Giant Hypervascular Lesion of the Sinonasal Tract Invading the Anterior Skull Base and Orbit: A Puzzling Case

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The sinonasal tract may be involved in a wide variety of hypervascular lesions, either benign or malignant, and differential diagnosis may be challenging. We present the case of a 26-year-old man with an aggressive ethmoid hypervascular mass invading the anterior skull base and orbit. Because of massive intraoperative bleeding and difficult dissection of the lesion from the periorbita, the planned craniendoscopic approach had to be converted into a standard craniofacial resection by a combined Lynch and coronal incision. The definitive diagnosis was consistent with lobular capillary hemangioma associated with intravascular papillary endothelial hyperplasia. Two years after surgery, the patient is free of disease. Lobular capillary hemangioma is a hypervascular lesion that may involve the sinonasal tract. The case presented herein is exceptional, both in the presentation and in the difficulties encountered in diagnosis and treatment, because of the concurrence of lobular capillary hemangioma and intravascular papillary endothelial hyperplasia.

Key Words: angiosarcoma, intravascular papillary endothelial hyperplasia, lobular capillary hemangioma, Masson's tumor, sinonasal tract.

INTRODUCTION

The sinonasal tract may be involved in a wide variety of hypervascular benign lesions (eg, juvenile angiofibroma, hemangioma, hemangiopericytoma, hemangioendothelioma, lobular capillary hemangioma [LCH], paragangioma) and malignant lesions (eg, angiosarcoma, Kaposi’s sarcoma, olfactory neuroblastoma, Merkel cell carcinoma, metastasis from kidney adenocarcinoma). Their endoscopic appearance is usually nonspecific, and in only a few of them, such as juvenile angiofibroma, do epidemiological data and imaging findings lead to a correct diagnosis without resort to biopsy, which is associated with a risk of massive hemorrhage. Furthermore, analysis of biopsy samples may frequently only suggest a diagnosis, which is definitely established after careful examination of the entire specimen.

Lobular capillary hemangioma commonly involves the nasal cavity as a small hypervascular lesion arising from the anterior third of the septum or the lateral wall. Its diagnosis is challenging in those rare instances of a large LCH that entirely fills the nasal cavity, which can be misdiagnosed as angiofibroma or low-grade angiosarcoma.

We report an intriguing case of LCH associated with intravascular papillary endothelial hyperplasia (IPEH) presenting as a huge expansile lesion involving the anterior skull base and orbit. Problems related to diagnosis and selection of the surgical approach will be discussed in detail.

CASE REPORT

In November 2004, a 26-year-old man began complaining of left nasal fossa obstruction with purulent nasal discharge, ipsilateral epiphora, and orbital pain. A diagnosis of nasal polyposis was made elsewhere, but no treatment was advised. Three months later, the patient was admitted to another hospital for fever, exacerbation of pain, and left periorbital swelling. An orbital complication from acute rhinosinusitis was suspected, and a computed tomography (CT) scan was obtained. The examination revealed a left nasoethmoidal mass with soft tissue density and strong enhancement after administration of contrast medium (Fig 1). Bone remodeling and possible invasion of both the anterior skull base and the left lamina papyracea were also detected. The left orbital contents were laterally displaced. At this point, the patient was referred to our department.

Upon endoscopy, the left nasal fossa was completely filled by a red, easily bleeding mass, apparently originating from the ethmoid sinus. Magnetic resonance imaging (MRI) scans (Fig 2) showed...
a left nasoethmoidal, solid neoplasm characterized by a nonhomogeneous signal and relevant enhancement after intravenous injection of gadolinium. The lesion displaced the left lamina papyracea; intraorbital extension was also suspected in relation to the encroachment on the rectus medialis muscle. The left nasolacrimal duct was compressed, with consequent dilation of the lacrimal sac. Also, the ethmoid roof was resorbed, with clear signs of cribriform plate infiltration; dural thickening was suspicious for involvement. Angiography was performed to analyze the vascularization pattern of the mass and to verify the possibility of preoperative selective embolization. The lesion was vascularized mainly by the internal carotid artery through the ethmoidal arteries, whereas the supply from the internal maxillary and sphenopalatine arteries was minimal. Embolization of the major feeding vessels was not performed, in view of the high risk of complications. A biopsy specimen was obtained under local anesthesia. Microscopically, a vascular lesion was seen, mainly characterized by capillary proliferation with focal endothelial papillary hyperplasia, but without elements suggestive for malignancy. A cranioendoscopic resection combining a transnasal endoscopic approach with a subfrontal craniotomy was offered to the patient, who was informed of the possibility of switching to an external transfacial procedure. During dissection of the mass from the periorbita, massive bleeding from the ethmoidal arteries was encountered, necessitating their quick exposure and coagulation through a Lynch incision. Although the lesion was intimately adherent to the periorbita, which was partially resected, there were no signs of involvement of the orbital contents. A similar relationship with the dura led us to perform an extensive resection and consequently a duraplasty by use of 2 layers of fascia lata and a third layer of pedicled pericranium. Transfusion of 2 prebanked blood units was required. The postoperative course was

**Fig 1.** Multislice computed tomography in A) coronal and B) axial planes. Lesion is demonstrated in left ethmoid sinus, causing extensive bone destruction at level of lamina papyracea, nasal septum, and ethmoid roof.

**Fig 2.** Magnetic resonance imaging. A) Gadolinium–diethylenetriamine pentaacetic acid (Gd-DTPA)–enhanced 3-dimensional gradient echo (GE) T1-weighted imaging with fat suppression in axial plane. B) Spin echo (SE) T2-weighted imaging in coronal plane. C) Gd-DTPA–enhanced SE T1-weighted imaging in sagittal plane. Left naso-ethmoid mass is visible, exhibiting vivid contrast enhancement and marked bone remodeling at level of nasal septum, lamina papyracea, and ethmoid roof. Effacement of extraconal fat pad and close contact between lesion and rectus medialis muscle suggest orbital infiltration. Cribriform plate is encroached on by tumor, and adjacent dura is thickened and enhancing.
uneventful, and the patient was discharged 10 days after surgery.

Histologic examination showed the lesion to be composed of papillary structures often fused to form a network of anastomosing vascular channels of various calibers. The papillary structures were composed of a single layer of endothelial cells, and the cores contained red cells or fibrinoid material (Fig 3). At the periphery of the tumor, a striking lobular pattern was present; larger-caliber vessels, often with a muscular wall, surrounded by congeries of small capillaries were also observed. No necrosis, no significant pleomorphism, and no abnormal mitoses in the endothelial cells were present. A large amount of thrombotic and hemorrhagic material was also present. Immunohistochemically, the endothelial cells of the papillae reacted with endothelial cell markers (factor VIII–related antigen, CD31, and CD34), and there was an abundance of actin-positive pericytes surrounding the vascular channels (Fig 4).

A diagnosis of a benign vascular lesion (not otherwise specified) associated with IPEH was made; a consulting pathologist confirmed the benign nature of the lesion, defining it as IPEH in LCH.

Two years after treatment, the patient has no radiologic or endoscopic evidence of recurrence (Fig 5).

DISCUSSION

Lobular capillary hemangioma was first described as “human botryomycosis” by Poncet and
Dor\textsuperscript{2} in 1897, and subsequently it has been commonly referred to as “pyogenic granuloma,” a term first introduced by Hartzell\textsuperscript{3} in 1904. Since the term “granuloma” could be misleading, erroneously suggesting an infectious or a granulomatous process, in 1980 Mills et al\textsuperscript{4} proposed the more accurate term “lobular capillary hemangioma” to highlight the main histologic features of the lesion, which is characterized by aggregates of capillaries arranged in lobules and embedded in an edematous and fibroblastic stroma. The skin, especially on contact surfaces, and the oral cavity mucosa are the most commonly involved sites, whereas its occurrence in the nasal cavity is quite rare. Because of the rarity of the lesion, no data on prevalence have been reported. A male predominance in patients younger than 18 years and a female predominance in patients in the 18- to 39-year age range have, however, been observed.\textsuperscript{4} Any gender predilection is no longer present after the age of 39 years.\textsuperscript{4} The cause of LCH is still unknown, even though direct trauma, hormonal influences, underlying microscopic arteriovenous malformations, viral oncogenes, and the production of angiogenic growth factors have all been hypothesized to be related to its pathogenesis. A specific form of LCH, the so-called “pregnancy tumor” typically observed in the oral or nasal cavity of pregnant women, is believed to be related to altered estrogen and progesterone stimulation.

Lobular capillary hemangioma of the nasal cavity most frequently arises from the anterior part of the nasal septum, thus supporting the effect of traumas, such as nose picking and nasal packing, in the development of the lesion. The most frequent presenting complaint is epistaxis, and nasal obstruction, facial pain, and impairment of olfaction are rarely reported. Its endoscopic appearance is usually quite typical, showing a small, red, easily bleeding mass involving the anterior third of the nasal cavity. Lobular capillary hemangioma usually has rapid growth and a marked tendency to bleed, especially when traumatized; it does not invade the surrounding structures, but may cause bone resorption.\textsuperscript{7}

Only 4 cases of huge sinonasal LCH, defined as “giant LCH,” have been reported in the literature.\textsuperscript{8-11} The lesion was secondary to nasal packing\textsuperscript{8,9} and to long-lasting nasal intubation\textsuperscript{11} in 2 cases and 1 case, respectively, and the remaining case was a pregnancy tumor.\textsuperscript{10} The site of origin was the inferior turbinate in 3 cases\textsuperscript{8-10} and the nasal septum in 1 case.\textsuperscript{11} Three cases were managed by endoscopic surgery,\textsuperscript{8,9,11} but the lesion in the pregnant patient required superselective embolization and lateral rhinotomy.\textsuperscript{10}

The present case is, to the best of our knowledge, the first “idiopathic” giant LCH reported in the literature. Furthermore, although the exact site of origin was not recognizable, the epicenter of the lesion was apparently in the anterior ethmoid sinus. Another finding that makes this case unique is the association with IPEH.

Intravascular papillary endothelial hyperplasia was first described in 1923 by Masson\textsuperscript{12} as a neoplastic process consisting of papillary hyperplasia of endothelial cells, with a consequent obliteration of the vascular lumen, later followed by degenerative changes for which he coined the term “vegetant intravascular hemangioendothelioma.” Currently, IPEH is no longer interpreted as a neoplasm, but instead as a reparative response of endothelial cells to inflammation and stasis within the vascular bed leading to an unusual form of thrombus organization.\textsuperscript{13,14} Intravascular papillary endothelial hyperplasia has been divided into 3 forms: a primary form that occurs within dilated vascular spaces; a secondary form that usually arises in a preexist-
ing vascular lesion; and a third form (extravascular) that arises in a hematoma. Intravascular papillary endothelial hyperplasia is rare, with a wide age distribution (9 months to 80 years) and a slight female predominance. It may occur in different sites, but it is most frequent in the dermis and subcutis of the head and neck, fingers, and trunk. From a survey of the literature, we identified only 4 cases of IPEH involving the paranasal sinuses. The first case was diagnosed in a 17-year-old boy; on CT it was found to originate within the right maxillary sinus and to involve the floor of the orbit and the ipsilateral ethmoid sinus. The lesion was excised via a Caldwell-Luc approach; at definitive histology a diagnosis of IPEH arising in a preexisting vascular malformation was made. The second case, a right maxillary sinus lesion extending into the ethmoid sinus, was observed in a 67-year-old woman. The tumor was removed endoscopically, and the histologic diagnosis was consistent with primary IPEH. In the third case, an ethmoidal-sphenoid mass extending to the sella and cavernous sinus was diagnosed in a 35-year-old man. A transfacial excision through a Weber-Fergusonus approach was performed. In this case the definitive diagnosis was also IPEH within a preexisting vascular lesion. Recently, Hooda et al reported the case of an ethmoid lesion in a 45-year-old woman with recurrent brisk epistaxis. Since neither intraorbital nor intracranial extension had been detected by imaging studies, the patient underwent Denker’s medial maxillectomy. The definitive histologic diagnosis was consistent with primary IPEH, even though an underlying hemangioma could not be ruled out. Two cases of nasal cavity IPEH, both involving the inferior turbinate, have also been reported. In the first case the lesion was removed by a lateral rhinotomy incision, whereas in the second patient complete surgical excision was obtained by a sublabial transmaxillary approach. These cases involving the paranasal sinuses demonstrate that in this site the lesion can display a locally aggressive pattern of growth with clinical and radiologic features mimicking a malignant tumor.

In dealing with a hypervascular lesion of the sinonasal tract or nasopharynx, differential diagnosis is essential to properly plan treatment. The imaging profile is rarely diagnostic, as in juvenile angiofibroma; in all the other masses some common features, including strong enhancement after contrast medium administration, are usually present. The tendency to erode the bone may be associated not only with malignancies, but also with aggressive benign lesions. Therefore, even in the present case, CT and MRI findings of a hypervascular lesion associated with extensive bony erosion of the anterior skull base and lamina papyracea could not lead to definition of the nature (benign versus malignant) of the lesion. On histopathology, the most critical differentiation was from a low-grade angiosarcoma. Primary angiosarcomas of the sinonasal tract are very rare, and in this specific site they tend to present in the low-grade form. Unlike benign vascular lesions, angiosarcoma is characterized by the presence of interconnecting vascular channels dissecting the underlying stroma. Neoplastic vessels are lined with atypical endothelial cells, arranged in papillary structures, with “hobnail” nuclei and epithelial tufts. These papillary structures may resemble those of IPEH, but they are associated with a more pronounced piling up of cells and more nuclear atypia. Furthermore, unlike LCH, angiosarcoma lacks a lobular architecture. Solid areas, necrosis, and hemorrhage may be prominent in high-grade tumors. At histology, several elements led us to exclude a diagnosis of angiosarcoma. First, there was the lack of tumor-related necrosis and atypical mitoses and the presence of only occasional nuclear pleomorphism. Second, a lobular arrangement in some areas of the tumor contrasted with the manner in which well-differentiated angiosarcoma dissects through connective tissue and creates irregular vascular spaces.

Finally, the presence of pericytes around vascular channels and the expansive pattern of growth without infiltration of the brain and soft tissues of the orbit was another element that excluded a diagnosis of angiosarcoma. The biologic aggressiveness coupled with the imaging and pathologic features of the present case excluded other vascular lesions (angiofibroma, hemangioma) from the differential diagnosis.

Our experience and reports of other groups confirm the possibility of removing hypervascular lesions of the sinonasal tract and nasopharynx with an endoscopic approach. A purely endoscopic approach should be considered as a viable alternative to external approaches, not only for small lesions, but also for larger tumors extending into the infratemporal fossa, the apex of the orbit, and the parasellar area. Extensive dural adherences, abundant vascular supply from the internal carotid artery, and encasement of the artery itself by the tumor lead to our department to selection of a combined or external approach.

In the present case, however, the imaging findings clearly showed intracranial extension and encroachment on the medial rectus muscle, together with a pattern of vascularization coming mainly from the internal carotid system through the ethmoidal arteries, which led us to plan a combined cranio-end-
scopic approach. However, massive bleeding from the ethmoidal vascularization and the firm adherences of the mass to the periorbita required prompt identification and coagulation of both ethmoidal arteries via a Lynch incision, first to control the bleeding and then to clearly identify the relationship between the mass and the orbital contents. The lesion was also found to be strongly adherent to the dura, which was excised and patched with fascia lata and vascularized pericranium. After definitive diagnosis confirming the benign nature of the lesion, the need for adjuvant radiotherapy was excluded, and the patient was strictly followed up with periodic endoscopic and MRI evaluations. Both LCH and IPEH, when radically excised, have indeed a favorable prognosis with a very low rate of local recurrence.1,13,14

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REFERENCES


CONCLUSIONS

This is the first report of a hypervascular mass of the paranasal sinuses with the coexistence of an LCH with IPEH, two disorders that are rarely encountered even independently in this anatomic area. The mass presented as a large expansile growth eroding the anterior skull base and the lamina papyracea, thus suggesting an aggressive behavior. Even the rare cases of giant LCH reported in the literature were never associated with a locally aggressive pattern of growth, which seems to be more characteristic of IPEH. It is possible that the association of these two different disorders might explain the local aggressiveness of the lesion, which was challenging to diagnose and treat.