CASE REPORT

Stridor: an unusual presentation of juvenile nasopharyngeal angiofibroma

Hitendra Prakash Singh, Sunil Kumar, Madhukar Vashishtha, Satya Prakash Agarwal

Department of ENT and Head-Neck Surgery, King George's Medical University, Lucknow, Uttar Pradesh, India

Correspondence to
Dr Hitendra Prakash Singh,
drhpsingh77@gmail.com

Accepted 9 March 2014

SUMMARY

Nasopharyngeal angiofibroma is a rare and benign disease, which is mainly found in adolescent male subjects. It is usually diagnosed on clinical grounds on the basis of its presenting symptoms of nasal obstruction, nasal mass and most importantly unprovoked recurrent moderate to severe epistaxis. Imaging studies are only needed to confirm the diagnosis and formulate the management plan. A case of juvenile nasopharyngeal angiofibroma is presented here, which presented to us with severe respiratory distress and stridor. Urgent tracheostomy had to be performed before definitive management could be started. Definitive treatment was excision through modified transpalatal with sublabial route, which resulted in successful outcome.

BACKGROUND

Juvenile nasopharyngeal angiofibroma (JNA) is a relatively uncommon, benign but locally aggressive tumour that arises from the lateral wall of the nasopharynx. It was first described by Hippocrates in 5th century and the term 'angiofibroma' was coined by Friedberg in 1940.¹ Owing to its locally aggressive nature, it involves the surrounding regions by expansion. The signs and symptoms at presentation depend on its extensions. The most frequent symptoms are repeated epistaxis, nasal obstruction and anaemia in an adolescent male patient. Rarely swelling of the cheek and proptosis may be the associated symptoms. Stridor is usually not the presenting symptom because the tumour is diagnosed early on the basis of epistaxis. This case is presented here because of its unique presentation.

CASE PRESENTATION

A 14-year-old boy presented to the Outdoor of ENT & Head-Neck Surgery Department with symptoms of progressively increasing respiratory distress and noisy respiration for the past 2 months. There was a positive history of progressively increasing nasal obstruction and intermittent unprovoked nasal bleeding for the past 2 years. The patient also gave history of mild difficulty in deglutition and change of voice for the past 6 months. At the time of presentation, the patient had moderate stridor with suprasternal and intercostal retraction, which used to increase on exertion. Drooling of saliva was present. The patient was admitted and was subjected to complete examination. On general examination, there was pallor and cyanosis of lips. Facial appearance of the patient was like adenoid facies, that is, elongated face, open mouth, pinched nasal ala (figure 1). There was no

Figure 1 The patient showing adenoid like facies, drooling of saliva with in situ tracheostomy tube.

proptosis. On examination of the oral cavity and oropharynx, there was a smooth bulge of the soft palate with the presence of a reddish-white fleshy mass occupying whole of the oropharynx (figure 2). There was pooling of saliva in the oral cavity. Anterior rhinoscopy showed large reddish lobulated mass in the left nasal cavity along with secretions. This was confirmed by nasal endoscopy. Nasopharynx was totally occupied by mass. Owing to stridor, laryngeal examination could not be performed. After taking informed consent from patient's parents, urgent tracheostomy was performed using no. 7 portex cuffed tracheostomy tube under local anaesthesia. After the patient became stable, proper laryngeal examination was performed which revealed the mass hanging in the oropharynx, obstructing the supra-glottic lumen; however, laryngeal structures were normal in shape, size and function.

Figure 2 The patient showing bulge in the soft palate with pooling of saliva.

CrossMark

To cite: Singh HP, Kumar S, Vashishtha M, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-202745

INVESTIGATIONS

Laboratory investigations were performed which showed Hb 9.5 g%; TLC 4500/cmm; DLC: N 84, L 16, E 0, M 0, B 0; S. electrolytes: S.Na+ 134 mmol/L, S.K+ 4.2 mmol/L, S.Ca²⁺ 0.97 mmol/L; S.urea 22 mg/dL; S.creatinine 0.5 mg/dL; random blood sugar 106 mg/dL. His platelet count was 2.0 lakhs/mm³ and bleeding and clotting times were within normal limits. The patient was subjected for contrast-enhanced CT scan of the paranasal sinuses and neck, which revealed a heterogeneous soft-tissue attenuation lesion in the region of the left sphenopalatine fossa causing its marked widening with strong non-homogeneous postcontrast enhancement (figure 3). The lesion was also seen extending into posterior part of the left nasal cavity, oropharynx, left infratemporal fossa, abutting the medial pterygoid muscle with partial obliteration of the left parapharyngeal space (figure 4). The tumour was causing near complete obliteration of the oropharyngeal lumen and was extending till the epiglottis (figure 5).

DIFFERENTIAL DIAGNOSIS

- ▶ Juvenile nasopharyngeal angiofibroma.
- ▶ Antrochoanal polyp.
- ▶ Soft-tissue sarcoma.

Antrochoanal polyp and soft-tissue sarcoma could be excluded on the basis of history and CT scan findings.

TREATMENT

The patient was posted for excision of angiofibroma through modified transpalatal with sublabial approach using 'S'-shaped incision. The transpalatal approach was preferred because it gave direct access to the tumour in comparison to other approaches and by combining it with sublabial incision, lateral part of the tumour can be easily accessed. Preoperative embolisation was not performed. Under general anaesthesia, the patient was positioned on the operating table with head extended on the atlanto-axial joint. The incision was started on the right side of the hard palate, continued on the left side and then curved round the alveolar margin to come in the sublabial

Figure 3 Axial CT scan of the patient showing heterogenous soft tissue attenuation lesion in the region of left sphenopalatine fossa causing its marked widening with strong non-homogeneous postcontrast enhancement extending into the posterior part of the left nasal cavity, oropharynx, left infratemporal fossa, abutting the medial pterygoid muscle.

Figure 4 Coronal CT scan of the patient showing involvement of pterygoid plates by the tumour.

region, where incision was continued till the Canine fossa. Posterior part of the hard palate was removed and pterygo-maxillary fissure was widened using high-speed drill. By retracting the cheek using a tongue depressor, the lateral part of the tumour was visualised. The tumour was freed from its soft-tissue attachments by a blunt dissection method. The feeding vessel was encountered while the tumour was being removed from its bed. It was held with artery forceps and ligated. After removal, tumour bed was inspected to exclude any possibility of residual disease using nasal endoscopes. Blood loss was around 400 mL during the procedure. The patient received 2 units of blood transfusion during his hospital stay. figure 6 shows specimen of the tumour after removal.

OUTCOME AND FOLLOW-UP

Postoperative period remained uneventful and the patient was decannulated off tracheostomy on fourth postoperative day (figure 7). The patient was discharged on the seventh postoperative day. Histopathological examination of the tumour confirmed the preoperative diagnosis of JNA (figure 8). The patient was advised to come quarterly in the first year and

Figure 5 Coronal CT scan of the patient showing a near-complete obliteration of the oropharyngeal lumen caused by the lesion.

Figure 6 The excised tumour.

Figure 8 Histopathological appearance of the excised tumour. Note the typical feature of dense fibro-collagenous tissue with interspersed slit-like and gaping vascular channels (H&E, $\times 20$).

biannually in the second year for his follow-up visits. During every follow-up visit, complete history, physical examination and nasal endoscopy was performed. After 2 years of follow-up, there was no evidence of any recurrence.

DISCUSSION

Nasopharyngeal angiofibroma is the commonest benign tumour of the nasopharynx.² These tumours occur mostly in adolescent boys. These are composed of a rich vascular network within a fibrous stroma with a tendency of local destruction.

Though most angiofibromas originate in the posterior lateral wall of the nasopharynx, the exact site of origin is still a matter of debate. According to some researchers, it arises from the superior tip of the sphenopalatine foramen at the junction of the pterygoid process of the sphenoid bone and the sphenoid process of the palatine bone³ while others believe that it arises from the bone of the vidian canal.⁴

Many theories have been put forward for the development of JNA, which include relation to pituitary androgen-oestrogen axis⁵ role of various factors such as vascular endothelial growth factor receptor-2, transforming growth factor β -1, insulin-like growth factor-2⁶ and vascular hamartoma⁷ or inflammatory reaction theories.

JNA grows laterally towards the pterygomaxillary area, infra-temporal fossa, cheek; superiorly towards orbit⁸ through the

inferior orbital fissure; anteriorly in nasal cavity; inferiorly to the oropharynx. Intracranial extension occurs in 20–36%.⁹

The most common presentation of JNA is unilateral nasal obstruction or recurrent epistaxis in an adolescent man. Although nasopharyngeal mass is listed among the nasopharyngeal causes of airway obstruction causing stridor/stertor¹⁰ the authors could not find a report of nasopharyngeal angiofibroma presenting with stridor on performing search on Medline using keywords such as stridor, angiofibroma causing stridor, respiratory distress due to nasopharyngeal angiofibroma.

Contrast-enhanced CT scan and MRI is the most commonly used modalities for imaging of JNA. The diagnosis is usually based on the physical examination findings and imaging.

The management for angiofibroma is primarily surgical; however, radiotherapy and other modalities have been tried for unresectable tumours. Surgical excision of tumour should be complete as incomplete excision is the leading cause for recurrence, which occurs in 46% of cases.¹¹

Learning points

- ▶ Nasopharyngeal angiofibroma should be suspected in young men who present with huge nasopharyngeal mass and stridor even when there is no or minimal history of nasal bleeding.
- ▶ Contrast-enhanced CT scan is the most commonly used modality for imaging of juvenile nasopharyngeal angiofibroma (JNA).
- ▶ Complete surgical excision of tumour is vital to definitive treatment of JNA.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Gullane PJ, Davidson J, O'Dwyer T, et al. Juvenile angiofibroma: a review of the literature and a case series report. *Laryngoscope* 1992;102:928–33.

Figure 7 The patient on the fourth postoperative day.

- 2 Ulf-Rüdiger Heinrich, Jürgen Brieger, Jan Gosepath, *et al.* Frequent chromosomal gains in recurrent juvenile nasopharyngeal angiofibroma. *Cancer Genet Cytogenet* 2007;175:138–43.
- 3 Radkowski D, McGill T, Healy GB, *et al.* Angiofibroma. *Arch Otolaryngol Head Neck Surg* 1996;122:122–9.
- 4 Maroldi R, Nicolai P, *Imaging in treatment planning for sinonasal diseases.* New York, NY, USA: Springer, 2004.
- 5 Schiff M. Juvenile nasopharyngeal angiofibroma: a theory of pathogenesis. *Laryngoscope* 1959;69:981–1016.
- 6 Coutinho-Camillo CM, Brentani MM, Nagai MA. Genetic alterations in juvenile nasopharyngeal angiofibromas. *Head Neck* 2008;30:390–400.
- 7 Girgis IH, Fahmy SA. Nasopharyngeal fibroma: its histo-pathological nature. *J Laryngol Otol* 1973;87:1107–23.
- 8 Enepekides DJ. Recent advances in the treatment of juvenile angiofibroma. *Curr Opin Otolaryngol Head Neck Surg* 2004;12:495–9.
- 9 Close LG, Schaefer SD, Mickey BE, *et al.* Surgical management of nasopharyngeal angiofibroma involving the cavernous sinus. *Arch Otolaryngol Head Neck Surg* 1989;115:1091–5.
- 10 Albert D. *Scott-Brown's otorhinolaryngology, head and neck surgery, Part I.* 7th edn. Great Britain: Edward Arnold Ltd; 2008. Chapter 86, Stridor:1115.
- 11 Fagan JJ, Snyderman CH, Carrau RL, *et al.* Nasopharyngeal angiofibromas: selecting a surgical approach. *Head Neck* 1997;19:391–9.

Copyright 2014 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