CASE REPORT

Massive juvenile nasopharyngeal angiofibroma: ode to the open surgical approach

Ravi Meher,¹ Nikhil Arora,¹ Eishaan Kamta Bhargava,² Ruchika Juneja¹

¹Department of ENT, Maulana Azad Medical College, New Delhi, India

²Department of ENT, Peterborough City Hospital, Peterborough, UK

Correspondence to
Mr Eishaan Kamta Bhargava,
eishaan@gmail.com

Accepted 2 July 2017

SUMMARY

The management of juvenile nasopharyngeal angiofibroma has undergone a significant evolution, with more surgeons moving towards the minimal invasive endoscopic approaches. Although considered the standard of care by most, an endoscopic approach may not be sufficient for extensive tumours, as exemplified by the current case of a young man presenting with the largest juvenile nasopharyngeal angiofibroma described in English literature until the present that was eventually excised via an anterior external approach.

BACKGROUND

The management of juvenile nasopharyngeal angiofibroma has undergone a significant evolution, with more surgeons moving towards minimally invasive endoscopic approaches.¹ Limited tumours with no significant intracranial, intraorbital and lateral infratemporal space extensions are best suited for endoscopic approaches. Though the endoscopic approach is considered the standard of care for removal of angiofibromas by most surgeons, on occasion there may be cases like ours where an open approach is necessary.

This case report discusses the challenges that may be faced by a surgical team when attempting removal of large angiofibromas such as this one, which, weighing in at 750 g, is the largest angiofibroma reported in English literature.

CASE PRESENTATION

An 18-year-old man, resident of Bihar, India, presented to the outpatient clinic with a right-sided cheek swelling of 6 months duration that had

Figure 2 MRI (postgadolinium enhancement) depicting a brilliantly enhancing homogenous mass extending superiorly until skull base (thin white arrow), inferiorly hanging below the mandible, posteriorly until the nasopharynx (black arrow), anteriorly extending into the soft tissue of the cheek (broken white arrow) medially pushing the septum to opposite side and laterally extending into infratemporal fossa (thick white arrow) and parapharyngeal space while abutting the masseter and temporalis muscles.

progressively increased in size to involve most of the right side of his face. He also had complaints of nasal obstruction and a nasal twang to his voice since the last 1 year, along with multiple episodes of unprovoked epistaxis that usually subsided spontaneously. He gave a history of a biopsy from the nose done in a different city, with no clear pathology report available at hand.

On examination of the face, a firm swelling of 20×15 cm dimension was seen on the right side of his face extending from the zygoma superiorly until the level of, and hanging beyond, the inferior border of the mandible, with obliteration of the nasomaxillary groove on the right side, along with a smaller firm swelling in the right temporal region extending anteriorly to the level of the right lateral orbital canthus and posteriorly up to the root of helix (figure 1). Mild proptosis of the right eye was also noted. Anterior rhinoscopy revealed a pinkish mass filling the right nasal cavity completely, extending up to the nasal vestibule, with the nasal septum deviated grossly to the left making it impossible to pass an endoscope to visualise the nasopharynx. Oral cavity examination revealed a diffuse bulge involving the hard and soft palate, almost completely obscuring the view of the oropharynx.

INVESTIGATIONS

Routine haematological and biochemical tests showed no abnormality.

Figure 1 Preoperative appearance, with right cheek swelling, temporal swelling and mild proptosis. The right side nasal mass can also be appreciated.

CrossMark

To cite: Meher R, Arora N, Bhargava EK, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2016-218731

Figure 3 Modified Weber-Ferguson incision.

Contrast enhanced CT and MRI scanning revealed a brilliantly enhancing homogenous mass extending superiorly until the skull base, causing thinning of the cribriform plate area, inferiorly hanging from the mandible, posteriorly until the nasopharynx, medially pushing the septum to the opposite side and laterally extending into the infratemporal fossa and parapharyngeal space while abutting the masseter and temporalis muscles, with a visualised widening of the sphenopalatine foramen. The tumour was also extending deep to the zygomatic arch to reach the temporal fossa. The maxilla on the right was significantly compressed, conceivably due to the pressure effect of the tumour and is indicated by an interrupted arrow on the axial scan. Orbital extension of the tumour can be discerned on the right side, through the floor of the orbit ([figure 2](#)). On MR angiography, feeding vessels were identified supplying the tumour from both the internal and external carotid system.

DIFFERENTIAL DIAGNOSIS

Considering the clinical and radiological features, a provisional diagnosis of juvenile nasopharyngeal angiofibroma was made,

Figure 4 Excised tumour specimen (2115 cm).

Figure 5 Postoperative appearance at follow-up.

keeping the possibility of a sinonasal malignancy as a differential diagnosis.

The patient's histopathology report was traced, confirming our provisional diagnosis of angiofibroma.

TREATMENT

Taking into consideration the size and extent of the tumour, it was decided that an external surgical approach via a modified Weber-Ferguson incision be performed. Unfortunately, preoperative embolisation could not be carried out, and the excision was done with hypotensive anaesthesia and a pre-excision operative control of the external carotid artery via the cervical approach. A modified Weber-Ferguson incision was fashioned with a lateral extension ([figure 3](#)), and a plane of dissection was developed between the tumour and skin. The tumour had a pseudocapsule, which was followed with blunt dissection separating it from the surrounding vital structures, namely the carotids, internal jugular vein, vagus nerve and skull base and was delivered in toto, measuring 21 x 15 cm in size and weighing 750 g, making it the world's largest juvenile nasopharyngeal angiofibroma removed till date([figure 4](#)). Postexcision, the sphenoid sinus and area of the sphenopalatine foramen were inspected endoscopically to confirm complete tumour removal. The estimated blood loss during the procedure (2200 mL) was managed with intraoperative fluid replacement with colloids and postoperative blood transfusion.

OUTCOME AND FOLLOW-UP

Following surgical removal, the patient remained under observation in the intensive care unit overnight and was discharged after 72 hours. The patient has been under regular follow-up for 18 months at present, has a well-healed scar with a cosmetic result that he finds acceptable ([figure 5](#)) and has developed no recurrence or cerebrovascular complications following surgery. Further plastic reconstructive surgery is being planned to improve the cosmesis.

DISCUSSION

Juvenile nasopharyngeal angiofibroma (JNA) is a rare fibrovascular tumour comprising only 0.5% of all head and neck tumours, most commonly affecting men in the prepubescent and adolescent age group. Histologically, the tumour comprises vascular and fibrous components with the vessels characteristically lacking the tunica media, hence the great propensity for haemorrhage. Although histologically benign, it may extend to adjacent tissues and cause bone destruction. Many theories have been proposed regarding its pathogenesis; however, the androgen dependence theory is the most widely accepted, which may explain why it afflicts the male sex. It was previously postulated that JNAs arise from posterosuperior lip of the sphenopalatine foramen, but recently it has been discovered that the vidian canal is usually the point of origin.² The most frequent symptom reported is epistaxis; however, due to a lack of education and awareness in various regions of rural India where JNA is prevalent, patients as well as their parents frequently overlook it. Diagnosis is usually based on a combination of a triad of clinical features (epistaxis, nasal obstruction and nasal mass) and radiological signs of a tumour causing widening of the sphenopalatine foramen, anterior bowing of the maxilla and a splaying of the pterygoid plates. Various therapeutic options have been used to treat JNA, namely surgery, radiotherapy and chemotherapy; surgical excision, however, remains the mainstay of management, with radiotherapy being reserved for certain cases with intracranial and intraorbital extensions. With advances in the field of endoscopy, endoscopic excision has emerged as the primary treatment modality for treatment of most JNAs.¹ The various advantages of less surgical time, absence of a scar and decreased risk of complications has led surgeons to prefer it over open surgical approaches.^{3–7} A large lateral extension of the tumour into the infratemporal and parapharyngeal space, however, makes an endoscopic approach not only technically difficult but also fraught with the risk of leaving residual tumour.⁸

In our case, massive lateral extension precluded any thoughts of an endoscopic approach, as was the case in a similar patient with a giant multicompartamental JNA reported by Koshy *et al*, which was excised via a extended osteoplastic maxillotomy approach.⁹

A further surgical challenge faced was the decision regarding preoperative embolisation due to feeding vessels arising from

both external and internal carotid circulations. The embolisation of complex lesions such as ours can be best described as a double-edged sword, with some authors promoting its use to decrease blood loss and others condemning it in view of the higher risk of cerebrovascular and ocular embolic complications and recurrence.^{10 11} We opted for a surgical control of the external carotids along with hypotensive anaesthesia to decrease blood loss, as has been described by those who have condemned embolisation.

Intracranial extension of angiofibroma presents yet another surgical challenge; some authors have adopted a combined intracranial and extracranial surgical approach to excision, while others advocate radiotherapy, as either a primary modality of treatment or as an adjunct to surgical excision to treat residual intracranial disease.^{7 8}

In summary, the primary aim of treatment is a complete excision of the tumour, and an open surgical approach may still be considered as a first-line approach for excision of JNA in a select group of patients where this primary aim may not be fulfilled by minimally invasive approaches, with adequate measures taken to minimise blood loss.

Contributors RM was the primary surgeon in the current case and, along with NA and EKB developed the learning points to be discussed. RJ wrote the first draft of the report, which was reviewed and edited by NA and the final draft approved by RM.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

© BMJ Publishing Group Ltd (unless otherwise stated in the text of the article) 2017. All rights reserved. No commercial use is permitted unless otherwise expressly granted.

REFERENCES

- Garofalo P, Pia F, Policarpo M, *et al*. Juvenile nasopharyngeal angiofibroma: comparison between endoscopic and open operative approaches. *J Craniofac Surg* 2015;26:918–821.
- Yüce S, Uysal İÖ, Doğan M, *et al*. A giant juvenile nasopharyngeal angiofibroma. *J Craniofac Surg* 2013;24:e207–e209.
- Pryor SG, Moore EJ, Kasperbauer JL. Endoscopic versus traditional approaches for excision of juvenile nasopharyngeal angiofibroma. *Laryngoscope* 2005;115:1201–7.
- Oliveira JA, Tavares MG, Aguiar CV, *et al*. Comparison between endoscopic and open surgery in 37 patients with nasopharyngeal angiofibroma. *Braz J Otorhinolaryngol* 2012;78:75–80.
- Nicolai P, Schreiber A, Bolzoni Villaret A. Juvenile angiofibroma: evolution of management. *Int J Pediatr* 2012;2012:1–11.
- Andrade NA, Pinto JA, Nóbrega MO, *et al*. Exclusively endoscopic surgery for juvenile nasopharyngeal angiofibroma. *Otolaryngol Head Neck Surg* 2007;137:492–6.
- Yiotakis I, Eleftheriadou A, Davilis D, *et al*. Juvenile nasopharyngeal angiofibroma stages I and II: a comparative study of surgical approaches. *Int J Pediatr Otorhinolaryngol* 2008;72:793–800.
- Mishra A, Mishra SC, Verma V, *et al*. In defence of transpalatal, transpalatal-circumaxillary (transpterygopalatine) and transpalatal-circumaxillary-sublabial approaches to lateral extensions of juvenile nasopharyngeal angiofibroma. *J Laryngol Otol* 2016;130:462–73.
- Koshy S, George M, Gupta A, *et al*. Extended osteoplastic maxillotomy for total excision of giant multicompartamental juvenile nasopharyngeal angiofibroma. *Indian J Dent Res* 2008;19:366–9.
- Janakiram N, Sharma SB, Panicker VB, *et al*. A drastic aftermath of Embolisation in Juvenile Nasopharyngeal Angiofibroma. *Indian J Otolaryngol Head Neck Surg* 2016;68:540–3.
- Tyagi I, Taneja HC, Phadke RV, *et al*. Juvenile angiofibroma: review of a case series and role of embolization. *Indian J Otolaryngol Head Neck Surg* 1998;50:20–5.

Learning points

- Preoperative assessment of the size and extent of the tumour is crucial for deciding the surgical approach to excise a juvenile nasopharyngeal angiofibroma (JNA).
- With large tumours with multiple extensions, a preoperative angiogram can help to plan adjunctive measures for minimising intraoperative blood loss.
- Complete excision of the tumour is crucial to prevent recurrence of greater importance in JNA due to younger age at presentation

Copyright 2017 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow