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Case Report

# Intravascular Papillary Endothelial Hyperplasia: An Unusual Tumor of the Paranasal Sinuses

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## Abstract

Intravascular Papillary Endothelial Hyperplasia (IPEH) also known as Masson's Tumor was first described in 1923 by French pathologist Dr. Claude L. Pierre Masson. It's a reactive vascular proliferation secondary to traumatic vascular stasis which unleashes the activation of FGF of the macrophages invading the site of trauma with proliferation of endothelial cells. The importance of Masson's tumor, of a benign nature, is the striking similarity it has with the angiosarcoma. This tumor represent 2% of all the vascular tumor, more frequently seen in the skin of fingers and blood vessels. Its presentation in the paranasal sinus is extremely rare, with only 10 published cases in medical literature. The present case is of a 41 year old female with paresthesia of the left side of the face and periorbital edema: treated with steroids and antibiotic without improvement. Physical exploration with left palpebral ptosis, diplopia and diminishes visual acuity. Endoscopic visualization of the nose with a submucosal purplish mass protruding of the left medium meatus expanding lateral nasal wall. CT enhanced imaging reveals a heterogeneous mass in the left maxillary sinus with an erosive expansion of the lateral nasal wall and orbit floor. Biopsy of the specimen is taken, which is diagnosed as IPEH. The tumor is resected entirely via a Denker endoscopic medial maxillectomy.

**Keywords:** Intravascular Papillary Endothelial Hyperplasia; Masson's Tumor; Denker; Lateral Nasal Wall

## Abbreviation

IPEH: Intravascular Papillary Endothelial Hyperplasia

## Introduction

The nose and paranasal sinuses, as a functional anatomic unit, can be aggravated by distinct histopathologic etiologies defined by the World Health Organization (WHO) as epithelial tumors, soft tissue tumors and tumors of the bone and cartilage [1]. Malignant tumors of the paranasal sinus represent less than 5% of all the tumors of the head and neck, with an incidence of 0.5-1 in 100,000 persons in occidental countries [2].

The Intravascular Papillary Endothelial Hyperplasia (IPEH) also known as Masson's Tumor, described in 1923 by the French pathologist Dr. Claude L. Pierre Masson, is the reactive vascular proliferation secondary to the traumatic vascular stasis with endothelial cellular proliferation. The importance of Masson's tumor, benign in nature, lies in the histopathological and radiological resemblance it has with tumors of malignant nature, e.g. angiosarcoma.

Vascular tumors of the paranasal sinuses are seldom encountered; nonetheless, when they are present, they pose a real challenge for the medical team to identify the benignant or malignant nature of the tumor and to establish the best course of action depending in each individual case.

In this article we present the case of a 41 years old woman with an expansive vascular tumor in the left maxillary sinus, in which we discuss the clinical, radiological and histopathological findings as well as the therapeutic protocol taken.

## Case Report

In April 2019, a 41 years old woman attends the emergency room of the Hospital Regional "General Ignacio Zaragoza" with a history of left hemi facial pain, periorbital edema and loss of visual acuity.

Three months previous to her admittance, she starts with pain in the left zygomatic region and edema in the ipsilateral lower eyelid. She goes with a private medical doctor who make the diagnosis of viral conjunctivitis prescribing topical analgesic and steroid drops, with mild relieve of her symptoms. Seven days previous to her admittance she manifest left eyelid ptosis, diplopia, and hyperalgesia of the left side of the face. She is admitted to the ENT ward on April 2019. She denies exposition to tobacco smoke or chemical aerosols.

Upon her admittance a full otolaryngologic examination is performed. At inspection it is observed swelling of the left zygomatic region and painful left lower eyelid edema; left eye is displaced superiorly to the interpupillary line. Visual acuity: OD 20/70 OS 20/300; left intraocular pressure 30 mmHg.

Endoscopic examination reveals a reddish mass of sub mucous aspect, friable protruding from the left middle meatus with medial displacement of the lateral nasal wall. It is of importance to denote that the patient didn't refer at any time nasal obstruction, rhinorrhea, or epistaxis.

CT scan of the head reveals a tumor occupying the totality of the left maxillary sinus, regular borders, heterogeneous and with great contrast enhancement. An expansive destructive pattern is observed in the anterior and medial wall of the maxillary sinus as well as the floor of the orbit with superior-lateral displacement of the left eye (Figure 1). Magnetic Resonance shows complete occupation of the left maxillary sinus, lateral displacement of the lateral nasal wall and floor of the orbit without compromise to the extraocular muscles (Figure 2). Angiography remarks a highly vascular tumor depending primarily of the terminal branches of the internal maxillary artery (Figure 3).

**Figure 1:** Contrast-enhanced facial CT scan with bone window. A) In the coronal plain there is a heterogeneous contrast-enhanced tumor within the left maxillary sinus which displaces the orbital floor and erodes the lateral nasal wall. B) In the axial plain there is erosion of the anterior and posterior wall of the left maxillary sinus.

**Figure 2:** Magnetic resonance imaging. A) T2-weighted sequence in the axial plain, it is noted a hyperintense heterogeneous tumor in the left maxillary sinus. B) T1-weighted sequence, observe the delimitation of the tumor confined to the left maxillary sinus.

**Figure 3:** Brain angioresonance. A and B) sagittal plain, observe the vascularity characteristic of a vascular tumor well confined to the left maxillary sinus. Note the branching of the internal maxillary artery as the main tumor artery.

A biopsy is taken in the operation room due to the high vascularity, evidenced by the CT scan and MRI, and the high risk of heavy bleeding; the biopsy is taken without any eventuality. Under microscope imaging, a vascular hyperplasic pattern is observed, of soft consistency, bright red, and a fibrous aspect suggestive a tumor of benign nature (Figure 4). A supra-selective embolization with ethylene vinyl alcohol (EVOH) copolymer (ONYX®) is realized to help diminish the tumors size, intra-operative bleeding and hospitalization.

**Figure 4:** Microphotography (H&E 10x). In the superior part of the image, mucosa of the maxillary sinus comprised of vascularized fibro connective tissue, presence of bone fragments without histologic alterations. In the inferior border, partial dilated vessel with the presence of hemorrhage and fronds of thrombotic appearance.

The patient is subjected to a full tumor excision by means of a type III endoscopic maxillectomy or endoscopic Denker procedure (Sturmann-Canfield). During the dissection a friable, necrotic tissue is observed, with expansive destruction of the lateral and anterior nasal wall; despite the lateral displacement of the left eye there was no evidence of orbital fat herniation. A full excision of the tumor is completed, with a mean blood loss of 1000 cc, no blood transfusion was required. Finally, hemostatic material (SurgiCell®) and an anterior nasal packing is placed.

Post-surgical evolution was favorable without systemic manifestations of infection, with improvement of visual acuity and without epistaxis at discharge 14 days after surgery.

Histopathological study of the tumor reveals a vascularized fibro connective tissue, with hemorrhage and thrombotic material forming papillary fronds characteristic of IPEH (Figure 5A). Similarly it is common to observe papillae constituted by fibrin and inflammatory cells; it is important to denote the no zones of mitosis or necrosis that would a malignant tumor dubious was observed (Figure 5B).

**Figure 5:** A) Microphotography (H&E 40 x and 100x). In early stages there is observed thrombotic material forming papillary fronds. B) Microphotography (H&E 100x). Fronds conformed of fibrin, mononuclear and polymorphonuclear cells, enveloped by endothelial cells.

The diagnosis of Intravascular Papillary Endothelial Hyperplasia is confirmed by the service of Pathology. The follow-up of the patient up to 6 months with better visual acuity: OD 20/70 OS: 20/80, PIO: 23 mm/hg, pain-free in the face and without radiological evidence of tumor recurrence.

## Discussion

Vascular tumors of the paranasal sinuses benign or malignant all together are rare, corresponding to hemangiomas as the most frequent vascular tumors [3]. As a group, hypervascular pathologies of the paranasal sinuses present very unspecific clinical signs e.g. unilateral or bilateral nasal obstruction, local facial pain, epistaxis, edema, rhinorrhea or epiphora [4]. Making the diagnosis even more challenging, imaging studies are not enough to determine the nature of the tumor; although we may observe some radiological findings which suggest a malignant nature e.g. erosive destructive pattern, invasion to local tissue o metastasis, it is not enough information to make a confirming diagnosis. As such, it is obligatory to realize an excisional or incisional biopsy conscious of the surgical comorbidities and hospitalization this procedures carry.

The Intravascular Papillary Endothelial Hyperplasia (IPEH) also known as Masson's tumor, was firstly described in 1923 by the French pathologist Dr. Claude L. Pierre Masson in anomalous lesions in the hemorrhoidal plexus in a patient which he described as "hémangioendothéliome végétant intra-vasculaire" [5-7]. Since its firs description it has received multiple eponyms until 1976 Clearkin and Ezinger coined the term "Intravascular Papillary Endothelial Hyperplasia" [8].

The pathogenesis of IPEH is currently considered as a reactive vascular proliferation consequence to the traumatic vascular stasis unleashing the activation of the fibroblastic growth factor (FGF) of the macrophages invading the site of trauma with a proliferation of endothelial cells [6]. There are three clinical types of IPEH: Type I) Primary localized in the dilated vascular spaces; most frequent type. Type II) Secondary or combined, localized in hemangiomas, varicose veins, pyogenic granuloma or arteriovenous malformations. Type III) localized in the extravascular space usually correlated with trauma [9,12].

This type of lesion compose 2% of all vascular tumors [10]; while it can present itself in any part of the body, the most frequent location is in the fingers and the bloody vessels of the head, neck and thorax [4,11]. Pins., *et al*. [22] reported in their literature revision that 56% of tumors corresponded to type I, 40% to type II and 4% to type III; 33% of the cases where localized in the head and neck region with a female to male relation of 1.2:1 [9]. Presentation in the paranasal sinuses is extremely rare. Its clinical manifestation are commonly nasal obstruction, hyaline/purulent nasal discharge and/or epistaxis. Interestingly in the present case none of these classic clinical symptoms of tumors of the paranasal sinuses where present, being visual acuity loss and facial edema the symptoms which motivated to seek medical attention.

Despite its benign nature the expansive pattern observed by the tumor (Figure 1) with erosion of the lateral nasal wall and floor of the orbit with lateral displacement of the eye, mimics the pattern of tumor of malignant etiology.

Stern., *et al.* emphasizes the importance of a differential diagnosis of IPEH especially considering other malignant pathologies like hemangiosarcoma [12]. Some of the histological findings encountered are: a) intravascular development of the tumor, b) lack of mitosis or cellular pleomorphism, and c) no necrosis induced by rapid cellular growth [8]. In early stages it is common to observe thrombotic material and papillary fronds while in late stages one can observe papillary fronds made up of hyaline material and fibroblasts enveloped by endothelial cells that could be confused as hemangiosarcoma (Figure 6). Immuno histologically, Akdur., *et al.* found that 100% of the patients presented positive markers CD 31 and CD 34 alluding to the vascular-thrombotic origin of the tumor; likewise FVIII, type IV collagen, SMA, MSA where positive in varying degrees [11].

**Figure 6:** A) Microphotography (H&E 100x). Fronds presenting hemosiderophages. B) Microphotography (H&E 100x). In late stages the fronds are constituted only of hyaline material and fibroblasts, enveloped by endothelial cells. C) Microphotography (H&E 100x). Delicate papillary fronds constituted only by endothelial cells; this areas are the ones which most get confused with hemangiosarcoma.

In our revision of literature and to the best of our knowledge there has been only 10 cases of IPEH of the nasal cavity and paranasal sinuses reported in the medical literature (Table 1). Interestingly the majority of tumors (60%) appeared in the maxillary sinus, in accordance to the epidemiology of the malignant tumors of the paranasal sinuses [13]. Following the maxillary sinus, the most frequent place of presentation was the ethmoidal sinus (20%) and nasal cavity (20%). It serves to highlight that conservative surgical treatments, without aggressive resection of the tumor, were the ones which presented the highest rate of recurrence; endoscopic or combined (endoscopic and open) techniques were the most satisfactory for total tumor resection.

| Author | Age/Gender | Clinical Symptoms | Location | Surgical Technique | Recurrence |
|---|---|---|---|---|---|
| (1) Stevens 1988 [17] | 21/Male | Nasal obstruction, frontal headache, hypoosmia | Right inferior turbinate. | 1st: septoplasty, marsupialization. 2nd: lateral rhinotomy. | 1st: yes 2nd: no |
| (2) Stern., *et al*. 1991 [13] | 17/Male | Right zygomatic pain, proptosis and headache. | Right maxillary sinus. | Caldwell-Luc Approach. | No |
| (3) Safneck., *et al*. 1995 [16] | 36/Female | Right nasal obstruction. | Right inferior turbinate and nasal cavity. | 1st: trans-nasal excision. 2nd: Sublabial transmaxilar excision. | 1st: yes 2nd: no |
| (4) Lancaster., *et al*. 1998 [22] | 67/Female | Nasal obstruction, anterior and posterior rhinorrhea. | Left maxillary and ethmoidal sinus. | Endoscopic surgery. | No |
| (5) Moon., *et al*. 2000 [5] | 35/Male | Visual acuity loss, ptosis, proptosis and headache. | Left ethmoidal and sphenoid sinus and sella turcica. | Weber-Fergusson approach. | No |
| (6) Hooda., *et al*. 2007 [23] | 45/Female | Epistaxis | Right ethmoidal sinus. | Denker's medial maxillectomy. | No |
| (7) Lombardi., *et al*. 2008 [20] | 26/Maale | Left nasal obstruction, purulent rhinorrhea, epiphora and ocular pain. | Left maxillary and ethmoidal sinus. Left orbit and anterior cranial fossa. | Mixed approach: Endoscopic nasal surgery and sub frontal craniotomy. | No |
| (8) Wang., *et al*. 2009 [21] | 42/Male | Left nasal obstruction, rhinorrhea, epistaxis and headache. | Left maxillary, ethmoidal and frontal sinus, left nasal cavity. | Endoscopic surgery. | No |
| (9) Al Qahtani KH 2016 [18] | 33/Female | Nasal obstruction, epistaxis and rhinorrhea. | Right nasal cavity, right maxillary, ethmoidal, frontal and sphenoid sinus. | CT-guided endoscopic surgery. | No |
| (10) D´Aguanno., *et al*. 2018 [19] | 67/Female | Right nasal obstruction, right zygomatic pain and rhinorrhea. | Right maxillary and ethmoidal sinus. Right nasal cavity. | Mixed approach: open and endoscopic surgery. | No |
| (11) Present case | 41/Female | Visual acquity loss, left hemi facial pain and periorbital edema. | Left maxillary sinus. | Denker's medial endoscopic maxillectomy. | No |

**Table 1:** Case reports of Intravascular Papillary Endothelial Hyperplasia in medical literature.

Masson's tumor, being a benign vascular neoplasia, it is important a complete surgical resection to avoid tumor recurrence which have been documented [14,15]. With the introduction of the endoscope to the otolaryngology field the possibility of resecting tumors from the nasal cavity and paranasal sinuses from within the nose has favored its usage displacing more aggressive or morbid open technique surgeries, e.g. lateral rhinotomy, Weber-Ferguson technique or open maxillectomy. In our patient an endoscopic Denker's technique was chosen, with which ample knowledge and experience we have with non-papilloma tumors of the paranasal sinus, since it offers a great visual and surgical field for the resection of suck pathologies.

## Conclusion

Masson's tumor is an unusual lesion consisting of an organized thrombus in a venous vein generally without history of trauma that can be associated with preexisting benign vascular lesion; of slow growth and with expansive destruction of the anatomical area in which it is located. Imaging studies and direct visualization of the tumor are essential complimentary studies for a good surgical planning. Nonetheless, it is the invariably the expertise of the anatomical pathology doctor which will determine the definitive diagnosis. IPEH has an excellent prognosis given it is resected in its totality. Surgical management will depend on the individual conditions of each case as well as the experience of the surgeon with open or endoscopic techniques. This article advocates the use of endoscopic techniques, especially Denker's endoscopic technique, for the excision of nasal cavity and paranasal sinus tumors for its great versatility and efficiency; under the right selected cases and in hands of an experienced surgeon, comorbidities are kept at minimum as well as hospitalization days and the aesthetic defects of an open approach.

## Conflict of Interest

The authors declare no conflicts of interest.

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