



## Case series

## Sinonasal hemangiopericytoma: Report of two cases and review of literature

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## ABSTRACT

**Introduction:** Hemangiopericytoma is a low grade malignant vascular tumor. It is rare and is characterised by a poorly clinical presentation and often confused with other fibrous tumor, which could poses difficulties on diagnosis. The nasal cavity is an uncommon site and have a distinct behaviour compared to others location.

**Case presentation:** We report the case of two women who consulted for epistaxis and nasal obstruction. Vascular masses were visualized with nasal endoscopy – one in the left nasal cavity and the second one had a destructive mass of the right hemiface. CT scan and MRI helped identifying the mass, its margins and its extirpability. The Diagnosis was confirmed by histology examination. In one patient, surgery was performed. The second patient had an aggressive and invading tumor for which chemotherapy was performed. The first patient remained free of disease after surgery for a follow-up of 3 years, the second one had a fatal evolution.

**Discussion:** Surgical treatment is the important mainstay, and it still controversial regarding whether the endonasal surgery could control tumor resection despite the wide performance, nowadays, of endoscopic techniques is still controversial. The use of embolization before surgical removal in place of chemotherapy is still unclear. We report our experience through two cases.

**Conclusion:** The high rate of recurrence of this tumor requires a long follow-up after the most radical surgery possible. The external way must be adopted easily in front of any doubt of complete removal by endoscopy. Endoscopic techniques were regarded as appropriate for small low-vascularized tumors only.

## 1. Introduction

Sinonasal hemangiopericytoma, recently called hemangiopericytoma, was categorized as a borderline or low-malignant-potential tumor by the World Health Organization (WHO) in 2005. It was defined as a sinonasal mesenchymal neoplasm demonstrating a perivascular myoid phenotype [1,2].

Hemangiopericytoma (HPC) was described in 1942 by Stout and Murray as a distinctive soft tissue neoplasm presumably of pericytic origin, which exhibited a characteristic well-developed “staghorn” branching vascular pattern [3].

It is a rare tumor representing approximately 1% of all vascular neoplasms, 3% of all soft-tissue sarcomas, and less than 0.5% of all sinonasal tumors [4]. This series has been reported in line with the SCARE criteria [5].

## 2. Case presentation

## 2.1. Case 1

A 48-year-old woman presented with a 2-month history of permanent left nasal obstruction and homolateral epistaxis that appeared 2 weeks before. Nasal endoscopy revealed a mass in the left nasal cavity extended to the middle concha. There was no active bleeding and no rhinorrhea. She had no rhinological symptoms and no visual changes. The head and neck examination highlight a bleeding polypoid mass of the left nasal fossa, located inside and above the middle turbinate, in addition, there were no cervical adenopathy or other disorders. A CT of the sinuses with contrast was obtained, which showed a well-defined homogeneously enhancing mass in the left nasal cavity with extension to the adjacent ethmoid cells (Fig. 1). Magnetic resonance imaging (MRI) of the sinuses with contrast showed a well-defined homogeneously

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Fig. 1. CT scan: tumor mass of the left nasal fossa, enhancing after PDC.

enhancing mass in the left nasal cavity emerging from the superior concha and pushing the middle concha outwards without signs of an invasion (Figs. 2, 3). The patient initially underwent an endoscopic biopsy that concluded with a hemangiopericytoma of the nasal cavity.

The patient underwent a surgical removal through an endoscopic approach. In fact, this approach permitted us an overview of the tumor

insertion and its extension. We objective a firm mass crossed by a vascular network on its surface. This mass originated from the superior concha filling the left nasal cavity, pushing the middle turbinate outward and filling the middle meatus; moreover, we noticed the presence of retention liquid in the maxillary sinus and ethmoidal cells. We therefore proceed with en bloc excision of the lesion by realizing tumorectomy, a middle meatus antrostomy, and ethmoidectomy and the specimen was sent for histopathological examination. The intervention was achieved without any complications. The definitive histological examination confirms the diagnosis of hemangiopericytoma. Besides, after 3 years of follow-up, the patient remains free of symptoms. There were no signs of recurrence nor distant metastasis on objective examination by endoscopic exam associated with MRI controls.

2.2. Case 2

A 44-year-old female was diagnosed with a nasopharyngeal angiofibroma at the age of 16 years old after a history of headache, nasal obstruction, right epistaxis, and decreased right visual acuity. The tumor was judged inextirpable after CT and MRI imaging. First chemotherapy followed by radiotherapy made it possible to reduce the tumor size to 40%.

The evolution was marked by the recurrence of the mass after 9 years with extension towards the retromolar trigone and internal side of the cheek. The patient had a reduction surgery but presented a second recurrence 2 years later with an extension to the infra temporal fossa. The histological examination concluded a solitary fibrous tumor. The patient had external irradiation.

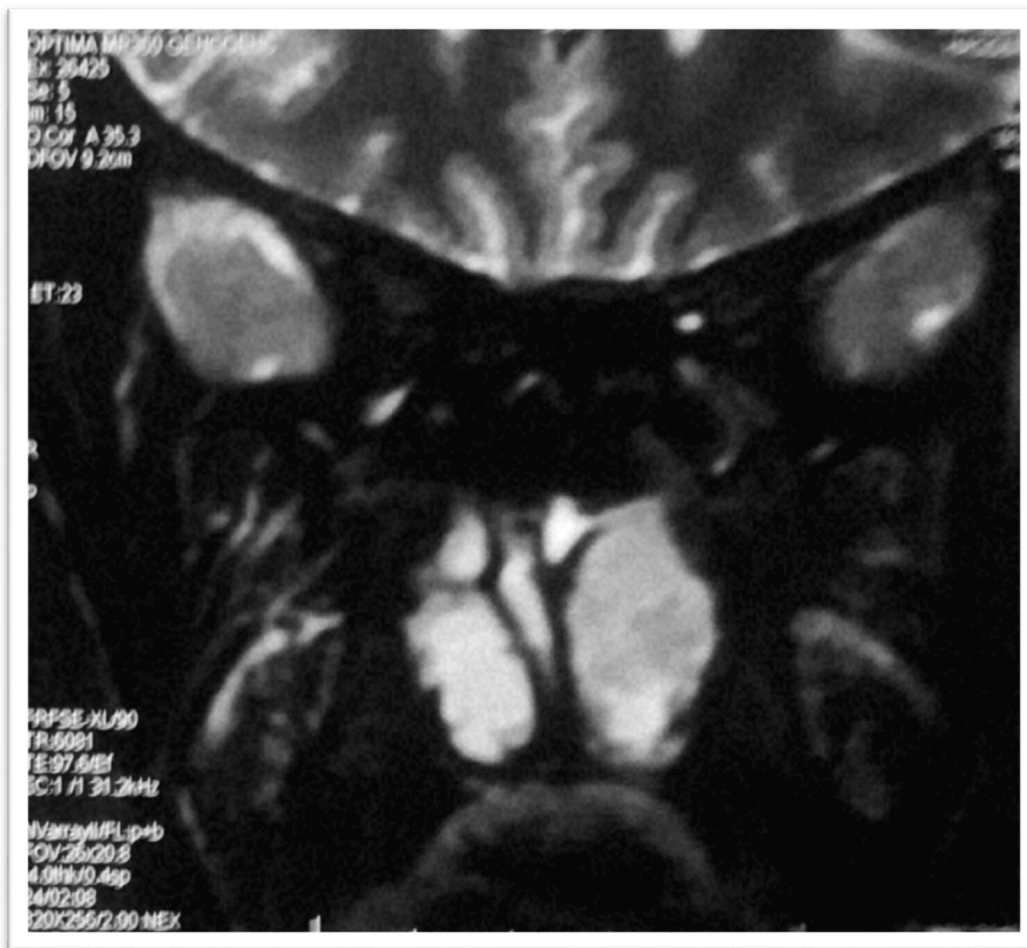


Fig. 2. Axial MRI: tumor in T2 hypersignal.

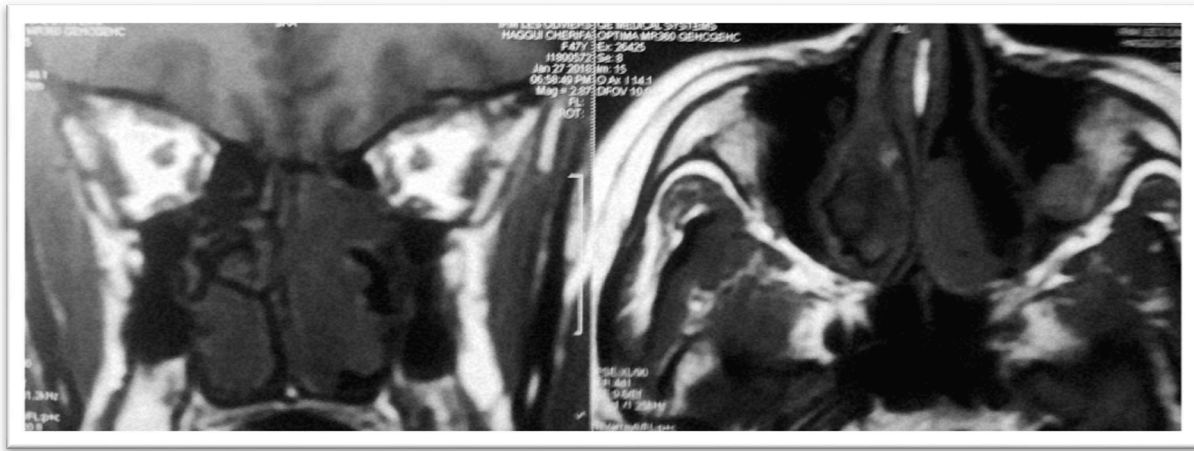


Fig. 3. Axial and coronal MRI: tumoral part of the FN dte in hyposignal T1.

A year later, the evolution was fulminant with a destructive mass of the right hemiface. A third biopsy showed monophasic synovial sarcoma. Imaging showed a very aggressive extension of the tumor enhancing after injection of PDC (Figs. 4, 5, 6 and 7). Faced with the unusual clinical and progressive aspect, a histological revision was carried out and the histological nature was corrected in favor of an initially low-grade hemangiopericytoma which transformed into a high-grade one. The disease didn't metastasize. We opted for chemotherapy but, unfortunately, the outcome was fatal.

### 3. Discussion

Over the years, it has appeared that hemangiopericytomas show a nonspecific growth pattern that is shared by numerous unrelated benign and malignant lesions. HPC was better considered as a diagnosis of exclusion [6].

It is found mostly in tissues within creased vascularity and mostly occurs in the low extremities, pelvic cavity, and retro-peritoneum [7].

Fifteen to sixteen percent of all is found in the head and neck region with a tendency to appear in the nasal cavity and paranasal sinuses. Sinonasal hemangiopericytoma (SNHPC) involves mostly the ethmoid and sphenoid sinuses [4,8].

These tumors can occur at any age, however, the peak incidence is usually between the 5th and 7th decades of life. An equal to slight female predominance [9,10] in our case, the two patients were women.

The etiology remains unknown; however, predisposing factors such as past trauma, hypertension, pregnancy, and the use of corticosteroids are considered [8,9]. We didn't identify any risk factors in our patients.

Clinical presentation is usually unilateral nasal obstruction, recurrent epistaxis, or both. Difficulty in breathing, visual disturbance, pain, and headache are less frequent symptoms [4,9]. In our study, nasal obstruction was the main symptom followed by the epistaxis, then headache was present in the second patient and revealed a locally advanced stage tumor.

On examination, SNHPC is usually unilateral, appears as a red to pink polypoid mass without surface ulceration that is around 3 cm in size, and can only be differentiated from similarly presenting tumors such as lobular capillary hemangiomas, solitary fibrous tumors, and glomus tumors through histochemical analysis [11]. In our cases, we highlight on the clinical exam a non-specific polypoid mass without any cervical lymphadenopathy.

CT findings are non-specific. On MRI, literature shows typically hyperintense signal on T2WI with vascular signal voids, a high mean ADC value, and a wash-in and washout pattern on DCE-MR imaging. MR imaging findings, including the ADC value and DCE-MR imaging pattern, can help differentiate glomangiopericytomas from other

hypervascular tumors in the head and neck, especially in the sinonasal cavity [12]. In our two cases, CT scan and MRI describe the extension tumor without giving any specific signs related to hemangiopericytoma.

Histological analysis shows that the neoplasm consisted of uniform, monotonous cells, exhibiting minimal pleomorphism. A few mitotic figures can be seen without significant apoptosis or necrosis associated with the lesion. The cells appear to focally palisade around the vessels. Immunohistochemistry can show strong expression of vimentin and focal expression of smooth-muscle actin (SMA) [6]. In the first case were ported, the histological exam confirmed the diagnosis, but in the second one, the diagnosis of hemangiopericytoma was missed then revised owing to the fact of unusual tumor progression.

Concerning treatment, surgery by realizing a total excision is still the gold standard. Although the high degree of vascularization, which makes the removal of these tumors usually challenging [13], the place of the pre-operative angiography in the management of SNHPC is still controversial. But many authors, in order to facilitate preoperative planning and to enable embolization, indicate angiography particularly in large tumors [9]. Afferent vessel embolization can help in surgical procedures. Indeed, the embolization of the ophthalmic artery was described by many authors before a massive sinonasal HPC was removed [6]. In addition, it has been noted a significant reduction in the risk of intraoperative hemorrhage after angiography [9].

In our report, concerning the first case, the tumoral resection was obtained solely by surgery; there was no need for preoperative embolization, we judged it inappropriate as tumor extension because it was localized. Then, the tumor resection with free margins was totally controlled by endoscopic approach.

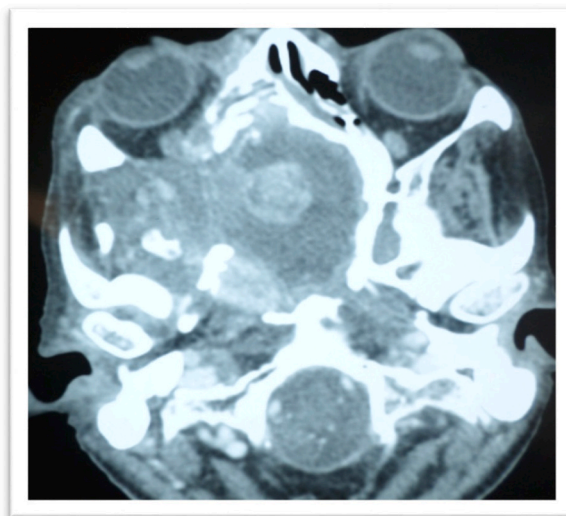
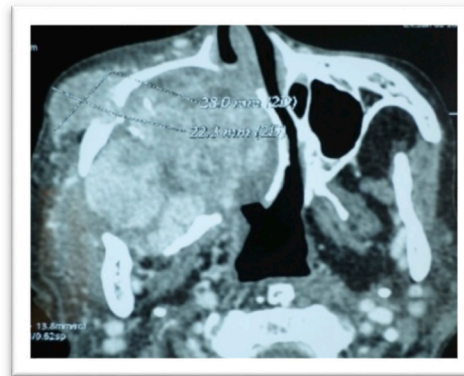
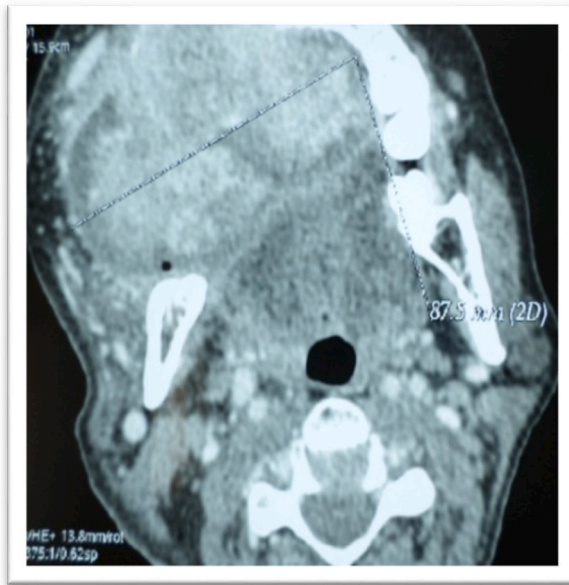
Traditionally, SNHPCs are treated by wide surgical excision through an open craniofacial approach. Recently, endonasal endoscopic removal has become popular with no significant statistical difference in the rate of recurrence reported in the literature between the two approaches [14]. In fact, nowadays, endoscopic surgery of the nasal cavity and paranasal sinuses is considered as the primary therapeutic option for the benign as well as the malignant neoplasms [9].

The most significant advantages of endoscopic surgery are improved visualization and simplicity of post-operative monitoring of the surgical site, preservation of the natural physiology of the nasal cavity and the paranasal sinuses, and avoidance of scars; another reason is that the external approach is known by cause more bleeding than the endoscopic approach [9].

However, open surgery still have a place in instances when the paranasal sinuses are heavily involved, when there is extension to the ocular structures and/or involvement of the superficial tissues, or the fossa pterygopalatina [9].

In comparison to the high rates reported for open procedures, major





**Figs. 4, 5, 6 and 7.** CT of the facial mass in coronal and axial section: poorly limited tumor mass, centered on the right ethmoid nasal region of 8.7 cm, intensely enhanced tissue and in a heterogeneous way, extending towards the maxillary sinus, the infratemporal fossa + the sphenoidal sinus + lysis of the orbital floor.

problems following endoscopic skull base surgery are quite infrequent [9].

Cerebrospinal fluid leaking is the most common significant complication following endoscopic skull base surgery. It occurs in around 5% of cases, and it may be treated with a lumbar spinal drain or further endoscopic surgery [9].

Radiotherapy and chemotherapy may be necessary in situations with an unresectable primary tumor or metastatic illness. Radiotherapy is used for non radical surgical resection, inoperable tumors, or metastases cases. Additionally, adjuvant radiation can be utilized to manage local illness in combination with surgery [6].

Regarding our first case, she was successfully treated by endonasal approach surgery. The course intervention was bloodless. The second case was treated by radiotherapy and chemotherapy because of the extension of the tumor which became unresectable.

The prognosis is usually favorable and depends on the mitotic activity in the tumor [15]. Such situation was the case of our second patient, in a way that extension tumor causes her death.

Contrary to the other localization of this tumor in the body, the sinonasal hemangiopericytoma is known by having a low tendency for metastasis. The prognosis of GPC is usually favorable and depends particularly on the mitotic activity in the tumor [4].

In his study on 104 patients with sinonasal HPC, Thompson et al. reported a disease-free survival rate of 74.2% at 5 years and 64.4% at 10 years. Overall the prognosis for patients with the sinonasal type of HPC is favorable, as the raw 5-year survival rate in the study by Thompson et al. was as high as 88% [16].

#### 4. Conclusion

Sinonasal hemangiopericytomas are slowly progressive tumors that can be highly vascularized, involving the skull base, and requiring extensive resection.

Surgical treatment is the gold standard for this type of tumor that is relatively radio-resistant. The role of preoperative embolization is yet to be defined.

Due to the indolent nature of these tumors, long-term follow-up is preferable to monitor any recurrences, however, the prognosis is usually favorable.

#### Consent

Written informed consent was obtained from the 2 patients for publication of this case report and accompanying images.

#### Provenance and peer review

Not commissioned, externally peer-reviewed.

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This is a case report study and ethical approval not required.

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#### Guarantor

Professor Mohamed Abdelkefi.  
Dr Meherzi Abir.

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#### CRediT authorship contribution statement

MEherzi Abir: conception and design of the study  
Omri Malika, Bellakhdher Mouna, Hwass Jihen: acquisition of data  
Meherzi Abir, Khalifa Mouna: article drafting  
Wassim Kermani, Mouna Bellakhdher, Mohamed Abdelkefi: final approval of the version to be submitted.

#### Declaration of competing interest

None declared.

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