Complete resolution of stage IIIB juvenile nasopharyngeal angiofibroma with radiation therapy

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Abstract Juvenile nasopharyngeal angiofibroma is a benign, locally aggressive, vascular tumour of adolescent males. Extension to intracranial cavity is not uncommon and presents difficulties in management. Here we report a patient who had Radkowsky stage IIIB lesion, with blood supply from internal carotid artery. In view of anticipated problems with surgery, the patient was treated with 30 Gy in 15# external beam radiotherapy. On follow up, tumour was noted to disappear gradually overtime and at 3 years complete disappearance was noted with normal return of vision. Hence we are re-affirming the earlier studies that angiofibroma mass regresses gradually after completion of radiotherapy. Radiotherapy is a useful way of treating angiofibroma with significant intracranial extension.

Keywords Angiofibroma · Intracranial extension · Radiotherapy

Introduction

Juvenile nasopharyngeal angiofibroma (JNA) is a histologically benign yet highly vascular tumor that primarily affects adolescent males. It accounts for 0.05–0.5 % of all head and neck tumors [1] and is considered the most common benign neoplasm of nasopharynx [2]. Although benign, the tumor may exhibit locally aggressive behaviour, eroding into adjacent structures and causing functional injury [1, 2]. Intracranial involvement has been reported to occur in 10–30 % of all cases with middle cranial fossa being the most common sites of intracranial extension [3, 4]. Significant morbidity and even death may occur from intra cranial extension or haemorrhage [5].

Surgery is the treatment of choice for lesions without intracranial extension but therapeutic management of JNA with intracranial extension still remains controversial [4, 6]. While some opine that such tumors can be safely resected, others argue that radiotherapy signifies the best form of treatment for such tumors. Both, either surgery or radiotherapy have been reported to achieve a control rate of 80% [4, 6]. Some surgeons even advocate partial resection and follow by radiating the residual tumour [7]. However, this seems inappropriate since recurrence may occur from recognized residual tumour anywhere in the tumour bed which would not be irradiated by such an approach [6].

We are reporting a patient of JNA with intracranial extension (Radkowsky Stage IIIB) who achieved a complete remission with external beam radiotherapy.

Case study

A 18-year-old male was admitted in July, 2002 with chief complaint of single episode of spontaneous self limiting epistaxis. The patient also complained of nasal obstruction for 1year, progressive left eye proptosis for the previous 2 months, blurring of vision and diplopia on left lateral gaze and altered sensation over left side of face. On examination,

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Fig. 1 Characteristic pre-radiotherapy showing soft tissue mass expanding the pterygo-palatine fissure with bowing and erosion of the posterior maxillary wall, erosion of the block of sphenoid sinus and erosion of the base of the pterygoid plates

Fig. 2 (a) 5 months after completion of radiotherapy, scan shows no significant regression of the tumor mass. (b) 10 months after completion of radiotherapy, scan shows mark regression of the tumor mass

there was pink, fleshy mass seen to occupy the left nasal cavity, pushing the nasal septum to the right. There was a firm mass felt through left upper gingivobuccal sulcus in infratemporal fossa. Patient had proptosis and the left eyeball was pushed down and out.

On the basis of history and clinical examination a provisional diagnosis of Juvenile nasopharyngeal angiofibroma (JNA) was considered most likely and patient was taken up for imaging studies for definitive diagnosis and further management. CECT paranasal sinuses and head showed highly vascular soft tissue mass occupying left nasal cavity, maxillary antrum, ethmoid sinus, nasopharynx, pterygopalatine fossa, infratemporal fosa, sphenoid sinus and eroding the roof of sphenoid sinus with intracranial extension. Characteristic radiological signs to confirm the diagnosis included 1) expansion of the pterygo-palatine fissure with bowing and erosion of the posterior maxillary wall 2) erosion of the floor of the sphenoid sinus and 3) erosion of the base of the pterygoid plates. Digital Subtraction Angiography showed highly vascular mass supplied by maxillary artery, internal carotid artery and draining into superior ophthalmic vein, draining anterograde via angular vein.

On the basis of clinical examination and radiological investigations a definitive diagnosis of JNA, Radkowsky stage III B was made. Treatment options of surgery and radiotherapy were considered and discussed with the patient, and an informed decision was taken by the patient for treatment with radiation therapy. Radiotherapy was administered to a total dose of 30Gy in 15 fractions from 16-08-02 to 05-09-02.

Following completion of the Radiation therapy, the patient had no further episodes of epistaxis, although other symptoms persisted. A repeat CT Scan 1 month post treatment (Oct 2002) showed decreased enhancement of tumor, possibly due to radionecrosis; but no decrease in the size of the tumor. Subsequent sequential scans however demonstrated initial significant regression of tumor (6 months post treatment -March 2003), and subsequent

Fig. 3 Approximately 3 years after completion radiotherapy, the scans shows complete regression of the tumor mass

complete remission of tumour (30 months post treatment – April 2005). On last follow up (Dec 2006) the patient is completely asymptomatic and nasal endoscopy reveals no residual or recurrent mass in the nasal cavity or nasopharynx. The sequential scans are shown in Figs. 1, 2a, 2b and 3 with respective dates.

Discussion

Although radiation has been reported to be an effective measure of therapy for JNA [1, 5, 6, 7, 8, 9, 10, 11], potential long term complications have discouraged many from pursuing this course of therapy. Thyroid carcinoma, bone and soft tissue sarcomas, basal cell carcinoma, hypopituitarism, cataract, osteoradionecrosis, optic nerve, atrophy, osteomyelitis of skull base, facial growth retardation in maturing adolescents, all have been reported by various authors [6, 10–14].

Surgery is the alternate treatment for lesions with intracranial extension and is routinely practiced at our institution for such tumours. Our own experience with surgical treatment of intracranial extensions of angiofibroma has been safe but the literature reports of a reported mortality of at least 1 in 500³, and isolated reports have indicated a much higher mortality (20%) for advanced tumours with intracranial extension [15]. Further significant morbidity includes life threatening hemorrhage [15, 16], optic nerve injury [17], cranial nerve palsy [18], meningitis [15, 18], risks related to general anaesthesia, angiography and blood transfusion with related problems. Thus from the experience of various authors, both form of therapy carry significant risk and fatal complications. Complications of surgery being immediate and devastating in terms of morbidity and mortality while complications of radiotherapy follow a protracted course.

The case reported here demonstrates the complete regression of a massive angiofibroma with significant intracranial extension when treated with radiotherapy. However,

the process of regression was slow and completed only after more than 2 years. One source of confusion about effectiveness of radiotherapy as treatment for JNA arises from slow regression of this tumor after radiotherapy. The literature indicates that a residual but asymptomatic tumor is not necessarily uncontrolled and on continuing follow up, some of these residual masses may eventually disappear [6, 8, 18] This report reiterates that radiation therapy remains a viable option for the treatment of extensive JNA especially with significant intracranial extension of the disease.

Conclusion

With improvement in radiotherapeutic technique and corresponding reduction in its morbidity, radiotherapy is a possible option for angiofibroma with significant intracranial extension.

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