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Case Report

Sinonasal Glomangiopericytoma: Bibliographic Review and Case Report

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Abstract

Sinonasal Glomangiopericytoma is an extremely rare tumour that, although benign, should be included in the differential diagnosis of masses in the nasal cavity.

An Argentinian male patient of fifty (50) years of age was admitted to Centro de Nariz, Garganta y Oído Concordia. His main symptom was nasal obstruction of four months of evolution predominantly on the right side. He reported no surgical history.

A nasal endoscopy showed an angiomatous tumour mass in the right nasal passage. A highresolution computed tomography was requested, showing a soft tissue density image in the ipsilateral cribriform region. After the results of the CT scan, the lesion was resected and biopsied. Its pathological and its associated immunohistochemical analysis gave the diagnosis of Glomangiopericytoma.

Keywords: Nasal Congestion; Hemangiopericytoma; Sinonasal Glomangiopericytoma; Endoscopic Resection

Introduction

Sinonasal Glomangiopericytoma (GPC) is an extremely rare benign neoplasm, previously known as hemangiopericytoma (HPC). The latter name dates from 1942 and was introduced by Stout and Murray to describe a tumour formed by cells arranged around blood vessels with a staghorn-like structure [1].

The sinonasal-type HPC, a term coined by Compagno and Hyams in 1976, constitutes a welldefined clinicopathological entity and is characterized by being a spindle-cell tumour with morphological features similar to those of the soft tissue hemangiopericytoma [2].

It was observed that when this lesion originated in the nasal cavity, it tended to behave in a more indolent manner than its soft tissue counterpart, suggesting that the sinonasal hemangiopericytoma represented a different entity [6].

Currently, its diagnosis is based on endonasal endoscopic examination, CT and MRI images, biopsy of the intraoperative specimen, pathological analysis and immunohistochemical differentiation. The gold standard for its treatment is complete endoscopic resection with free margins, since a 16.8% local recurrence rate has been reported due to incomplete excision. It is further classified as a borderline tumour of low-grade malignancy. Therefore, it is important to include it in the group of nasal cavity masses.

We present a rare case of Glomangiopericytoma of the right nasal passage in a 50-year-old Argentinian male patient who was treated with surgical resection. Currently there is no local recurrence.

Case Report

A 50-year-old male patient was admitted to Centro de Nariz, Garganta y Oído Concordia on March 2021, with a predominantly right nasal obstruction of four months of evolution, associated arterial hypertension and no surgical history.

On physical examination, nasal endoscopy revealed a reddish heterogeneous mass in the right nasal passage (Figures 1 A, B).

Radiological investigations: high-resolution tomography in which an image compatible with a soft-tissue-density lesion with right ethmoidal involvement was observed, without osteolysis of the orbital or cribrotic regions (Figure 2). Magnetic resonance imaging was not ordered due to the location of the neoplasm. Preoperative biopsy was not performed because of the high possibility of bleeding complications.

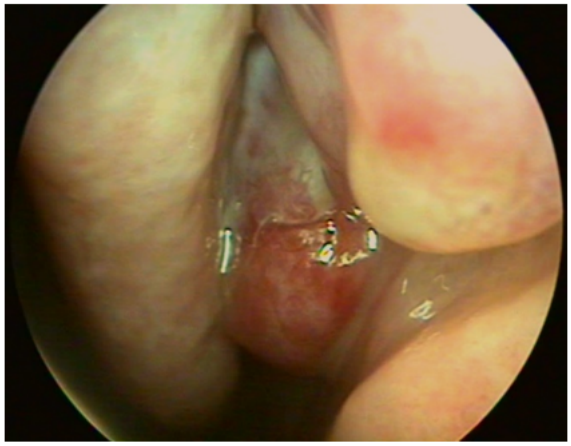

Figure 1A: Reddish tumour lesion in the right nasal passage; the septum can be seen medially and the middle turbinate bone can be seen laterally.

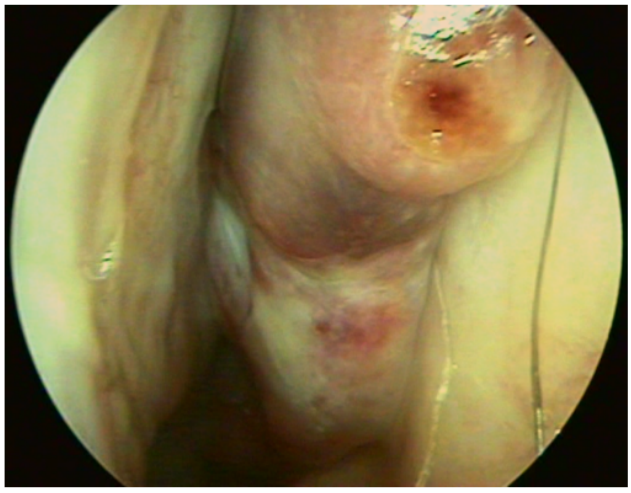

Figure 1B: The anterior portion stands out, being similar in color to the underlying mucosa

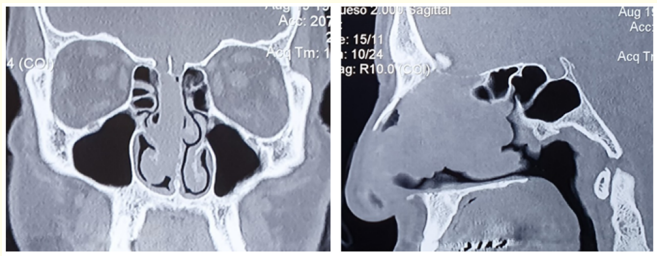

Figure 2: High resolution computed tomography showing a polypoid lesion occupying the right nasal passage, without bone compromise.

The excision was performed by a senior surgeon of the staff. Under general anaesthesia and orotracheal intubation, assisted by a zero-degree rigid endoscope, after topical application of lintines soaked in oxymetazoline and infiltration of 4% carticain hydrochloride plus adrenaline 1:100,000, a complete tumour excision was performed. The surgical specimen was subsequently sent for pathological study.

The specimen histological analysis indicated a hemangiopericytoma, whose immunohistochemistry confirmed to be a GPC. The negative result for SMA stood out. (Figures 3 A-F).

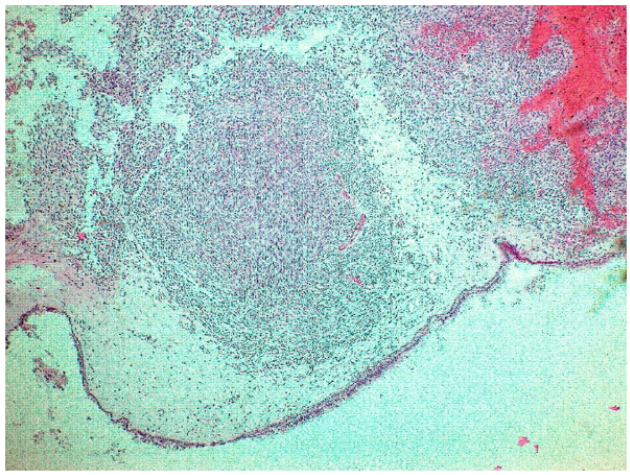

Figure 3A: Sinonasal Glomangiopericytoma. Proliferation of cells localised in submucosa lined by respiratory epithelium. H and E.

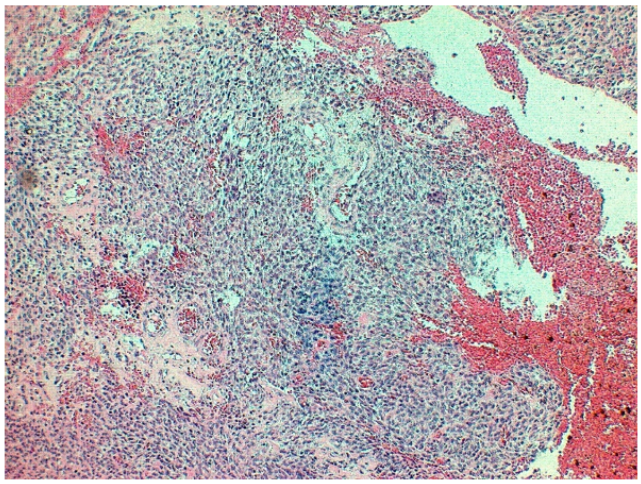

Figure 3B: Sinonasal Glomangiopericytoma. Proliferation of spindled and round cells with prominent blood vessels. Focal bleeding. H and E.

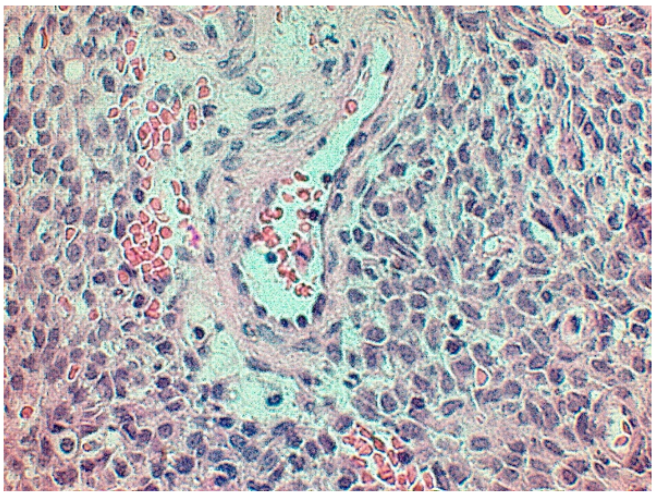

Figure 3C: Sinonasal Glomangiopericytoma. Pattern of prominent perivascular growth with perivascular hyalinization. H and E.

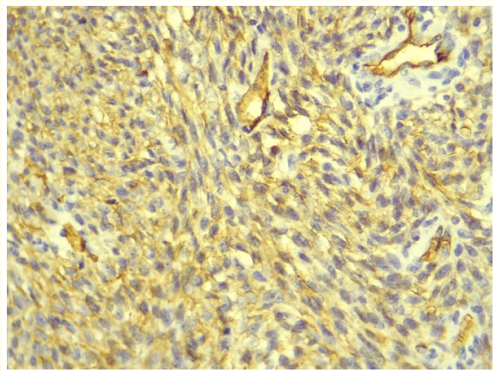

Figure 3D: Sinonasal Glomangiopericytoma. CD34. Diffuse positivity.

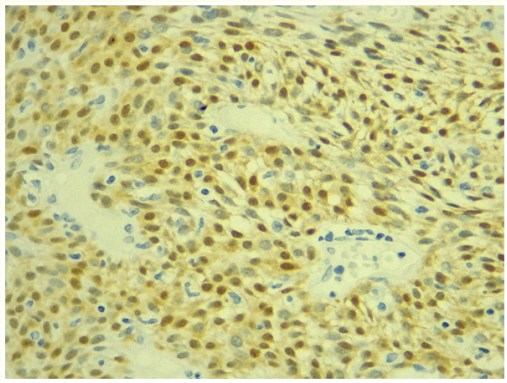

Figure 3E: Sinonasal Glomangiopericytoma. β catenin. Diffuse positivity.

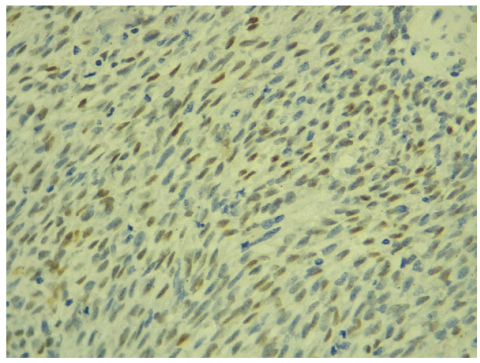

Figure 3F: Sinonasal Glomangiopericytoma. Cyclin D1. Diffuse positivity.

At the moment, 8 months after the procedure, no local recurrence is observed.

Discussion

GPC is a soft tissue tumour with perivascular myoid differentiation, which accounts for 0.5 to 1% of all sinonasal tumours. It is a rare mesenchymal neoplasm with an average age of occurrence at around 70 years of age and an average age range from 5 to 90 years old. There is no evidence of recognized family tendency or sexual predilection [1,3,4].

Regarding its aetiology, it is unknown, but factors such as trauma, arterial hypertension, pregnancy and the use of corticosteroids are described in the literature [3]. In our report, arterial hypertension stands out.

According to a study by Kono., *et al*. 23 patients with GPC were identified from 2005 to date, in which the median age was 60, with a more marked tendency of occurrence in women (16:7). The most frequent clinical presentation was epistaxis (78%), followed by nasal obstruction (52%) and headache (17%) [5]. Our patient consulted for nasal obstruction.

Regarding its location, it generally involves the nasal cavity in isolation, but it can be located at the maxillary and ethmoidal level. It occurs unilaterally, while bilaterality is rare [6]. The reported case showed cribriform region compromise.

In a review by Asimakopoulos., *et al*. The study of this type of lesion was performed under nasal endoscopy, which has the advantage of allowing the specialist to do the biopsy at their office. In our case, we did not carry it out due to the vascular nature of the lesion and the risk of bleeding [7].

As for imaging, computed tomography was the selected diagnostic technique. Suh., *et al*. Reported that GPCs stand out as homogeneous soft tissue masses, showing erosive bone remodelling. They also present an avid and homogeneous enhancement in both CT and MRI, intermediate signal intensity in T2WI, high ADC values and rapid washout patterns in DCE-MR images. They are also accompanied by vascular signal gaps in T2WI. Together, these elements could suggest a GPC with malignant potential [8]. Our patient's CT evidenced a polypoid mass without bone erosion.

Characteristic histology shows epithelioid cells in a perivascular pattern with frequent hyalinization. These cells are histologically positive for cytoplasmic SMA, vimentin, and nuclear β-catenin in 80 to 100% of cases; they do not show strong diffuse staining for CD34 and are basically negative for AE1 / AE3, Bcl-2, CD34, CD99, CD117, factor VIIIR Ag, protein S-100 and STAT6 [5,7,14]. Our case matches the description found in the literature; however, it was negative for SMA.

At the same time, a differentiating diagnosis should be made against conventional HPC and all those lesions that present a similar pattern. Solitary fibrous tumours present variable cellularity with hypo and hypercellular areas together with a collagen deposition similar to a keloid. The former stain diffusely and strongly for CD34, while the GPC stains more for SMA. Glomus tumours lack nuclear β-catenin expression and CTNNB1 mutations, whereas fibroblastic neoplasms such as nasopharyngeal angiofibroma and desmoid-type fibromatosis exhibit nuclear β-catenin expression and CTNNB1 mutations. For this same reason, Kono., *et al*. Propose the genetic analysis of oncogenes to distinguish vascular neoplasms that originate in the head and neck region [5,7,9,10].

Regarding the current management of this type of lesion, the gold standard is resection with free margins, without emphasising any particular approach. The possibility of carrying out prior embolization is considered in order to achieve a more adequate control [7,11,13]. The approach we selected was a videoendoscopic endonasal one.

Two main complications may occur: bleeding and cerebrospinal fluid fistula [7]. Our patient did not develop any intraoperative nor postoperative complications.

Finally, Thompson., *et al*. Reported a disease-free survival rate of 74.2% at 5 years and 64.4% at 10 years. The recurrence rate was 17%, varying from a few weeks to 12 years from the initial occurrence, and attributing it to an incomplete excision when happening within one year of the surgical procedure [12]. We have not observed local recurrence in our patient after 8 months.

Conclusion

This case shows a sinonasal Glomangiopericytoma. Its computed tomography evidenced a polypoid mass in the right nasal passage. An endoscopic resection was performed and its subsequent

pathological study confirmed a GPC on immunohistochemistry. There has not been local recurrence after 8 months of the intervention.

In this report, the diagnostic suspicion suggested a vascular lesion and hence the importance of including it within the differential diagnoses of masses in the nasal passage.

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