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Hemangio Pericytoma of the Nasal Cavity

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Authors' contributions

This work was done in collaboration with all authors. Authors RA, YO designed the bibliographic research and wrote the first draft of the manuscript. Authors SR, MR managed the discussion and analysis of the study. Author MM managed the documentary research. All authors have read and approved the final manuscript.

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Case Study

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ABSTRACT

Hemangiopéricytoma (HP) is a rare, slow growing intranasal vascular tumour. Initially not very symptomatic, it is difficult to diagnose. The clinician should be alerted by a persistent unilateral nasal symptomatology and by the endoscopic appearance of a gray polypoid tumor. The diagnosis of certainty is histological, but biopsies are delicate, given the risk of hemorrhaging. The malignant potential of this tumor involves surgical removal. The pre-therapeutic assessment includes a naso-sinus scanner with iodine injection and magnetic resonance imaging (MRI). Some propose arteriography with pre-operative embolization. The reference surgical technique remains the para-latero-nasal pathway but endoscopic excisions have recently been reported.

Keywords: Hemangiopéricytoma; rhino-sinus tumor; endonasal surgery.

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1. INTODUCTION

The haemangiopericytoma is a rare vascular tumor. The first case of nasal hemangiopericytoma was described by Stout and Murray in 1942. This tumor grows from capillary pericytes. The nasal location is less aggressive than other areas of the body, but it has higher recurrence rates [1,2]. The main rhinologic clinical manifestations are recurrent epistaxis and nasal obstruction. The diagnostic difficulty is a major problem especially as the clini- cal features are nonspecific and the architectural study must set aside several diagnostics to conclude this histological type. A post operative care is recommended for diagnosing recurrences that could occur years later.

2. CASE REPORT

Ms E., 34 years old and in an excellent general condition, has been referred for a fourth recurrence of intranasal tumor hemangio-péricytoma type. It had been evolving for a year after three surgeries for the same tumor, combined with a pansinusite resistant to different medical treatments, combining antibiotic and corticosteroid therapy. During interrogation, he complained of a bilateral nasal obstruction that had been developing for a year after his surgery and was gradually worsening, associated with daily frontal headaches, and a posterior rhinorrhea. The rhinological examination objectified a polypoid lesion, of purplish colour, fleshy and firm on palpation, not hemorrhagic, filling the entire nasal cavity and externalized to the right (Fig. 1).

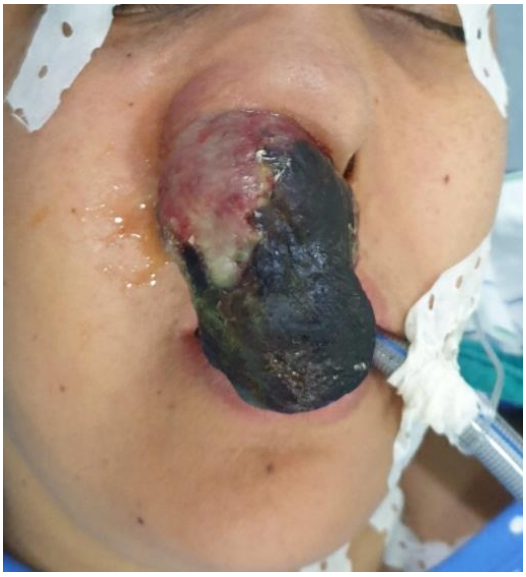

Fig. 1. The entire nasal cavity and externalized to the right

CT scan showed a tissue process centered on the right naso-sinus cavities, taking the contrast in a heterogeneous way, measuring approximately 111mm 51mm 68mm with filling of the maxillary, spheroidal and ethmoidal sinuses and lysis of their walls, it extends beyond the spheroidal sinus at the upper wall of the cavum. It invades the skull base with lysis of the round and oval foramen with basic-temporal endocranial extension. It invades the cavernous sinus, parapharyngeal and pre-stylian space (Fig. 2).

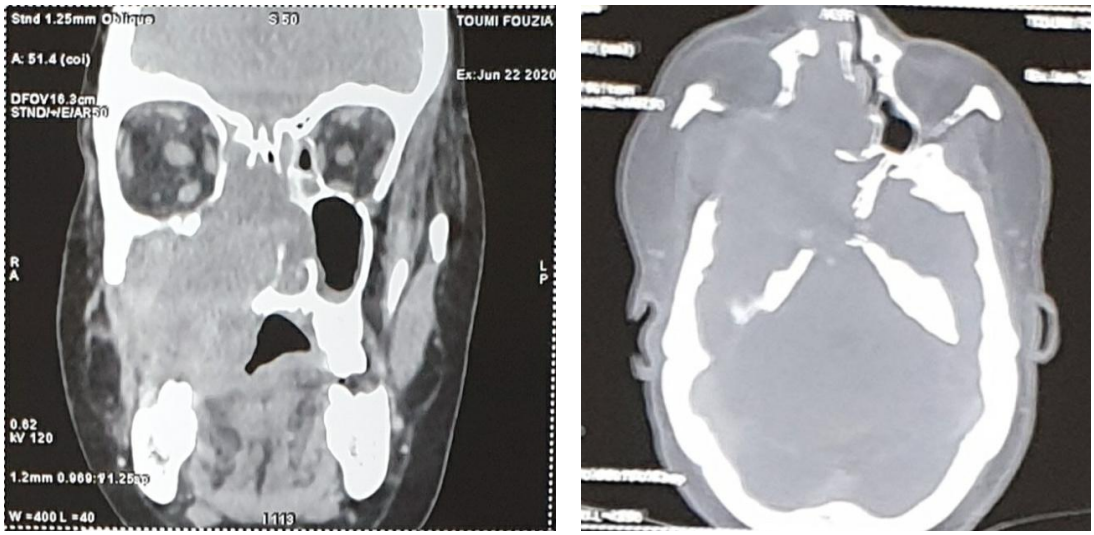

Fig. 2. CT scan view of cavernous sinus, parapharyngeal and pre-stylian space

It fills the entire right infra-temporal pit. At the top it lyses the orbital floor with separation edging with the lower right muscle. The CT scan has been completed by an MRI. The exploration made it possible to identify a lesional process centered on the right nasal cavity in free hyposignal homogeneous in T1 and in heterogeneous signal in T2. The lesion appeared hypervascularized, enhanced after gadolinium injection, with endocranial extension. Given the atypical nature of this bilateral "polyposis", new biopsies were carried out. The histopathological examination revealed a benign tumor lesion consistent with HP. A surgical treatment was proposed and the pre-therapeutic assessment was supplemented by an arteriography, in order to specify the tumor vascularization and to evaluate the possibility of pre-operative embolization. In the case of Ms E., the arteriography showed an intense vascular blush, developed only at the territory of the anterior ethmoidal artery. There was no vascularization from the branches of the external carotid artery; embolization was therefore not performed. Surgical removal of the tumour was performed by endonasal route, endoscopic guidance and computer-assisted navigation. Once the area of insertion of the lesion and its contacts to the orbital wall and skull base were clearly identified, the excision was carried out centrifugally until the maximum of the tumor attached to the anterior and posterior ethmoid region was released. The excision was macroscopically incomplete because of the extension to the skull base, and the part was sent to histological examination. During the procedure, the tumor bled little despite the absence of pre-operative embolization. The roller gauze was gradually removed from the second post-operative day. The procedures were simple, with multidaily saline washes for a month. The histopathological examination found a proliferation of fusiform cells without cytonuclear atypia, richly vascularized and compatible with HP. No signs of necrosis were found in favor of a malignant character. Immuno-labeling was performed with smooth anti-actin muscle antibodies and CD34. Their positivity confirmed the diagnosis of sinus-nasal HP.

3. DISCUSSION

HP is a rare tumor of vascular origin, ubiquitous topography. It develops from the pericytes, cells localized in the walls of the capillaries. HP represents only 1% of all vascular tumours and is found in 15% of cases in the head and neck region. Only 5% of these tumors of the ENT sphere sit in the rhino-sinus cavities. Most studies report a sex ratio close to 1 [1], with the exception of a recent series (104 cases) indicating a discreet female predominance [3]. HP can occur at any age with an average close to 60 years [3]. Often, as in the case of Ms E., the clinical picture evolves for several months after three previous surgical procedures for the same tumor. In the Thompson series, the time between diagnosis and onset of symptoms ranges from 1 to 60 months, with an average duration of 10 months [3]. The two most common initial clinical symptoms are unilateral nasal obstruction and recurrent epistaxis. Other symptoms can complete the symptomatology: headaches, feeling heavier, a mucous or purulent rhinorrhea or odor disorders [3]. The clinical picture is often incomplete and not very specific. In the case of our patient, unilateral nasal obstruction and headache were the present, dominant and persistent symptoms. More than the symptoms themselves, their evolution towards "chronicity" and, above all, their persistence despite appropriate medical treatments motivated the consultation with an ENT and the continuation of investigations in search of a tumor process. The clinical examination makes it possible to note the presence in the nasal cavity of a unilateral mass, which may have a polypoid appearance and whose size is on average 3,1 cm [3]. In the literature, its tanned appearance varies from purplish red to pinkish gray, and its texture appears soft and grainy. It is described as friable and hemorrhagic on palpation, but never ulcerated [2]. Its development is usually intranasal, rarely strict intra-sinusal [3].

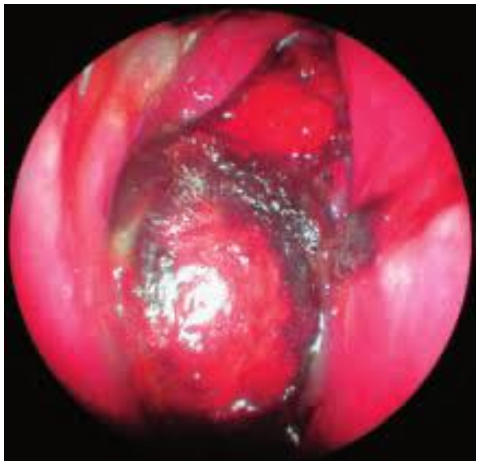

Fig. 3. Endoscopic view after exeresis of the extra nasal part of the tumor

The initial X-ray includes a millimetre scan of the rhino-sinus cavities, as well as an MRI. These two tests make it possible to carry out the loco-regional balance of the tumor extension and to identify its exact limits. The CT scan is ideally performed with contrast injection. The tumor has a density close to that of soft tissues. The difference in density between tumour tissue and mucous secretions makes it possible to distinguish them and individualize the tissue mass enhanced by the contrast agent. Unfortunately, the associated contrast of the inflammatory sinus mucosa may lead to errors in the analysis of tumor extension limits. MRI is more specific. T1-weighted iso-signal and iso-, or hyposignal T2 characterize the solid tumor. Furthermore, the gadolinium injection strengthens the tumor's signal and allows the search for invasiveness of the dura [2]. The assessment may be supplemented secondarily by carotid arteriography [2]. The interest of carotid arteriography is discussed in the literature. It allows the analysis of tumor vascularization for pre-operative embolization [4], in order to reduce the risk of hemorrhagic during tumor excision. During the management of Ms E.,an arteriography was requested, but the tumor could not be embolized. Despite this, the tumor resection was little hemorrhagic. The diagnosis of certainty is histological [3], using a study in immuno-histochemistry and sometimes requiring several biopsies [5]. The study immuno-histochemical is done with several markers. There are no specific markers of sinus-nasal HP. Unlike soft tissue HP, it preferentially expresses muscle markers (smooth muscle actin), but also factor VIII, laminine and CD34. The immuno-histochemical and histological aspect of sinusonasal HP is very similar to that of solitary fibrous tumours, initially described at the level of the pleura. This similarity suggests the existence of a common cell differentiation, and calls into question sinus-nasal HP as a distinct entity [1,3]. The aggressiveness of this tumour is assessed according to its histological grade, size and limitations. There is no consensus in determining the criteria for malignancy. The histological elements to be taken into account are the existence of necrosis or haemorrhage and nuclear abnormalities. According to the literature, the existence of mitotic elements is suggestive of malignancy. The evolution can then be marked by loco-regional recidivism as well as by the occurrence of metastases. The reported local recidivism risk varies, depending on the team, from 25% to 40%. The metastatic potential is also unevenly estimated between 5% and 57% [2,6]. Metastatic locations are described at the cerebral, pulmonary, bone and hepatic levels. The therapeutic attitude depends on the initial clinical presentation and the evaluation of the degree of tumor aggression. According to the various studies, the curative treatment is surgical [3,6]. Tumor resection is preferably carried out by para-lateral-nasal route. The endonasal pathway may be proposed in cases where the tumour is strictly intranasal or developed at the ethmoid or the sphenoid sinus, where the limits of tumor excision are well controlable endoscopically [5]. In the case of our patient., the loco-regional extension was important at the level of the nasal cavity, ethmoidal cavities, right orbital cavity and the skull base. The examinations performed showed no other localization at the neck. Based on these results, endonasal surgical treatment was chosen. Additional treatment with radiotherapy is performed by certain teams, according to histological malignancy criteria and the risk of local recurrence [6].

4. CONCLUSION

HP is a rare tumor of the naso-sinus cavities, of vascular origin, which requires a broad exeresis given the risk of local recurrence and its malignant potential which remains poorly defined.

CONSENT

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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