CASE REPORT

Endonasal endoscopic laser-assisted resection of septal glomangiopericytoma

Sana Sheikh,¹ Fatima Sarwar,¹ Nageeb Ullah Khan,¹ Muhammad Sarwar Khan¹

¹Department of Ear, Nose & Throat, KRL Hospital, Islamabad, Pakistan

²Department of Epidemiology, KRL Hospital, Islamabad, Pakistan

Correspondence to
Dr Nageeb Ullah Khan,
nageeb16@yahoo.com

Accepted 30 March 2018

SUMMARY

Glomangiopericytoma is a rare vascular neoplasm characterised by a pattern of prominent perivascular growth with myoid phenotype. It is categorised as a borderline low-malignancy tumour by WHO and accounts for less than 0.5% of all sinonasal tumours. After curative resection, patients of glomangiopericytoma need long-term endoscopic follow-up due to high risk of recurrence.

We report a case of a 23-year-old man complaining of nasal obstruction off and on and frequent epistaxis. A reddish mass in the right nasal cavity was observed on endoscopy and treated with endoscopic excision. Biopsy revealed this to be glomangiopericytoma arising from the septum of right nasal cavity, which was excised in toto with endonasal endoscopic approach using diode laser.

BACKGROUND

According to WHO 2005 classification of head and neck tumours, glomangiopericytoma is a sinonasal tumour demonstrating a perivascular myoid differentiation.¹ It is a rare neoplasm accounting for less than 0.5% of all sinonasal tumours and is usually of borderline low malignant potential.

CASE PRESENTATION

A 23-year-old man, presented in our tertiary care facility with a history of progressive right-sided nasal blockage and epistaxis over the last 1 year. Nasal endoscopy revealed a reddish pink polypoidal mass with smooth surface occupying the right nasal cavity. Thorough endoscopic examination did not demonstrate any other potential bleeding site. No cervical lymphadenopathy was noted.

INVESTIGATIONS

Haematological, microbiological and biochemical parameters were within normal limits.

CT scan showed a well-defined soft-tissue density area measuring 2.1×1.9×1.5 cm arising from nasal septum extending into right nasal cavity laterally abutting the right middle turbinate and extending inferiorly up to right inferior turbinate (figure 1). Frontal, sphenoid and bilateral anterior ethmoid sinuses were clear with minimal mucosal thickening in the maxillary sinuses. MRI demonstrated a homogenous enhancement in the lesion (figure 2). The CT scan and MRI confirmed the absence of nodal involvement.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis include glomus tumours, solitary fibrous tumours, angiofibromas, lobular capillary haemangiomas, myoepitheliomas, leiomyomas, olfactory neuroblastomas and rarely leiomyosarcomas and fibrosarcomas. Immunohistochemistry helps to differentiate between glomangiopericytoma and other differentials.²

TREATMENT

Complete endonasal endoscopic resection of the polypoidal mass was performed using Diode 980 nm Laser (figure 3). A margin of approximately 1.5 cm of septum adjacent to the mass was also removed, and both middle and inferior turbinates were reduced on the right side. The anterior table of sphenoid and part of rostrum was excised on posterior aspect of the tumour to achieve a wider clear margin, and superiorly the resection margin was extended up to the skull base. Laser-assisted resection ensured good haemostasis and avoided the need for perioperative embolisation which may often be required.³ Our patient did not need any kind of nasal packing at the end of operation.

A biopsy was performed and histological examination of the biopsy specimen showed respiratory mucosa with an underlying spindle-cell neoplasm with haemangiopericytoma-like vessels, perivascular hyalinisation and capillary-sized vessels. Cells had three syncytial cytoplasm and bland nuclei. Pleomorphism, necrosis and increased mitosis were not

Figure 1 CT scan shows soft-tissue mass in the right nasal cavity.

To cite: Sheikh S, Sarwar F, Khan NU, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-223752

Figure 2 Sagittal MRI of soft-tissue mass in the right nasal cavity.

present. Immunohistochemistry showed negative reactions with S-100 protein, desmin and myogenin.

OUTCOME AND FOLLOW-UP

Histological examination of the surgical specimen was consistent with glomangiopericytoma. Biopsies were also taken for tumour margin evaluation from anterior and posterior end of septum and were all negative, ensuring clear margins.

The postoperative course was uneventful, and the patient was discharged 3 days later. The patient remains disease free with no recurrence in his 18-month endoscopic follow-up.

DISCUSSION

Glomangiopericytoma represents less than 0.5% of all sinonasal neoplasms and has female predominance with peak age of incidence in the seventh decade.¹ Glomangiopericytoma is a rare mesenchymal tumour that falls within the category of borderline and low malignant potential soft-tissue tumours of the nose and

Figure 3 Resected mass from the right nasal cavity.

paranasal sinuses. Initially, this tumour was reported as haemangiopericytoma in 1942 which was later described by Compagno in 1976 as 'haemangiopericytoma like,' as it has a lower rate of metastasis and mortality.³ The WHO classified this tumour as glomangiopericytoma in 2005. It commonly presents as unilateral nasal obstruction with or without recurrent epistaxis. Other less common complaints include difficulty in breathing, visual disturbance and headache.⁴

Gross examination of the tumour usually reveals a polypoid mass, beefy red to greyish pink in colour, soft, fleshy or friable in consistency and oedematous or haemorrhagic in appearance, which bleeds to touch. Regional lymph nodes are rarely involved.⁵ CT scan demonstrates a non-calcified, soft-tissue mass in nasal fossa or a paranasal sinus and may be associated with sinusitis, displacement of adjacent structures and bone erosion. Contrast-enhanced imaging demonstrates the vascular nature of the tumour and is preferred to differentiate between tumour mass and inflammatory fluid in obstructed paranasal sinuses.⁶

The diagnosis of a glomangiopericytoma is based on histopathology. Microscopically glomangiopericytomas are submucosal polypoid tumours rimmed by a border of superficially spared stroma and have overlying intact respiratory epithelium with pushing and/or locally infiltrative borders. These tumours are composed of short, bland spindle cells forming fascicular, storiform or whorled patterns.⁷ Tumour nuclei are uniform and evenly spaced and tumour cells have a moderate amount of eosinophilic cytoplasm with indistinct cell membranes imparting a syncytial appearance. A characteristic feature is perivascular hyalinisation, and tumour cells may be seen oriented perpendicular to these hyalinised vessel walls. Glomangiopericytoma has limited mitotic activity, atypia and less amount of haemorrhage and necrosis as compared with haemangiopericytoma.

Immunohistochemically, glomangiopericytoma has characteristic diffuse reactivity for actin, vimentin and factor XIIIa with variable staining for desmin, S100 protein and CD34. There is negative staining for Bcl-2, CD99, CD117 and cytokeratins.

Success of the treatment depends on complete tumour resection; preferred presently is transnasal endoscopic laser-assisted resection which maintains the natural physiology of nose with many more advantages, like better view of the tumour and its extension, minimal blood loss, reduced risk of damaging lacrimal structures and decreased rate of recurrence and excellent prognosis. We recommend the use of Diode 980 nm Laser on account of its ability of good coagulation and rather 'bloodless' cutting. A meticulous surgical technique may obviate the need of perioperative embolisation.

The overall survival rate of glomangiopericytoma is high with a 5-year survival rate greater than 90% after surgical resection.⁸ Local recurrence is reported to be 7%–20%, mainly because of incomplete surgical resection; however, aggressive behaviour is rare which is expected in tumours with size of more than 5 cm, bone invasion, profound nuclear pleomorphism, increased mitotic activity (>4/10 high power fields), necrosis and a higher proliferative index (>10%).⁹ Metastatic disease and disease-related mortality is extremely rare (3%). Thus, a regular follow-up postoperatively is essential.

Contributors SS managed the whole of this case and selected it for publication; she wrote the abstract, case presentation, investigations, treatment and discussion sections of this article. FS assisted the operation and did extensive literature search for this case report. She also helped in writing of the investigations and treatment sections and provided references for the article. NUK critically reviewed the whole manuscript, edited and drafted it again and gave a final touch to the report and made it ready for publication. MSK is the main surgeon who operated this case. He also contributed in the drafting and revision of this document.

Learning points

- ▶ Glomangiopericytoma is a rare, indolent, unique sinonasal tumour usually of elderly age group with a characteristic perivascular myoid phenotype.
- ▶ It differs from conventional soft-tissue haemangiopericytoma in its location, biological behaviour and morphology.
- ▶ These tumours usually have a benign course and have very little chance of metastasis.
- ▶ Complete endonasal endoscopic laser excision is the treatment of choice since this is associated with little or no bleeding, rare need of preoperative embolisation and achieves a good clearance.
- ▶ Regular postoperative lifelong follow-up is recommended.
- ▶ In the absence of laser surgical facility or lack of expertise, however, perioperative embolisation should be considered due to the strong potential of these tumours to bleed.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

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) 2018. All rights reserved. No commercial use is permitted unless otherwise expressly granted.

REFERENCES

- 1 Anon. Rare sinonasal perivascular tumor. *J Med Sci* 2013;33:107–11.
- 2 Thompson LDR, Fanburg-Smith JC, Wenig BM. *et al* Borderline and low malignant potential tumours of soft tissues. In: Barnes L, Eveson JW, Reichart P, Sidransky D, . eds. *World Health Organization classification of tumours: pathology and genetics of head and neck tumours*. Lyon: IARC Press, 2005:43–5.
- 3 Roy NP, Desai D. Glomangiopericytoma of nasal cavity. *Indian J PatholMicrobiol* 2015;58:554–6.
- 4 Higashi K, Nakaya K, Watanabe M, *et al*. Glomangiopericytoma of the nasal cavity. *Auris Nasus Larynx* 2011;38:415–7.
- 5 Wang CC, Chu ST. Glomangiopericytoma of nasal cavity: a rare sinonasal perivascular tumor. *J Med Sci* 2013;3392:107–11.
- 6 Psoma E, Karkos PD, Dova S, *et al*. Sinonasal glomangiopericytoma treated with preoperative embolisation and endoscopic sinus surgery. *Ecancermedicalscience* 2016;10:692.
- 7 Oysu C, Verim A, Ertugay C. A rare tumor of nasal cavity: glomangiopericytoma case reports. *Otolaryngology* 2014. Article ID 282958.
- 8 Stout AP, Murray MR. Hemangiopericytoma: a vascular tumor featuring zimmermann's pericytes. *Ann Surg* 1942;116:26–33.
- 9 Anzai T, Saito T, Tsuyama S, *et al*. A case of glomangiopericytoma at the nasal septum. *Head Neck Pathol* 2017.

Copyright 2018 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