

CASE REPORT

Sinonasal hemangiopericytoma – case report and literature review

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ABSTRACT

Sinonasal hemangiopericytoma is a rare vascular tumor with malignant behaviour in what it concerns the local invasion characteristics and recurrence rate.

Three cases of nasal hemangiopericytoma are reported, emphasizing the diagnostic work-up and the therapeutic management for each one.

In the literature review section, the authors present the epidemiological and morpho-pathological characteristics of this tumoral pathology, pointing-out the complexity of the clinical, radiological and histopathological diagnostic algorithm needed in this situation. The particularities of the therapeutic protocol are presented, showing the advantages and disadvantages of each surgical approach.

KEYWORDS: sinonasal hemangiopericytoma, vascular tumor, endoscopic approach

INTRODUCTION

As Stout and Murray described it in 1942, hemangiopericytoma is a rare vascular tumor derived from Zimmermann's pericytes¹ - modified smooth muscle cells with mesenchymal origin, with pronounced morphologic heterogeneity².

Because the pericytes presumably are modified contractile smooth-muscle cells found in the capillaries and post-capillary venules structure^{3,4}, hemangiopericytomas may appear in any capillarized tissue. Nevertheless, the literature describes them primarily in the lower extremities and the retroperitoneum, where they are known to have malignant behaviour^{3,5}.

In the head and neck region arise 25% of the hemangiopericytomas, representing a small part of head and neck tumours – about 5%⁶. The most common locations in this area are the nasal cavity and paranasal sinuses, especially the ethmoid and sphenoid sinuses, where they have a lower risk of malignancy and metastasis compared with any other sites^{7,8,9}.

As the sinonasal hemangiopericytoma was studied, it was concluded that it represents a distinct entity, on one

hand because of its lower malignancy grade, on the other hand because the sinonasal lesions have distinctive morphologic features and show true pericytic differentiation. Compagno and Hyams named these lesions hemangiopericytoma-like intranasal tumors in 1976¹⁰⁻¹².

The etiology is still unknown, even if it was thought that hormonal disturbances, prolonged steroid use or trauma could be involved. There is no sex predilection and it seems to appear at any age, but more frequently in the 6th and 7th decade of life^{13,14}.

The main symptomatology consists in frequent episodes of epistaxis and nasal obstruction⁷.

Effective management requires wide surgical excision with clear resection margins, as these tumors are relatively radioresistant. Recent advances in endoscopic surgery have led to the development of techniques that now permit endoscopic resection.

This article discusses three clinical cases of hemangiopericytoma treated in “Sfanta Maria” ENT Department between 2012 – 2014. The particularities of diagnosis and treatment are presented together with a short literature review regarding sinonasal hemangiopericytoma.

CASE REPORT

Case 1: V.M., 37-year-old female, was admitted in our Department in January 2013 for right chronic nasal obstruction and right chronic antero-posterior rhinorrhea. Symptomatology enhanced in the previous 3 months, being accompanied by frequent episodes of anterior epistaxis and right nasal pyramid tumefaction.

The patient was diagnosed in 1995 with hemangiopericytoma of the right nasal fossa which has been repeatedly surgically excised in 1996, 1997 and august 2012.

The clinical examination showed a vegetant tumor situated in the superior half of the right nasal fossa with left septal displacing. In its medial third, the nasal septum was perforated. Due to repeated surgical interventions, the patient had an iatrogenic lack of the right nasal turbinates and of the posterior part of the lateral nasal wall.

The native CT sinuses examination showed an extensive tumoral process, 33/25 mm in diameter, occupying the right anterior nasal cavity and causing mass effect on the nasal septum. It infiltrated the soft parts and extended to the subcutaneous grease, right maxillary and caused erosion of the nasal bones (Figure 1).

Our decision was to remove the tumor using the endoscopic endonasal approach under general anaesthesia. Intraoperatively, we found a red, vegetant tumor, with soft consistency, which easily bled at instrumental touch. We performed total tumoral excision using the "peace-meal" resection method (Figure 2, Figure 3).

The histopathological examination reconfirmed hemangiopericytoma.

Case 2: Female patient, 80-year-old, known with nasal polyposis, presented for bilateral nasal obstruction, anosmia and anterior rhinorrhea that occurred 1 year previously. The clinic examination showed an edematous tumor, which filled the entire left nasal fossa, the right inferior and middle meatus and also the posterior nasal aperture.

Native and contrast enhanced CT scan described proliferative process of the nasopharynx occupying



Figure 1 Cranio-facial CT examination (coronal slice) aspect showing an extensive process, occupying the right anterior nasal cavity and causing mass effect on the nasal septum. It infiltrates the soft parts and extends to the subcutaneous grease, right maxillary and causes erosion of the nasal bones.

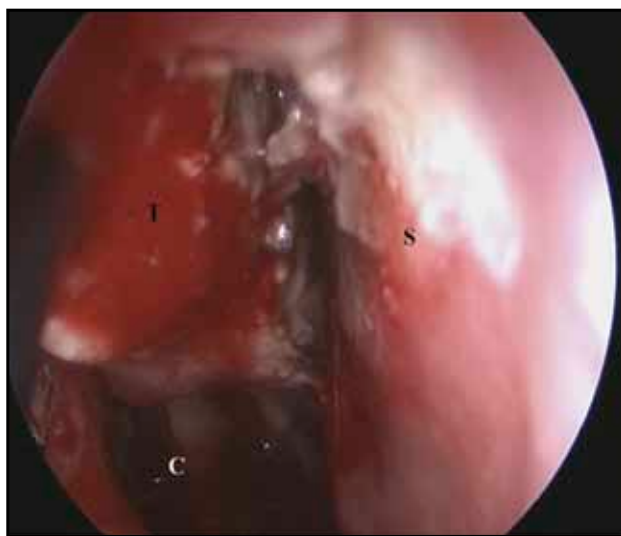


Figure 2 Intraoperative aspect; S= nasal septum; C= right choana; T= red, vegetant tumor, with soft consistency. The lack of anatomical structures of the lateral nasal wall because of the previous surgical procedures can be seen

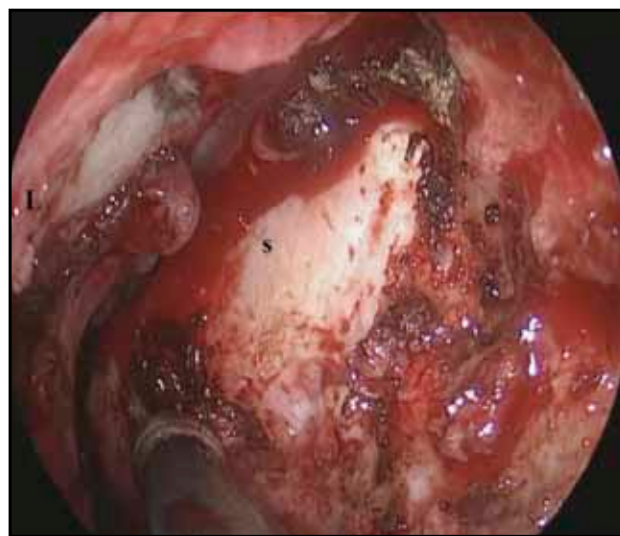


Figure 3 Post-tumoral resection right nasal fossa cavity; S = nasal septum (perpendicular plate of the ethmoid); L= the anterior part of the lateral wall of the nasal fossa

the entire left nasal fossa, with ethmoid and sphenoid sinuses extension (Figure 4).

Intraoperatively, a voluminous, smooth tumor has been discovered, extended to the left nasal fossa, the rhinopharynx and the posterior third of the right nasal fossa, which infiltrated 1/3 of the posterior of the nasal septum, the left ethmoid cells and the left sphenoid sinus.

A tumoral fragment was sent to extemporaneous anatomopathological examination, revealing a malignant tumor – possibly low-grade sarcoma or lymphoma. Considering this possible diagnosis and the age of the patient, the intraoperative decision was to complete endoscopically the tumoral debulking in a piece-meal fashion.

The final histopathological result, confirmed by immunohistochemistry tests, was hemangiopericytoma. No recurrence was noticed 6 months postoperatively.

Case 3: A 29-year-old female patient was admitted to our institution complaining of right nasal obstruction appeared 6 weeks previously. She also reported recurrent episodes of anteroposterior epistaxis during this period.

Head and neck examination revealed a red, pulsatile, vegetant tumor localized in the posterior part of the right nasal fossa, with partial obstruction of the right choana, without clinical cervical adenopathy.

A CT scan of the paranasal sinuses revealed a compact mass located in the right pterygopalatine fossa,

extended to the posterior 3rd of the right nasal cavity and to the nasopharynx, measuring 25/16 mm in diameter. It had a mass effect on the posterior maxillary sinus wall and inferior sphenoidal wall, being in contact with the right superior and middle turbinates and also with the nasal septum (Figure 5).

The MRI showed an expansive tissular mass situated in the right nasal cavity from the inferior and middle turbinates, well defined, with intense gadolinophilly, 20/20/27 mm, apparently with external carotid artery vascularization (Figure 6).

The biopsy taken in another ENT clinic showed hemangiopericytoma.

We decided to perform tumoral resection using the endonasal approach after tumor embolization, because of the high bleeding risk. Intraoperatively, two tumors apparently with sphenopalatine foramen origin – one soft, friable tumor, which easily bled at instrumental touch, and another tumor of high consistency - were discovered in the posterior third of the right nasal cavity (Figure 7, Figure 8). Both tumors were completely excised using the endoscopic endonasal approach (Figure 9).

The treatment of this kind of tumors does not end after the total resection, the subsequent treatment being very important together with the long-term follow-up. The postoperative treatment consists in emollient ointments and lavage with saline solutions. There was registered no relapsing tumor during the follow-up period in the presented cases.

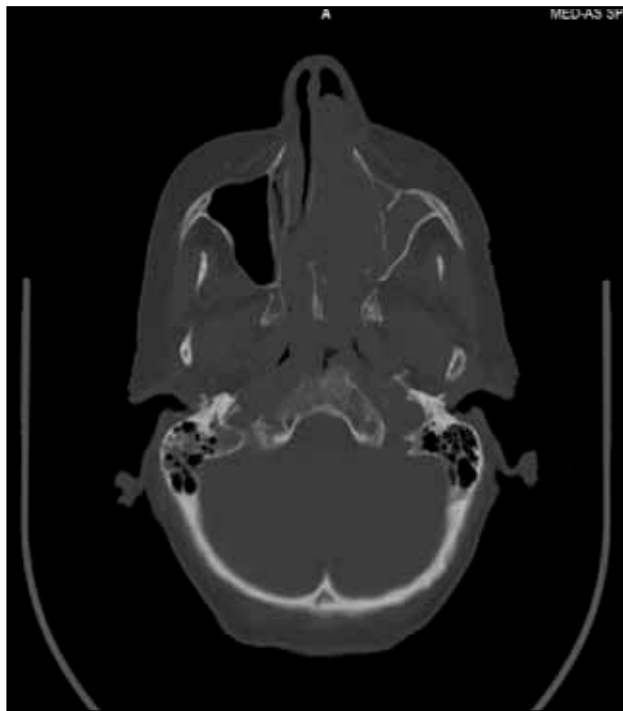


Figure 4 Crano-facial CT examination (axial slice) showing the proliferative process of the nasopharynx occupying the entire left nasal fossa, with ethmoid and sphenoid sinuses extension



Figure 5 Axial CT scan revealing a compact mass located in the right pterygopalatine fossa, extended to the posterior 3rd of the right nasal cavity and to the nasopharynx

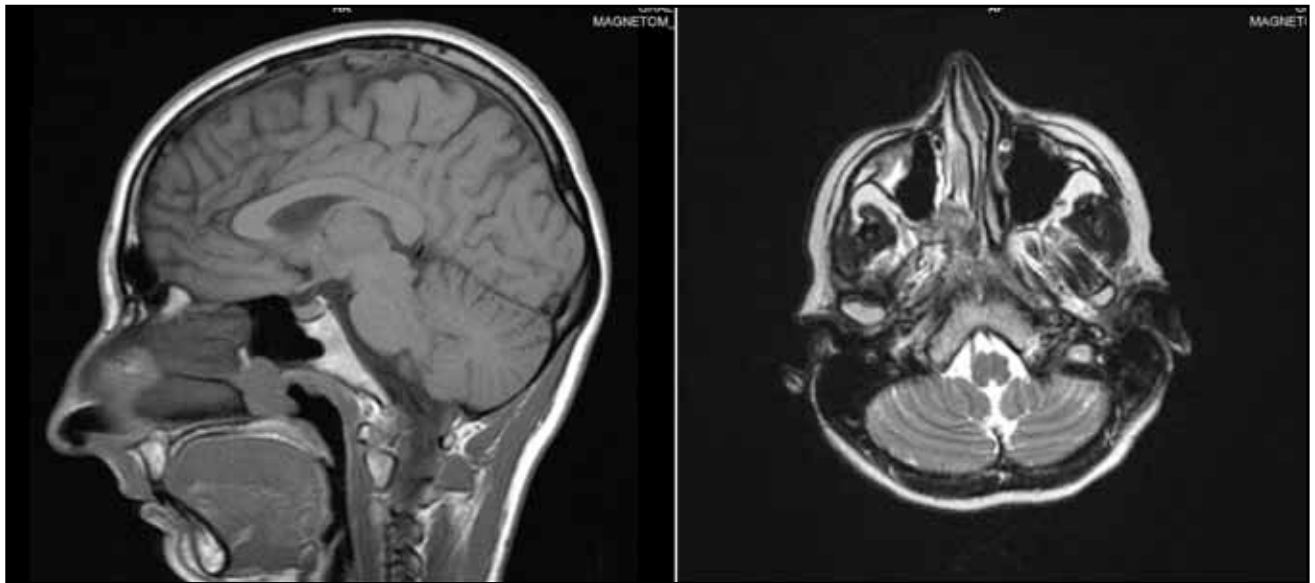


Figure 6 Cranio-facial MRI scan (sagittal and axial slices) showing expansive tissular mass situated in the right nasal cavity from the inferior and middle turbinates, well defined, with intense gadolinophilly

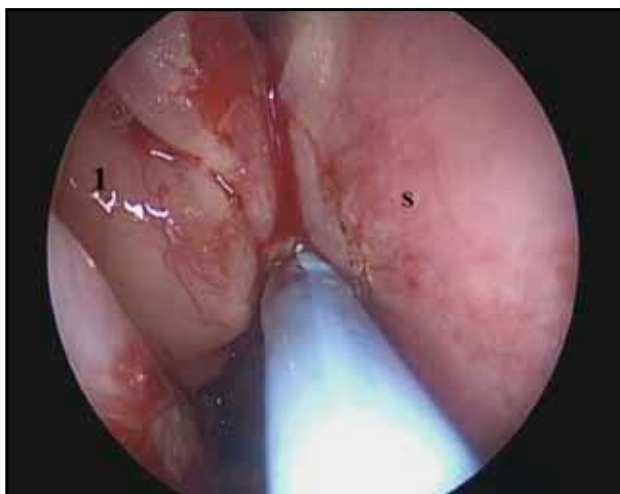


Figure 7 Intraoperative aspect: 1= High consistency tumor; S= Nasal septum

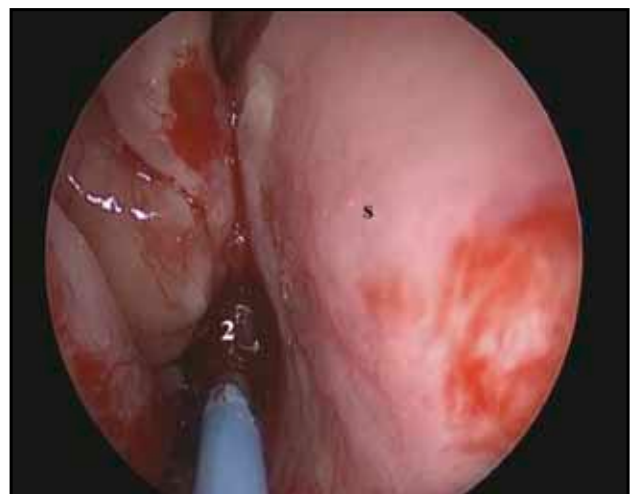


Figure 8 Intraoperative aspect: 2= Soft, friable tumor; S= Nasal septum



Figure 9 Intraoperative aspect – tumoral excision from the pterygopalatine fossa (T – tumor, Pf - pterygopalatine fossa)

DISCUSSIONS AND LITERATURE REVIEW

In 2001, more than 60 cases of nasal hemangiopericytoma have been described in the literature¹⁵, while in 2013 the number got to almost 250¹⁶. There is no absolute term used for this type of tumour, some authors prefer “intranasal hemangiopericytoma - like tumors”¹⁰; others consider that there is a strong bound with soft tissue hemangiopericytoma, so they use the term “sinonasal hemangiopericytoma”¹. In 2005, The World Health Organization proposed the denomination “glomangiopericytoma” due to the similarity with glomus tumours^{17,18}. Nowadays, in practice, the general term “hemangiopericytoma” has become accepted for all tumours with hemangiopericytoma-like histology, after exclusion of other tumor entities¹⁹.

Regarding gender and age of occurrence, sinonasal hemangiopericytoma mainly affects middle-aged patients, in the 6th-7th decade of life, with no gender predilection^{13,20,21}.

Over time, there have been many hypotheses concerning the histopathological structure of these tumors, depending on location. Compared with the soft tissue types, sinonasal hemangiopericytomas are usually composed of more uniform, smaller cells, having a very good prognosis. They hardly present nuclear polymorphisms with no hemorrhage, no necrosis, nor any other characteristics of the somatic sites of hemangiopericytoma. Compagno et al.¹⁰ used the term of hemangiopericytoma-like tumor in 1976, while Eichorn et al.²² affirmed, based on ultrastructural and immunohistochemical findings, that nasal hemangiopericytoma does not differ from similar tumors in other sites. Nevertheless, they observed focal smooth muscle actin immunoreactivity in nasal tumors, suggesting myoid differentiation, particularity rarely found in soft tissue hemangiopericytomas.

Chowdhary et al.²³ describes the sinonasal hemangiopericytoma as an uncommon upper aerodigestive tract tumour, of uncertain cellular differentiation, with the majority of them behaving in a benign manner, with excellent long-term prognosis (88% for 5 years survival) after surgery alone. In microscopy, their appearance is light, with an immunophenotypic profile resembling glomus tumours.

Sinonasal hemangiopericytoma has been subtyped as²⁴:

(a) *Soft tissue-type hemangiopericytoma*: an aggressive tumour, locally destructive, that may produce metastasis to regional lymph nodes and lungs. It does not show myoid differentiation. Smooth muscle actin and other myoid markers are negative.

(b) *True hemangiopericytoma*: it has a benign behaviour with no metastasis risk. It has a myogenic appearance and shows convincing pericytic differentiation. The smooth muscle actin is positive. These tumors may register recurrences.

The differential diagnosis of sinonasal hemangiopericytoma includes - glomus tumour, angiofibroma, fibrohistiocytic tumour, solitary fibrous tumour and lobular capillary hemangioma²⁵.

Immunohistochemistry is important in the diagnosis of any type of tumour. There are several markers that can help to diagnose the sinonasal hemangiopericytoma. Vimentin and CD34 are considered the only antigens to be reliably detected in tumor cells of sinonasal hemangiopericytoma, but they are not specific because they can be positive also in other neoplasms of the entity of solitary fibrous tumors²². Recent studies emphasize that the vessels of the sinonasal hemangiopericytoma partially show a positive staining for D2-40, podoplanin antibody. In case of positivity for this

marker, a diagnosis of sinonasal hemangiopericytoma may be established, because D2-40 is not expressed by any solitary fibrous tumor^{19,26}.

Clinically, the most encountered symptoms are nasal obstruction and repeated episodes of epistaxis^{27,28}. Pain may appear as a sign of local infiltration, but it occurs rarely, also as headache, vision impairment and local swelling²⁹. The most frequent localization is the nasal cavity²⁷.

There are several impediments in the establishment of the diagnosis. On one hand, in the ENT examination, it can be mistaken for inflammatory polyps³⁰ and, on the other hand, histology can find the final diagnosis, but the biopsy is not recommended because of the high risk of hemorrhage²⁷.

The radiological examination is very important. A CT scan of the paranasal sinuses shows a soft-tissue mass with strong enhancement after contrast administration, but it does not make any difference between tumor mass and inflammatory fluid. Therefore, the next step is an MRI scan. On T1-weighted MRI, sinonasal hemangiopericytoma appears as a solid isointense mass with strong contrast intake. On T2-weighted imaging, the intensity of the tumoral mass varies from isointense to low intense, in contrast to inflammatory fluid reflecting high-intense signals³¹. The conventional digital angiography offers the best visualization of the vessel supply of the sinonasal hemangiopericytoma, and it helps to plan a preoperative embolisation³².

The gold standard treatment is wide-field excision with negative margins^{33,34}. In the past, the external surgical approach was used – lateral rhinotomy, but currently endoscopic surgery is preferred, because it offers more advantages compared with the external surgery^{35,36}. First of all, the overview is better and can establish more precisely the tumor insertion and its extension³⁷. Further, the loss of blood is decreased compared with the external approach, the natural physiology of the nose can be maintained and the risk of damaging the lacrimal structures is reduced. Also, the lack of postoperative scars is another advantage^{27,38}. Total tumoral resection is recommended, but it depends on tumor size and location.

The reason for the low-grade behaviour of hemangiopericytoma is still unknown. There are theories which consider that chronic nasal obstruction and epistaxis lead to earlier diagnosis and treatment. Moreover, it was discovered the tendency to polypoid growth instead of tissue invasion, as occurs in other somatic hemangiopericytoma sites. According to the case reports presented in the literature, the prognosis of a well performed conservative treatment is very good. The conservative approach consists in debulking the tumor to obtain relief of the sinonasal obstruction. Further, a careful follow-up is necessary, with se-

rial endoscopic examinations and imaging studies. Thiringer et al.⁹ recommend aggressive resection only if the tumor shows a propensity for growth and local invasion. Head and neck hemangiopericytoma located outside the sinonasal tract has demonstrated a more malignant character and requires a more aggressive treatment plan^{39,40}.

The reported recurrence rate of HPC is quite varied, ranging from 7% to 50%, with an average time of recurrence of 6–7 years^{9,10,41}. While an incomplete primary excision has been identified as the primary factor in recurrent disease, severe nuclear pleomorphism, osseous invasion, large tumor size (more than 5 cm) and a high mitotic or proliferation rate also appear to significantly increase the risk of recurrence⁴²⁻⁴³. Long-term follow-up is essential for optimal clinical management.

CONCLUSIONS

Sinonasal hemangiopericytoma is a rare vascular tumor derived from Zimmermann's pericyte, with malignant behaviour in what it concerns the local invasion characteristics and recurrence rate.

The diagnosis algorithm is complex, involving the ENT examination, radiological investigations, and anatomopathological examination with immunohistochemistry. It has to be taken into consideration that the biopsy is not recommended because of the high risk of hemorrhage.

The complete surgical resection is the treatment of choice, and it can be obtained endoscopically, by external approach or by combined approach.

Long-term follow-up of all cases is essential for optimal clinical management.

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