CASE REPORT

Nasopharyngeal angiofibroma in a 32-year-old man

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SUMMARY

Nasopharyngeal angiofibroma (NA) is a benign, highly vascularised tumour of the nasopharynx, which typically occurs in young males aged 14–25 years. We report an interesting case of an NA arising de novo in a 32-year-old male. He was referred to our facility for severe nasal haemorrhage after biopsy of a left nasopharyngeal mass. In the operating room, extensive bleeding was noted, and an endoscopic medial maxillectomy was performed, and the left internal maxillary artery was ligated allowing for near total resection of the lesion. The pathological specimen confirmed the diagnosis of NA. To our knowledge, this is one of the oldest patients presenting with a de novo NA, in his fourth decade of life, confirming that this diagnosis must be considered in all those with large nasopharyngeal masses.

BACKGROUND

Nasopharyngeal angiofibroma (NA) is a benign, highly vascularised tumour of the nasopharynx, which typically occurs in young males aged 14–25 years. This tumour originates most often from the superior aspect of the sphenopalatine foramen in the posterolateral nasal cavity. Atypical sites have been described, including the lacrimal sac, inferior turbinate, tonsil and sphenoid, ethmoid and maxillary sinuses.^{1,2} Growing in the submucosal plane, it spreads medially into the nasal cavity and nasopharynx, laterally into the pterygopalatine fossa (PPF) and cranially into the inferior orbital fissure

Figure 2 H&E stain of patient's pathology specimen at 100x. Tissue stained strongly for beta-catenin and CD31 (vascular marker). Androgen receptor showed weak positivity. These findings support the diagnosis of nasopharyngeal angiofibroma.

and orbital apex. Several staging systems have been reported.¹

NAs derive from the fibrovascular stroma of the posterolateral nasal cavity, specifically centred at the junction of the sphenoidal process of the palatine bone and the horizontal ala of the vomer and pterygoid process. Characteristic CT findings include broadening of the PPF, sphenopalatine foramen and vidian canal (figure 1). The posterior maxillary sinus wall may be displaced anteriorly with large tumours. On MRI, the tumour shows flow voids and contrast enhancement on postcontrast T1 images.¹

The histopathology of NAs shows myofibroblast derivatives surrounded by a fibrous pseudocapsule (figure 2). The muscular layer may be present, but elastic fibres are often absent, preventing vessel contraction contributing to the profuse bleeding often seen clinically.³ Blood supply to NAs is typically from the ipsilateral internal maxillary artery, although contributions from the ascending pharyngeal artery and vidian artery are frequently seen. Contralateral vascular supply, although rare, has also been reported.¹

CASE PRESENTATION

We present a case of a 32-year-old male who underwent revision endoscopic sinus surgery (ESS) at an outside institution for sinusitis and polypoid masses in the nasal cavity and nasopharynx. His medical history is significant for a diagnosis of a right intraorbital rhabdomyosarcoma requiring resection and radiation as well as prior ESS for nasal polyposis. Prior sinus surgery was not complicated

Figure 1 Axial section of preoperative CT sinus (2016) just prior to revision ESS, aborted due to bleeding from posterior nasal cavity mass. Note broadening of right PPF (bracketed line) with mass filling nasopharynx (*). PPF, pterygopalatine fossa. ESS, endoscopic sinus surgery.

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by bleeding, and nasal polyps removed demonstrated no atypical features on histopathology. During his most recent surgery, extensive intraoperative bleeding from the left posterior nasal mass could not be controlled without full nasal packing, and the case was aborted. Urgent embolisations were performed on the first and second postoperative days but failed to allow for packing removal. He was then transferred to the University of Virginia Health System for further management. Review of his radiographic imaging prior to his initial sinus surgery revealed no nasopharyngeal or PPF masses. He was taken to the operating room where extensive bleeding was noted on manipulation of the mass, which appeared to be pedicled at the left sphenopalatine foramen.

TREATMENT

An endoscopic medial maxillectomy was performed, and the left internal maxillary artery was ligated allowing for near total resection of the lesion. Residual tumour was left in the area between the Eustachian tube and clivus due to the proximity of the paraclival carotid. He had an uncomplicated postoperative course and did not require any blood transfusions. Pathological evaluation of the mass revealed strong immunohistochemical staining for beta-catenin, diffuse nuclear positivity and weak androgen receptor positivity. These features were consistent with the diagnosis of NA despite preoperative concerns for a radiation-induced sarcomatous lesion. Gross tumour resection was performed, but margin status was not assessed resulting in the assumption of residual disease. Given the patient's prior history of radiotherapy, the decision was made by patient and physician to proceed with observation and reserve additional therapy should ongoing growth be noted.

OUTCOME AND FOLLOW-UP

The patient recovered well postoperatively. At his 6 month follow-up, he was found to have polypoid lesions that were biopsied and returned as granulation tissue polyps. The patient has repeat MRI scheduled 18 months postoperatively for ongoing surveillance.

DISCUSSION

We obtained approval from the University of Virginia Institutional Review Board to present this case report. The presentation of de novo NA in an adult male >30 years is exceedingly rare. There has been one case report in the literature of a 56-year-old male patient with NA confirmed by immunohistochemistry.⁴

The biological mechanisms involved in development of these lesions remains unclear. Given its restriction to young, often adolescent males, NA has been thought to be driven by the pituitary–androgen–oestrogen axis. A number of NAs express oestrogen and androgen receptors, although definite endocrinological anomalies have not been identified in affected individuals.⁵ In vivo study has revealed growth of NA in the setting of androgens. The largest report published, seven cases, demonstrated that using androgen antagonism preoperatively had a minimal reduction in tumour size (maximum tumour reduction of 11.1%).⁵

While the NA in this case expressed androgen receptors, its relevance is uncertain. Our patient was not on any androgen therapy and did not have a known endocrine abnormalities. His history of prior rhabdomyosarcoma does raise the concern for an underlying genetic predisposition for mesodermal derivative neoplasia.

Among the several staging systems that exist for juvenile nasopharyngeal angiofibroma, Snyderman *et al*⁶ developed an endoscopic staging system accounting for two key prognostic indicators: route of skull base extension and residual vascularity after embolisation. In this system, recurrent tumours were only seen in higher stages (III–V) and recurrence rates of 15%–50% have been reported in the literature. This staging system was also predictive of immediate morbidity—higher stages were associated with increased blood loss and need for staged operations.

This case further supports the notion that NA is not restricted to adolescent patients, as the usual moniker *juvenile nasopharyngeal angiofibroma* suggests. This tumour is an important consideration for the otolaryngologist in patients with a posterior nasal cavity mass, regardless of age. Additionally, significant morbidity to the patient may be avoided with proper suspicion and radiological evaluation and by approaching these lesions with appropriate preoperative planning and embolisation.

Learning points

- ▶ Nasopharyngeal angiofibroma (NA) is a benign, highly vascularised tumour of the nasopharynx.
- ▶ NA is not restricted to adolescent patients and is an important consideration for all patients with posterior nasal cavity masses regardless of age.
- ▶ When NA is suspected, patient morbidity can be avoided with appropriate preoperative planning and embolisation.

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