Glomangiopericytoma of the nasal cavity: A case report

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ABSTRACT
Introduction: Glomangiopericytoma is a rare perivascular tumor that accounts for less than 1% of all nasosinusal tumors. It belongs to the category of low malignancy tumor with a good prognosis.
Case report: We describe a case of 48-year-old female presented to our department with 2 months' history of progressive, permanent left nasal obstruction and nasal bleeding. Endoscopic examination of the nasal cavity revealed a left-sided, friable, vascular mass abutting to the superior turbinate. Computed tomography (CT) scan and Magnetic resonance imaging (MRI) showed a soft tissue mass occupying the left nasal cavity with intense contrast enhancement on MRI. The tumor was treated by endoscopic resection. Histological and immunohistochemical examination retained the diagnosis of glomangiopericytomas. During the 12-months follow-up no recurrence was observed
Conclusion: Glomangiopericytomas are rare, vascular tumors. It generally arises in the nasal cavity and may extend into the paranasal sinuses. Successful management depends on complete resection.
Key words: Glomangiopericytoma, Sinonasal cavity, Endoscopic surgery.

INTRODUCTION
Glomangiopericytoma (GPC) is a rare sinonasal mesenchymal neoplasm arising from the pericytes surrounding the capillaries. It accounts less than 0.5–1% of all sinonasal tumors [1]. It was first reported as a hemangiopericytoma in 1942 by Stout and Murray; however, in 2005 the World Health Organization (WHO) classified this disease as GPC. It was defined as a sinonasal neoplasm demonstrating a perivascular myoid phenotype [2]. It belongs to the category of borderline and low-malignant-potential soft tissue tumors of the sinonasal tract [2]. Our aim was to discuss diagnosis and therapeutic approaches of this rare tumour.

CASE REPORT
A 48-year-old female presented to the ENT Department of Farhat Hached Hospital with 2-month history of progressive, permanent left nasal obstruction and nasal bleeding. She had no anosmia no ophthalmologic symptoms and no headache. Endoscopic examination of the nasal cavity revealed a left-sided, friable, vascular, nasal mass abutting the superior turbinate. The floor and the nasal septum were not involved. There was no lymphadenopathy on the cervical palpation. A computed tomography (CT) scan showed a soft tissue mass of 3 cm x 1.5cm occupying the left nasal cavity with a heterogeneously enhancing. There was no bony destruction or invasion of the surrounding tissue (Figure 1).

Magnetic resonance imaging (MRI) showed a polypoid mass isointense on T1-weighted, hyperintense on T2-weighted with intense contrast enhancement on the left nasal cavity (Figure 2).
Pre-operative tumor embolization was not performed in light of the small size of the tumor. Considering the size, limited expansion and the location of the tumor, an endoscopic resection was performed. The bleeding was controlled without difficulty by compression with pledgets soaked in diluted adrenalin. Extemporaneous histology examination confirmed the absence of signs of malignancy.

Histological examination showed normal respiratory epithelium with a proliferation of short spindle-shaped cells with slightly branching vascular structures. Stromal bleeding was also noted; however, neither necrosis nor cytologic atypia were observed. The tumor cells were strongly positive for β-catenin and α-smooth muscle actin, and negative to CD34 and Pan-Cytokeratine (Figure 3). These findings were compatible with glomangiopericytoma. During the 12-months follow-up no recurrence was observed.

DISCUSSION

Glomangiopericytoma of the nasal cavity is a rare mesenchymal tumor. It develops in the nasal cavity and/or paranasal sinuses, where it represents less than 0.5% of all sinonasal neoplasm [1]. GPC belongs to the category of borderline and low-malignant-potential soft tissue tumors of the nose and paranasal sinuses. It can occur at any age; however, previous case series have suggested a peak incidence during the sixth and seventh decades in both genders, though, as in our case, it can occur earlier [1,3,4]. Some series shows a slight female predominance [1,4].

GPC are of unknown etiology. High vascularization caused by previous trauma, hypertension, and long-term steroid use are reported to be possible causes [1]. Clinically, as in our case, patients most often present with epistaxis and/or nasal obstruction. It may, rarely, provoke vision impairment, headache and local swelling because of local infiltration [1,3,5].

On examination, it usually appears as a quite vascular mass that can vary in color, size and consistency. According to some authors, this tumor is predominantly located in the nasal cavity, although some cases in the paranasal sinuses have been reported [6].

The radiological imaging techniques (CT and MRI) can provide adequate information with regards to extension of the tumor. The CT scan shows a soft-tissue mass with strong enhancement after contrast medium. It can demonstrate bone destruction in the sinonasal cavity and adjacent structures [1,7]. On MRI the mass appears solid isointense on T1-weighted images with strong contrast enhancement, while on T2-weighted images the signal is variable [7]. The angiography is used to plan a preoperative embolization [7,8].

According to the literature, the management of this tumor has been achieved through a variety of approaches. The open surgery was the only strategy to achieve free margins [9]. Actually, the endoscopic approach is the treatment of choice with comparable outcomes [1,5,10].
The preoperative embolization prior to the endoscopic ex-
cision is recommended in case of a large or highly vascularized tumor [8].
Other treatment modalities such as adjuvant radiotherapy
or chemotherapy are still controversial [4,11]. However, in
patients with a positive margin, adjuvant therapy could be
considered [1,4].
The histological examination is important for the diagnosis.
The diagnosis of GPC is often based on the architectural
features, especially the capillary pattern. The Hematoxy-
lin–eosin staining shows elongated or ovoid cells, surround-
ing the normal respiratory epithelium. These tumor cells
are characterized by round, punched-out central nuclei and
pale eosinophilic cytoplasm [4,7,12].
The immunohistochemical profile of glomangiopericyto-
ma is characterized by diffuse reactivity to vimentin, act-
in, β-catenin and negative staining for CD34, CD31 and
FVIII-Rag [4,10]. In our case, cells were strongly positive
for β-catenin and α-smooth muscle actin, and negative to
CD34. In addition to its histological and immunohistoche-
mmical features, mutations in exon 3 of the gene coding for
β-catenin (CTNNB1) and its nuclear expression were re-
cently discovered in GPC [12]. The prognostic factors for
aggressive behavior are a large tumor size (> 5 cm), bone
invasion, profound nuclear pleomorphism, increased mito-
tic activity, necrosis, and a higher proliferative index [2].
Life-long follow-up is generally required because of the risk
of local recurrence [1]. Complete surgical resection could
enhance disease-free survival, and adjuvant treatment
could be helpful to prolong survival when complete resec-
tion is impossible[1,4].

CONCLUSION
Glomangiopericytoma is a rare vascular tumor which diagno-
sis algorithm involves endoscopy, imaging (CT and MRI) for
lesion characterization and extension.
Successful management depends on complete resection.
Recent advances in nasal endoscopic surgery have en-
abled the complete resection of these tumors, minimizing
morbidity and facilitating subsequent surveillance of the
operative site.

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terest.

REFERENCES
1. Park ES, Kim J, Jun S-Y. Characteristics and prognosis of glomangio-
1909.
2. Barnes L, Eveson JW, Reichart P, Sidransky D, eds. Pathology and
Genetics of Head and Neck Tumours. World Health Organization Clas-
3. Arpaci RB, Kara T, Vayışoğlu Y, Ozgur A, Ozcan C. Sinonasal glo-
4. Thompson LDR, Miettinen M, Wenig BM. Sinonasal-type hemangio-
periyoicytoma: a clinicopathologic and immunophenotypic analysis of 104
cases showing perivascular myoid differentiation. Am J Surg Pathol.
5. Sheikh S, Sarwar F, Khan NU, Khan MS. Endonasal endoscopic
laser-assisted resection of septal glomangiopericytoma. BMJ Case
6. Terada T, Kato T. Sinonasal-type hemangiopericytoma of the nasal
7. Palacios E, Restrepo S, Mastrogiavanni L, Lorusso GD, Rojas R.
Sinonasal hemangiopericytomas: clinicopathologic and imaging find-
8. Ledderose G J, Gellrich D, Holtmannspötter M, Leunig A. Endo-
copic Resection of Sinonasal Hemangiopericytoma following Preope-
rative Embolisation: A Case Report and Literature Review. Case Rep
9. Morrison EJ, Wei BPC, Fancourt T, Lyons B. Glomangiopericytoma:
10. Anzai T, Saito T, Tsuyama S, Toh M, Ikeda K, Ito S. A Case of Glo-
mangiopericytoma at the Nasal Septum. Head Neck Pathol. 2017; 22
https://doi.org/10.1007/s12105-017-0870-6.
11. Gillman G, Pavlovich JB. Sinonasal hemangiopericytoma. Oto-
12. Suzuki Y, Ichihara S, Kawasaki T, Yanai H, Kitagawa S, Shimoya-
ma Y, et al. β-catenin (CTNNB1) mutation and LEF1 expression in si-
nonasal glomangiopericytoma (sinonasal-type hemangiopericytoma).

M. Bellakhdhar and al
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