy was performed followed by unilateral orbitozygomatic osteotomy (Samii et al, 1989). The lesser and greater wings of sphenoid, orbital roof and its lateral wall and the middle fossa base were drilled away to expose the Superior orbital fissure, V₃ and V₂ nerves. The intracranial part of the tumour was then exposed, dissected and freed from dura, it was then removed along with the residual tumour of sphenoid sinus wherever it was present. The defect in skull base was repaired by temporalis muscle and pericranium (Fig. Nos. 3, 4 & 5). Similar procedure for removing intracranial extension in middle cranial fossa was described by Lyons and Donald (1992), where a lateral infratemporal/middle fossa approach was used. The approach adopted by us gives a better

view of skullbase because we had performed orbitozygomatic osteotomy. Resection of part of temporal lobe was not necessary even when tumour was large (Fig. No. 3).

A good tumour exposure before its removal reduces the tumour removal time and hence the blood loss. We are also in favour of wide tumour exposure and incising periosteum around the tumour attachment on skullbase as far as possible and then elevating the healthy periosteum in

Fig.6

Lateral view of selective left external carotid angiogram shows a vascular tumour supplied by branches from distal internal maxillary artery

continuity with the tumour, this helps in complete and easy removal of tumour. Similar observation was made by Maharaj and Fernandes (1989). We could achieve complete tumour removal in 13 out of 14 operated cases. In 10 cases tumour removal was done in one stage and in another 4 cases who had large intracranial extension second stage surgery was done for the removal of intracranial part of the tumour. One of these patients (Case No. 4) has shown a small tumour in the floor of middle cranial fossa in a follow up CT scan one year after surgery though his preoperative and first postoperative CT scan had not shown any tumour in that region. Embolization was very effective in our cases since we could minimize the blood loss to 500-2500 ml. (average 1450 ml.). Four of our patients

Fig.7

Post-embolization superselective internal maxillary artery angiogram-lateral view shows significantly reduced vascularity and tumour stain and internal maxillary artery branches which are distally occluded. A normal filling middle meningeal artery is seen

(Case Nos. 2, 5, 10 & 13) who had blood loss of 2000 ml. and 2500 ml. had blood supply to the tumour from internal carotid system also (Fig. No. 8) which was not embolized which explains more blood loss (Table Nos. II & III). In another series with similar large tumours, where embolization was not done average blood loss was 3000 ml. (Duvall et al 1987). There are reports of blood loss exceeding 2000 ml. even after embolization is also important since gelfoam particles tend to dissolve and disintegrate after 5-7 days, development of collateral circulation after embolization too has to be kept in mind.

One more effective way of reducing the tumour vascularity is hormone therapy. Preoperative hormone therapy by estrogen is no more recommended for obvious side effects, however testosterone blocker is still used by some (Kuttner et al 1977 and Gates et al 1992). But they have the disadvantage of prolonged administration before they can be effective. Estrogens have added disadvantage of feminising the patient and responses of reducing vascularity can be variable, therefore embolization is the most effective method of reducing the tumour vascularity.

Table - I

Case No.	Age (in Yrs)	Duration of Symptoms (in Yrs.)	No. of Prev. Surg.	Extent		Stage
				Clinical	Radiological	
1.	24	2.0	01	NP+NC	NP+NC+SP+PF+IF+MC	IV
2.	12	2.5	03	NP	NP+NC+SP+PF+MC	IV
3.	20	1.0	02	NP	NP+NC+SP+PF	III
4.	16	2.0	04	NP+CH	NP+NC+SP+PF+IF+CH	III
5.	24	4.5	02	NP+NC	NP+NC+SP+PF	III
6.	30	2.0	01	NP+NC	NP+NC+SP+PF+MC	IV
7.	14	1.0	02	NP+NC	NP+NC+SP+PF	III
8.	14	2.5	03	NP+NC	NP+NC+SP	II
9.	18	2.5	01	NP+NC	NP+NC+SP+PF	III
10.	15	2.0	02	NP+NC	NP+NC+SP+PF	III
11.	14	2.0	01	NP+NC	NP+NC+SP+PF	III
12.	14	3.0	01	NP+NC	NP+NC+SP+PF+IF	III
13.	18	2.5	02	NP+NC	NP+NC+SP+PF+MC	IV
14.	11	3.0	00	NP+NC+OR	NP+NC+SP+PF+IF+OR+MC	IV
15.	12	4.0	00	NP+NC	NP+NC+SP+PF+MC	IV

Table showing clinical and radiological details of cases. Nasopharynx (NP), Nasal cavity (NC), Cheek (CH), Orbit (OR), Sphenoid Sinus (SP), Pterygomaxillary Fossa (PF), Infratemporal Fossa (IF) & Middle Cranial Fossa (MC)

Table - II

Case No.	Approach	Tumour Removal	Blood Loss	Followup	
				Status	Duration
1.	TP+RM+TM	C	1000ml	NR	3.0 years
2.	TP+PT*	C	2000ml	NR	2.0 Years
3.	TP+RM+TM	C	1500ml	NR	2.0 Years
4.	TP+RM+TM	I	1100ml	RT	1.5 Years
5.	TP+RM	C	2000ml	NR	1.0 Year
6.	NO	—	—	—	—
7.	TP+RM	C	1000ml	NR	1.0 year
8.	TP+RM	C	500ml	NR	1.0 Year
9.	TP+RM	C	1000ml	NR	9 Months
10.	TP+TM	C	2500ml	NR	9 Months
11.	TP+RM+TM	C	1500ml	NR	6 Months
12.	TP+RM+TM	C	1800ml	NR	6 Months
13.	TP+RM+TM+PT*	C	2000ml	NR	3Months
14.	TP+RM+TM+PT*	C	1200ml	NR	1 Month
15.	TP+RM+TM+PT*	C	1200ml	NR	1 Month

Table showing surgical approaches used, Preoperative Blood Loss and Followup Status, Transpalatal (TP), Retromolar (RM), Transmaxillary (TM), Pterional Craniotomy (PT), Surgery done in two stages(*), Not Operated (NO), Complete removal (C), Incomplete removal (I), No Recurrence (NR) & Residual Tumour (RT).

Fig.8

Lateral view of selective internal carotid angiogram shows tumour stain presumably supplied by cavernous branch of internal carotid artery

CONCLUSION

Preoperative detailed radioimaging techniques help in planning surgical approach. Preoperative embolization reduces the operative blood loss and therefore helps in complete tumour removal. Wide exposure of tumour and its removal with a rim of healthy periosteum in nasopharynx also helps in its complete removal and prevents

Table - III

Case No.	Feeding Vessels	Vessels Embolized	Surgical Interval (in days)
1.	IM(L)+MM(L)+AM(L)	IM(L)+MM(L)+AM(L)	02
2.	IM(R)+IM(L)+ICS(L)	IM(R)+IM(L)	01
3.	IM(L)	IM(L)	02
4.	IM(L)	IM(L)	02
5.	IM(L)+ICS(L)	IM(L)	01
6.	IM(R)+IM(L)	IM(R)+IM(L)	–
7.	IM(R)	IM(R)	04
8.	IM(R)+IM(L)	IM(R)+IM(L)	02
9.	IM(L)	IM(L)	01
10.	IM(R)+IM(L)+ICS(R)	IM(R)+IM(L)	01
11.	IM(R)	IM(R)	01
12.	IM(R)+IM(L)	IM(R)+IM(L)	02
13.	IM(R)+ICS(R)	IM(R)	02
14.	IM(R)+ICS(R)	IM(R)	03
15.	IM(R)+IM(L)+ICS(L)	IM(R)+IM(L)	02

Table showing vessels feeding the tumour as seen on DSA, vessels embolized and the interval between embolization and surgery. Internal Maxillary Artery (IM), Middle Meningeal Artery (MM), Accessory Meningeal Artery (AM) and Internal carotid system (ICS).

recurrence. Small intracranial extension can be removed through transpalatal approach.

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