Sinonasal hemangiopericytoma: two case reports and review of literature.

abir Meherzi (abir-meherzi@hotmail.fr)
University of Sousse: Universite de Sousse
Mouna Khalifa
University of Sousse: Universite de Sousse
Malika Omri
University of Sousse: Universite de Sousse
Mouna Belakhdher
University of Sousse: Universite de Sousse
Jihen Hwass
University of Sousse: Universite de Sousse
Wassim Kermani
University of Sousse: Universite de Sousse
Mohamed Abdelkefi
University of Sousse: Universite de Sousse

Research Article

Keywords:

Posted Date: March 30th, 2022

DOI: https://doi.org/10.21203/rs.3.rs-1404876/v1

Abstract

Hemangiopericytoma is a rare tumor that arises from pericytes, classified as a low-grade malignant vascular tumor. It is characterised by a poorly clinical presentation and histologically, often confused with other fibrous tumors, which could pose difficulties on the diagnosis. The nasal cavity is an uncommon site. The rarity of this tumor imply the absence of management guidelines. So far, it still controversial regarding wether the endonasal surgery could control the tumor resection despite the wide performance, nowdays, of endoscopic techniques. It still unclear also conserning the use of embolization before surgery removal and the place of chemotheray. The high rate of recurrence impose long-term follow-up. So that, we report our experience through two cases and we aimed to discuss the clinical features and the differents treatement procedures. In fact, they are 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 and MRI helped identify the mass, its margins and its extirpability. DiagnosIs was confirmed by histology. In one patient, surgery was performed. The second patient had an agressive and invading tumor for which chemotherapy was performed. The first patient remained free of disease after surgery. The second one had a fatal evolution.

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]

We present our findings in two cases, with the goal of discussing clinical characteristics and treatment options.

Case Presentation

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 scan 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 homogenously enhancing mass in the left nasal cavity emerging from the superior concha and pushing the middle concha outwards without signs of an invasion (fig2,3).

The patient initially underwent an endoscopic biopsy that concluded with a hemangiopericytoma of the nasal cavity. She underwent endonasal surgery: middle meatus antrostomy, ethmoidectomy, and tumorectomy. The postoperative course was simple; there was no relapse after 3 years of regular follow-up by endoscopic examination associated with MRI controls.

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 juged 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 infratemporal fossa. The histological examination concluded a solitary fibrous tumor. The patient had external irradiation.

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 agressive extension of the tumor enhancing after injection of PDC (fig4, 5 ,6 and7). 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.

Discussion

Over time, Hemangiopericytomas have been found to have a general growth pattern that is shared by a variety of unrelated benign and malignant lesions over time. HPC was better considered as a diagnosis of exclusion. [5]

It appears mostly in tissues with increased vascularity and is mostly occurs in the lower extremities, pelvic cavity, and retroperitoneum.[6]

Fifteen to sixteen percent of all are found in the head and neck region with a tendency to grow in the nasal cavity and paranasal sinuses. Sinonasal hemangiopericytoma (SNHPC) involves mostly The ethmoid and sphenoid sinuses. [4, 7]

These tumors can occur at any age, however, the peak incidence is usually between the 5th to 7th decades of life. An equal to slight female predominance was noted [8, 9].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[8–10].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, 8]. 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. It measures on average 3 cm. Only histochemical examination can distinguish it from tumors that show similaritie, such as lobular capillary hemangiomas, solitary fibrous tumors, and glomus tumors. [11]. In our cases, we highlight on the clinical exam a non-specific polypoid mass without any cervical lymphadenopathy.

CT scan findings are non-specific. On MRI, literature shows typically hyperintense signal on T2WI (T2 weighted image) with vascular signal voids, a high mean ADC (apparent diffusion coeficient) value, and a wash-in and washout pattern on DCE-MR imaging dynamic contrast-enhanced MRI.

MR imaging findings, including the ADC value and DCE-MR imaging pattern, can help differentiate hemangiopericytoma from other hypervascular tumors in the head and neck, especially in the sinonasal cavity[12]. In our two cases, CT scan and MRI describes 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). [5] in the first case we reported, 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 is still the gold standard.The high degree of vascularization makes the removal of these tumors usually challenging. A lateral rhinotomy is traditionally performed. Endoscopic approaches are, however, being increasingly used in managing sinonasal and skull tumors. [13]

Traditionally, SNHPCs are treated by wide surgical excision through an open craniofacial approach. Recently, endonasal removal has become popular with no significant statistical difference in the rate of recurrence reported in the literature between the two approaches [14]. Regarding our first case, she was successfully treated by external approach surgery.

Radiotherapy is used for nonradical surgical resection, inoperable tumors, or metastases cases. 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.

Preoperative angiography in the management of SNHPC is still controversial.

To facilitate preoperative planning and to enable embolization, many authors indicate angiography in large tumors [8]. It has been noted a significant reduction in the risk of intraoperative hemorrhage after angiography[8]. In our patients, the tumoral resection of the first case was obtained by solely surgery ; there were no need to a preoperative embolization.

The prognosis of hemangiopericytoma is usually favorable and depends on the mitotic activity in the tumor [4]. Indeed, in a study of 104 patients with sinonasal HPC, Thompson and 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].

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 natural behaviour of these tumors, and the high rate of recurrence a long-term follow-up is preferable to monitor any relapse, however, the prognosis is generally favorable as long as conditioned by a complete resection.

Abbreviations

WHO : world health organization

HPC : hemangiopericytoma

CT scan : computed tomography.

MRI : magnetic resonance imaging.

SNHPC : Sinonasal hemangiopericytoma.

ADC : apparent diffusion coeficient.

DCE-MRI : dynamic contrast-enhanced MRI.

SMA : smooth-muscle actin.

T2WI : T2 weighted image.

Declarations

Ethicsapproval and consent to participate:we don’trequire ethical approval in our institution for reporting individual cases, and we obtain verbal consent.

Consent for publication: the 2 patients gave their permission to publishtheir cases through a written consent

Consent for publication:Written informed consent was obtained from the patient for its anonymized information to be published in this article.

Availability of data and material: All data and material of this case are available.

Competing interests:we have no competing interests

Funding: none.

Author’scontributions: MA: charged of data collection, following up the case,participating in writing and supervising the manuscript preparation; KM: formulated the idea and participate in writingthe manuscript preparation; OM,BM, and HJ participate in analyzing and interpreting data, KW and AM: revising and approved the final version

Acknowledgments “Not applicable” for this section.

Figures

Figure 1

CT scann : tumor mass of the left nasal fossa, enhancing after PDC

Figure 2

Axial MRI: tumor in T2 hypersignal

Figure 3

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

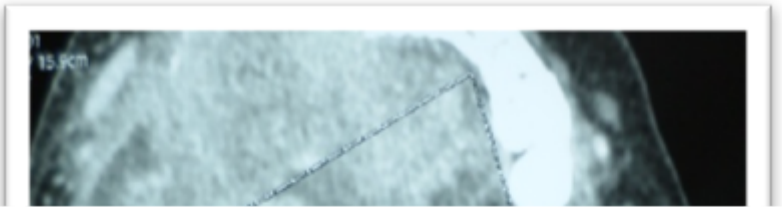

Figure 4

CT of the facial mass in coronal and axial section: poorly limited tumor mass, centered on the right ethmiodo nasal region of 8.7 cmn, intensely enhanced tissue and in a heterogeneous way, extending towards the maxillary sinus, the infratemporal fossa + the spheniodal sinus + lysis of the orbital floor

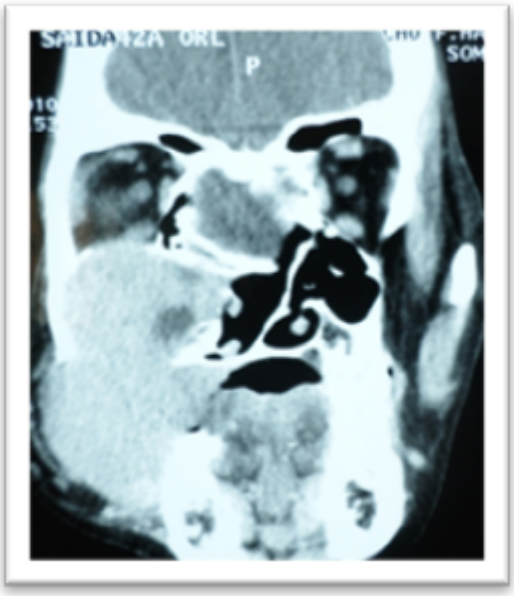

Figure 5

CT of the facial mass in coronal and axial section: poorly limited tumor mass, centered on the right ethmiodo nasal region of 8.7 cmn, intensely enhanced tissue and in a heterogeneous way, extending towards the maxillary sinus, the infratemporal fossa + the spheniodal sinus + lysis of the orbital floor

Figure 6

CT of the facial mass in coronal and axial section: poorly limited tumor mass, centered on the right ethmiodo nasal region of 8.7 cmn, intensely enhanced tissue and in a heterogeneous way, extending towards the maxillary sinus, the infratemporal fossa + the spheniodal sinus + lysis of the orbital floor

Figure 7

CT of the facial mass in coronal and axial section: poorly limited tumor mass, centered on the right ethmiodo nasal region of 8.7 cmn, intensely enhanced tissue and in a heterogeneous way, extending towards the maxillary sinus, the infratemporal fossa + the spheniodal sinus + lysis of the orbital floor

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- CAREchecklistEnglishhemangiopericytome.docx