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Case Report

Sinonasal Haemangiopericytoma: A Rare Case

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Abstract

The haemangiopericytoma is a rare vascular tumor. It is developed from capillary pericytes. His nasal sinus location only 0.5%. Its diagnosis is mainly based on the biopsy with histological study. The treatment is depends on the evolutionary stage. It uses radiotherapy, endovascular embolization or surgery. The authors report a case of a young 45 year old patient who presented a tumor sinonasal rapidly progressive in the diagnostic assessment, pathologic study showed a haemangiopericytoma.

Keywords: Haemangiopericytoma; Vascular Tumor; Nasal Sinus Location; Radiotherapy; Endovascular Embolization; Surgery

Introduction

The haemangiopericytoma is a rare vascular tumor [1,2]. The first case of nasal hemangiopericytoma was described by Stout and Murray in 1942 [3]. This tumor grows from capillary pericytes. The nasal location is less aggressive than other areas of the body, but it has higher recurrence rates [1,3]. The main rhinologic clinical manifestations are recurrent epistaxis and nasal obstruction [4]. The diagnostic difficulty is a major problem especially as the clinical 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 [1-3]. The aim of this work is to highlight the scarcity of nasal sinus haemangiopericytoma and talk through our observation and literature review about diagnostic features, treatment and prognosis.

Case Presentation

We report the case of a 45-year-old male patient without particular medical history or facial trauma, presented the consultation of Otorhinolaryngology (ENT) and Neck Surgery for chronic rhino-sinus dysfunction 6 months ago, combining a nasal obstruction with right anterior rhinorrhea complicated multiple episodes of epistaxis average abundance. The symptomatology was complicated a three months after by a decrease in the right visual acuity, right proptosis and exotropia (Figure 1). Nasal endoscopy found a vascularized tumor bleeding on contact. The rest of the ENT examination was unremarkable including no cervical lymphadenopathy. A CT scan performed with contrast material injection showed a tumor vascular process of the right nasal cavity extended to left with lysis intersinuso-nasal wall and extending to the right orbit, infiltration of muscles and discharge of the eyeball (Figure 2).

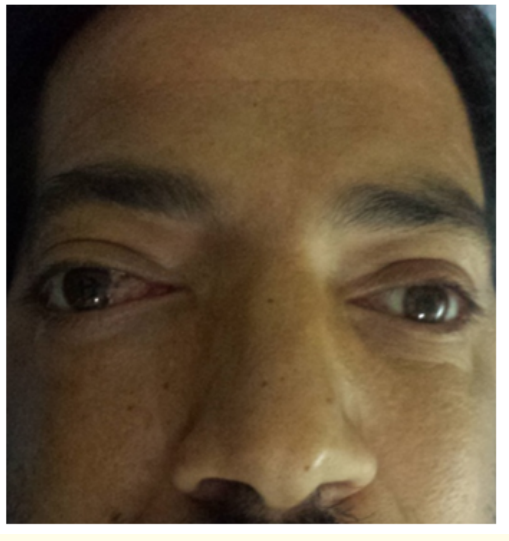

Figure 1: Clinical appearance of divergent strabismus and non axile right exophthalmia.

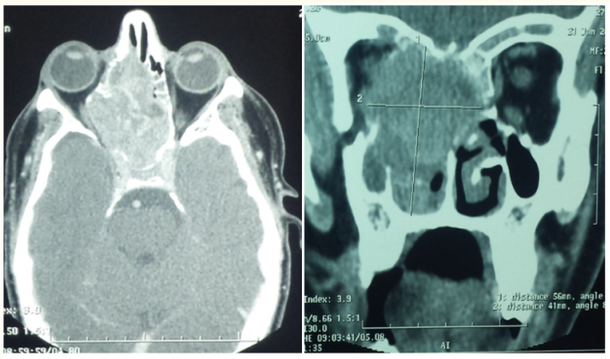

Figure 2: Sinus scanner through the vascular tumor process.

Axial (right): Filling the nasal passages with lysis intersinuso - nasal wall and extension to orbit infiltration of muscles and optic nerve and the eyeball refoulement.

Coronal (left): extension to the right maxillary sinus and invasion of the mastoid Crista Gali (black arrow) and the base of the skull.

The patient underwent a endonasal biopsy under general anesthesia, the surgical procedure was marked by severe bleeding (Figure 3). The pathological examination had showed small round cells to medium size, with large eosinophilic cytoplasm and myxoid aspects, they have around arteriolar vessels are thickened muscle wall endothelium and turgid. Immunohistochemical study was positive for vimentin with a negativity of CD34, the HMB45 and PS1000 and was retained diagnosing a sinonasal hemangiopericytoma (Figure 4). The patient underwent an endovascular embolization 24 hours before the surgical procedure (Figure 5). Endoscopic surgery has been performed with reduction in tumor volume. The bleeding was profuse despite hemostasis by bipolar electrocautery and adrénalinées wicks. The operation was stopped and the patient had received postoperative radiotherapy. The patient was reviewed in consultation with significant reduction in tumor volume and followed with a decline of one year.

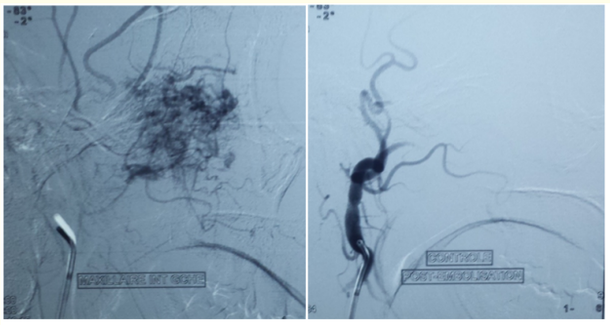

Figure 3: Carotid angiography before endovascular embolization (left) and after embolization of vascular tumor of the nasal cavity (right).

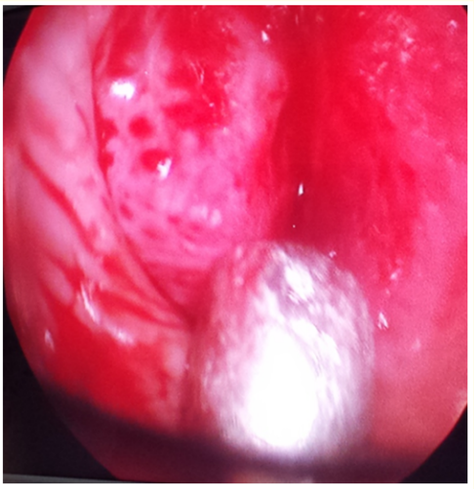

Figure 4: Endoscopic vascular tumor of the right nasal cavity.

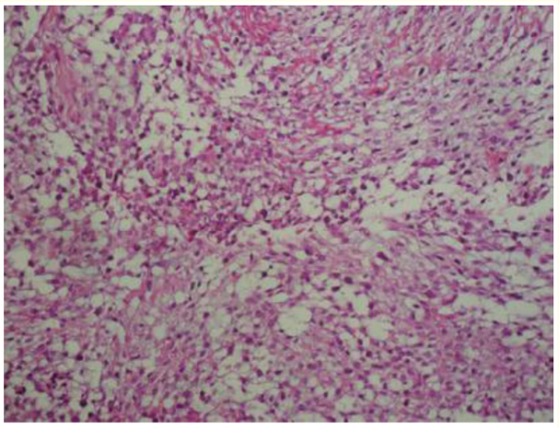

Figure 5: Histology of the haemangiopericytoma with proliferation of spindle cells.

Discussion

The haemangiopericytoma nasal sinus is a vascular tumor, developed from mesenchymal cell differentiation péricytaire, described for the first time by Stout and Murray [3]. It is classified by the World Health Organization (WHO) among tumors with low malignant potential [5]. The sinonasal hemangiopericytomas represent less than 0.5% of tumors of the nasal cavity and para nasal sinuses [5]. All ages can be affected with a slight female predominance [4,6]. No predisposing factor was highlighted, apart from a notion of facial trauma that is found in the history of several patients. The pathophysiological hypothesis mentioned in this study was a proliferation of capillary vessels and pericytes in the healing process after trauma [7]. In the case of our patient, there was no notion of facial trauma.

The most common functional signs are unilateral nasal obstruction and epistaxis. Other rarer signs were reported as visual disturbances, otitis média with effusion, pain in the face and headache [8,9]. The endoscopic appearance is a polypoid tumor reddish gray to pink (Figure 3). The histological diagnosis of haemangiopericytoma is difficult because the tumor has no histological profile and immunohistochemical characteristics. Since 2002, WHO classifies the tumor fibroblast and myofibroblast group [5]. It is a diagnosis of exclusion for which to exclude the differential diagnoses that may share with him histological similarities. Indeed, it is not difficult to distinguish hemangiopericytomas sarcomas that are clearly smarter, difficulty arises especially for tumors with low malignant potential such as the myogenic granuloma, solitary fibrous tumor, leiomyoma and 'angiofibrome [10]. Thanks to immunohistochemical examination, we can distinguish sinonasal hemangiopericytomas from other locations by their increased expression of vimentin, actin and XIIIA factor and a lower expression of CD34. The Bcl-2, CD99 and CD117 were negative. [5] In the case of our patient, immunohistochemistry showed positivity for vimentin and negativity of D34 and protein 100.

Imaging should include a computed tomography (CT) scans to assess bone disease and a magnetic resonance imaging (MRI) to better appreciate the characteristics of the tissue damage These examinations show a tissue lesion enhancement after contrast injection in CT, and gadolinium MRI [10,11]. Several authors recommend angiography within large tumors, for embolization which would reduce the risk of bleeding intraoperatively [12]. Surgery is the treatment of choice for sinonasal hemangiopericytomas. The endoscopy is recommended as first-line because of its lower morbidity [13].

Instead of radiotherapy and chemotherapy for sinonasal hemangiopericytomas remains controversial. Chemotherapy has not proven his efficiency [6]. External radiation therapy is indicated in cases of extensive tumors as is the case with our patent [8-10]. The recurrence rate of sinonasal hemangiopericytomas varies from 7 to 50%, with an average time to recurrence of 6 to 7 years [12]. The main factor implicated in recurrence is complete or incomplete resection. Other factors also increase the risk of recurrence, such as large tumor size, bone disease, a severe nuclear pleomorphism and a high mitotic index [9,11]. In the case of our patient, there is an associated bone disease, a higher than average tumor size and non-healthy resection margins. Distant metastases are extremely rare for sinonasal hemangiopericytomas [12], which is consistent with the benign course of these tumors with a survival rate of over 90% at 5 years [4].

Conclusion

The haemangiopericytoma nasal sinus is a rare tumor of vascular origin, which differs from other locations of haemangiopericytoma by its low grade malignancy and better survival rates. The difficulty of the histological diagnosis is a major problem especially as the clinical features are nonspecific and the architectural study must rule out differential diagnoses. A post operative care is recommended for diagnosing recurrences that can occur years later.

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