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CASE REPORT

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Endoscopic Resection of a Rare Case of Fronto-ethmoidal Glomangiopericytoma: A Case Report

ABSTRACT

We report a case of a 58-year-old man with a history of nasal obstruction and recurrent epistaxis who underwent videonasolaryngoscopy after 9 months of symptoms. The first images showed a hyperemic mass affecting the left middle turbinate up to the nasal cleft. Computed tomography described a mass with an expansive effect occupying the entire left frontal sinus and most of the left ethmoid cells, widening the frontal sinus drainage path, and creating a continuity break of the cribriform plate, the left papyraceous lamina, and the upper-third of the nasal septum. Magnetic resonance imaging suggested T1-isointensity and T2-hyperintensity, intense contrast uptake, and no involvement of meningeal or brain tissues. The patient underwent extended endoscopic surgery without previous endovascular embolization or adjuvant therapies. A contralateral inferior turbinate graft was applied over the cribriform plate. Histopathological examination suggested glomangiopericytoma (GPC), and immunohistochemistry confirmed the diagnosis with positive beta-catenin, smooth muscle actin, and cyclin D1. The patient presented no nasal symptoms up to a 9-month follow-up. Nasal endoscopy showed no tumor recurrence signal. Although fronto-ethmoidal GPC is a rare tumor and presents challenging surgical access, it can be safely excised by endoscopic surgery. However, careful short- and long-term endoscopic follow-ups remain necessary to prevent postoperative complications and maintain surveillance of recurrences.

Keywords: Endoscopic sinus surgery, epistaxis, glomangiopericytoma, rare diseases, sinusitis

INTRODUCTION

Glomangiopericytoma (GPC) is a rare indolent nasal tumor with a prevalence of less than 0.5% among all sinonasal neoplasms, a 5-year survival rate of 90%, and a tendency for local delayed recurrence (rates reported as 17.8%-40%).[1-5] It is classified as a mesenchymal neoplasm with perivascular myoid phenotype cell differentiation,[6] including branching vessels and small vessels perivascular hyalization.[4,7] The World Health Organization (WHO) considers it a borderline low-malignant-potential tumor.[7,8] Patients generally present with nasal obstruction and epistaxis, but in more advanced cases, visual disturbance, chronic sinusitis, and/or headache are also present. Specific risk factors are still unclear, but the most related ones are the use of corticosteroids, high blood pressure, and trauma.[1] Glomangiopericytoma usually presents a macroscopic appearance of a reddish polypoidal mass, which may result in an uncertain initial diagnosis due to a variety of potential mimics.[3,6,7] Preoperative exams include nasal endoscopy, computed tomography (CT), and magnetic resonance imaging (MRI) to evaluate the size, extension, and characteristics of the mass for accurate presurgical planning.[9] The images show a soft tissue mass with erosive bony remodeling, typically hyperintense on T2-weighted images with vascular signal voids, a high mean Apparent Diffusion Coefficient (ADC) value, and a wash-in/wash-out pattern on dynamic contrast-enhanced MRI.[10]

A complete surgical excision of GPC by endoscopic sinus surgery remains the optimal treatment with excellent prognosis, minimizing morbidity, and facilitating surveillance.[11-13] Differential diagnoses include glomus tumor, hemangioma, hemangiopericytoma, leiomyoma, leiomyosarcoma, malignant melanoma, solitary fibrous tumor, and synovial sarcoma.[5] The histopathological findings are indispensable for differentiation, and an extended immunohistochemical panel that includes at least smooth muscle actin, beta-catenin, cyclin D1, STAT6, and CD34 (besides, complementarily, CD99, pan-cytokeratin

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cocktails, S100, vimentin, factor 13, and SOX10) helps to support the diagnosis without the need for extra molecular and mutational studies.[3,6,13,14]

Actin and CD34 could be used as independent prognostic indicators of GPC.[8] Tumors that present positive-actin and negative-CD34 have a better prognosis after a complete surgical excision.[3] Techniques such as preoperative embolization or lateral rhinotomy/external ethmoidectomy should be considered for large and extensive lesions.[5] Embolization with percutaneous direct puncture by direct intratumoral injection of permanent liquid polymer agents has evolved significantly over the past few decades.[15] However, there is still no consensus regarding adjuvant therapies or endovascular embolization.[11]

CASE PRESENTATION

We report a case of a 58-year-old man with a history of persistent unilateral facial pain and nasal obstruction, hyposmia, and recurrent but not intense episodes of ipsilateral epistaxis that started in January 2022, evolving after 8 months into 2 voluminous epistaxis episodes that needed anterior nasal packing. Past medical history of hypertension and no previous nose surgeries. After 9 months of symptoms, the patient underwent a videonasolaryngoscopy, endoscopic otoscopy, oral examination, a CT scan, and an MRI. Initial endoscopic images showed a nasal hyperemic expansive lesion affecting mainly the left middle turbinate up to the nasal cleft, covered with purulent secretions and crusts. Computed tomography-scan showed a soft tissue density mass with an expansive effect, occupying the entire left frontal sinus, widening its drainage path, and most of the left ethmoid cells and middle meatus (Figure 1), determining a break in the continuity of the ethmoidal intercellular septa, the cribriform plate, and the papyraceous lamina on this side (Figure 2), as well as the upper third of the nasal septum. Patient presented free spheno-ethmoidal recesses. Magnetic resonance image suggested lobulated tissue with an isointense signal on T1 and hyperintense on T2 (Figure 3), intense contrast uptake, and no involvement of meningeal and brain tissues or the left ocular globe (Figure 4).

MAIN POINTS

- Glomangiopericytoma is a rare sinonasal tumor that is a differential diagnosis for unilateral chronic rhinosinusitis and epistaxis. Although it is a low-malignant-potential tumor, it has a high incidence of delayed recurrence.
- Tumors affecting the sinonasal region and skull base are usually diagnosed late, as their symptoms are often banal and, therefore, easily overlooked.
- Surgical resection is the standard of care, with no consensus regarding endovascular embolization or adjuvant therapies.
- Complete surgical excision by endoscopic sinus surgery is the optimal treatment with excellent prognosis, minimizing morbidity, and facilitating postoperative surveillance.
- The use of an inferior turbinate graft prevents cerebrospinal fluid leak and entails faster healing and a lower risk of synechia.

An extended endoscopic surgery for tumor resection with a contralateral inferior turbinate graft over the cribriform plate was the proposed intervention, without previous endovascular embolization or adjuvant therapies. The patient was previously informed about a possible cerebrospinal fluid (CSF) leak and was orientated about restrict postsurgical care to prevent leaking persistence. The contralateral inferior turbinate graft was applied to prevent CSF leak, to obtain a faster healing, and to lower the risk of synechia. The graft from the contralateral side was chosen to lower the chances of presence of tumoral cells and avoid a later tumoral recurrence. The endoscopic surgery opened and drained the nasal left ethmoidal cells, left frontal sinus, and carefully removed the tumor over the cribriform plate, left papyraceous lamina, and upper third of the nasal septum, resulting in a sample of 40 × 20 × 3 mm. The cribriform plate resection was not planned as part of the first intervention. The histopathological findings were suggestive of sinonasal glomangiopericytoma. Complementary immunohistochemistry confirmed the diagnosis with positivity for beta-catenin, smooth muscle actin, and cyclin D1, and negativity for STAT6. Endoscopic postsurgical follow-ups were scheduled in the first month weekly, followed by every 6 months. The patient

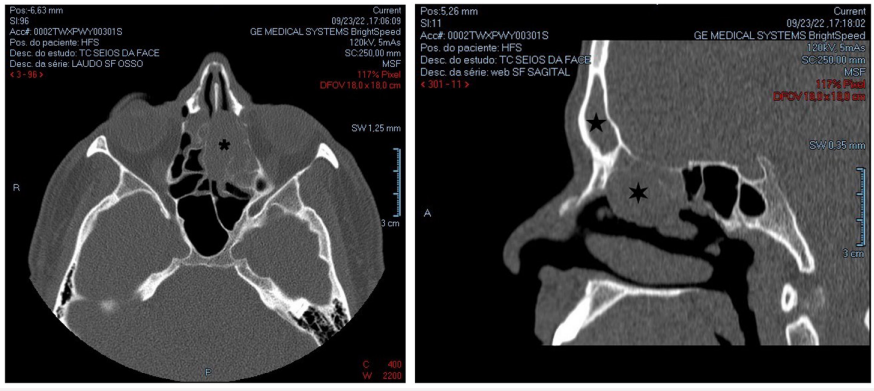

Figure 1. Contrast-enhanced computed tomography of nose and paranasal sinuses, axial and sagittal sections, before surgery. *Presence of tumor in the left fronto-ethmoidal region in axial section. ⁎Free spheno-ethmoidal recesses in sagittal section.

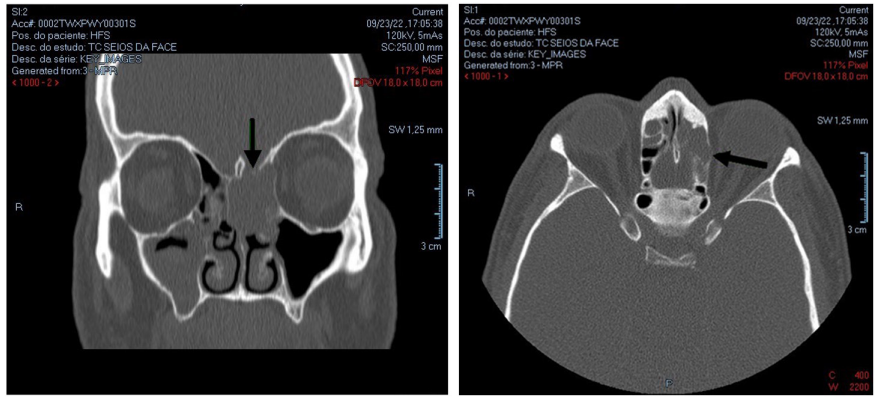

Figure 2. Contrast-enhanced computed tomography of nose and paranasal sinuses, before surgery. Focus on the continuity break (arrow) of the cribriform plate, in coronal section, and the papyraceous lamina (arrow), in axial section.

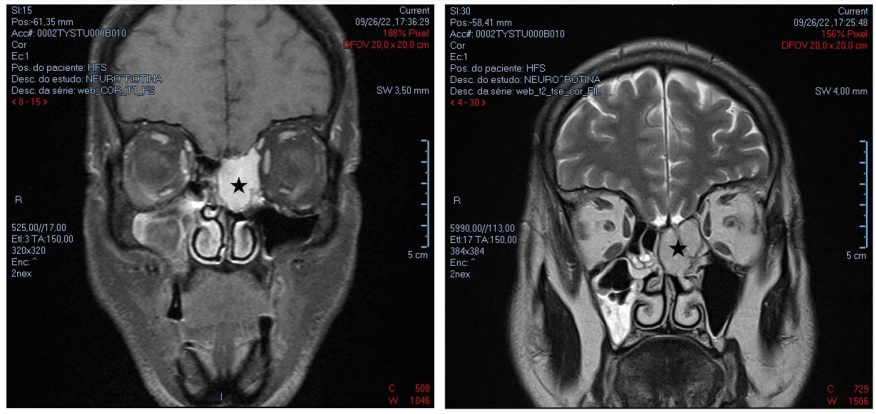

Figure 3. Gadolinium-enhanced magnetic resonance imaging of nose and paranasal sinuses, coronal sections, before surgery. ⁎Lobulated tissue with isointense signal on T1 and hyperintense signal on T2.

presented no nasal symptoms or signals such as facial pain, nasal secretion, nasal obstruction, nosebleed, anosmia, or CSF leak up to a 9-month follow-up. Nasal endoscopy presented healthy mucosa recovering the entire left nasal cavity, absence of left middle turbinate, and no signal of tumor recurrence. This case report obtained the patient's consent for publication.

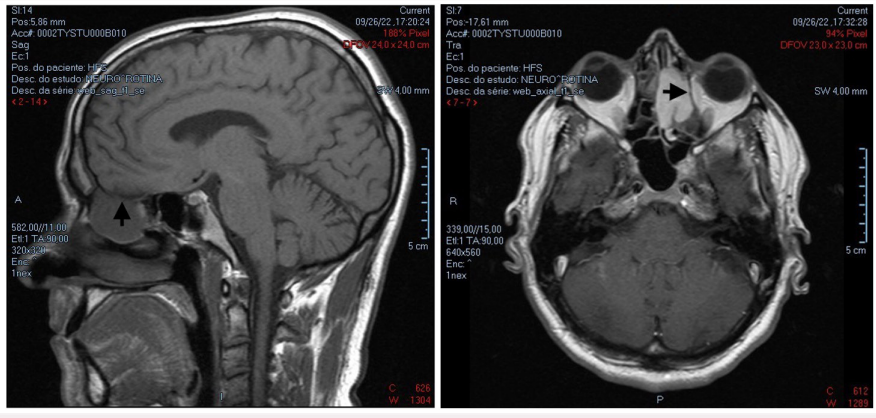

Figure 4. Gadolinium-enhanced magnetic resonance imaging of nose and paranasal sinuses, T1-weighted images, before surgery. Focus on the absence of involvement of meningeal brain tissues (arrow), in sagittal section, and absence of involvement of left ocular globe (arrow), in axial section.

DISCUSSION

Initially deemed as hemangiopericytoma, this tumor was recognized as a distinct diagnosis by the WHO in 2005.[2] Since then, specialists in the area have been more alert in considering this rare and borderline low-malignant-potential tumor as a differential diagnosis.[7] Our patient's case is similar to most other cases reported in the literature, showing the most typical initial symptom and signal of this tumor, which is nasal obstruction and recurrent epistaxis. In addition, the patient has high blood pressure, one of the known specific risk factors.[1] Also, his diagnosis was delayed because of common potential mimics, as the mass morphological aspect is similar to a nasal polyp.[3,6] He underwent the protocol of physical and image exams as preconized,[9,10] receiving as the first result a suggestive benign tumor. After an extended literature revision, the surgery team decided in agreement with the patient for an extended endoscopic sinus surgery, aiming at a complete excision of the tumor, followed by in-surgery control of damages.[11,12] No adjuvant therapies, external access surgery, laser surgery, or endovascular embolization were considered as first-line options.[5,11,15] Histopathological analysis and immunohistochemistry were applied following the proposed extended panel of markers specific to the most suggestive tumor.[3,13] The differential diagnoses were excluded with no need for further molecular and mutational analysis.

The patient is in the first-year post-surgery follow-up. Even though this case does not yet present a long-term postoperative result, such as cases already reported,[11,12] we aim to contribute to the literature with a case that is challenging due to this tumor's location and close connection to noble structures. Although fronto-ethmoidal GPC is a rare tumor and presents challenging surgical access, it can be safely excised by endoscopic surgery. Nevertheless, careful short- and long-term endoscopic follow-ups remain necessary to prevent postoperative complications and to maintain surveillance of recurrences.

Informed Consent: Informed consent was obtained from the patient who agreed to take part in the study.

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Declaration of Interests: The authors have no conflict of interest to declare.

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