

AN EXCEPTIONAL NASAL TUMOR: ANGIOLEIOMYOMA OF THE NASAL CAVITY

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ABSTRACT:

Angioleiomyoma is a benign tumor that arises from smooth muscle. It is rare in the head and neck area and less than 1% occur in the nasal cavity with 38 cases reported so far. The only way to make the diagnosis is the histologic examination. The treatment of choice is surgical excision and it is curative. Occurrence of this type of neoplasm in the nasal cavity is extremely exceptional and has rarely been described in the literature. We present a new case of angioleiomyoma in the nasal cavity and a review of the literature

KEYWORDS- Angioleiomyoma, nasal cavity, management

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INTRODUCTION:

Benign sinonasal tract masses, either neoplastic or non-neoplastic are commonly encountered in the clinical practice. Angioleiomyoma is a benign tumor arising from smooth muscle cells and it is also known as vascular leiomyoma. They are rarely found in the head and neck area. Maesaka et al [1-3] reported the first case of angioleiomyoma of the nasal vestibule in 1966. Since then, to our knowledge, only 38 cases of nasal cavity angioleiomyomas have been reported [2] This rare tumor shows prevalence in female patients (in a 2:1 ratio) between the fourth and sixth

decades of life and can be originated mainly the inferior nasal conchae. The clinical features of this tumor include: nasal obstruction, facial pain, headache and epistaxis. Computed tomography (CT) and magnetic resonance do not conclude the diagnosis. Cytological examination does not provide a conclusive diagnosis. Surgical excision with histologic examination is the only way to make a definitive diagnosis [2]. Recurrence is extremely rare after total excision.

CASE REPORT:

We present a case of a 49 year old male patient with a 12 month history of painless left nasal obstruction and recurrent epistaxis. He had a personal clinical history of an orbital exenteration when he was 2 years old due to an orbital tumor of unknown details. Physical examination revealed an elastic, brownish mass occupying the back of his left nasal cavity [Figure 1a]. Computerized Tomography scan (CT scan) of paranasal sinuses showed a well circumscribed, multilobulated soft tissue density mass originating from the left middle turbinate that strongly enhanced with intravenous contrast. The radiological suspicion was of a hemangioma. [Figure 1b]

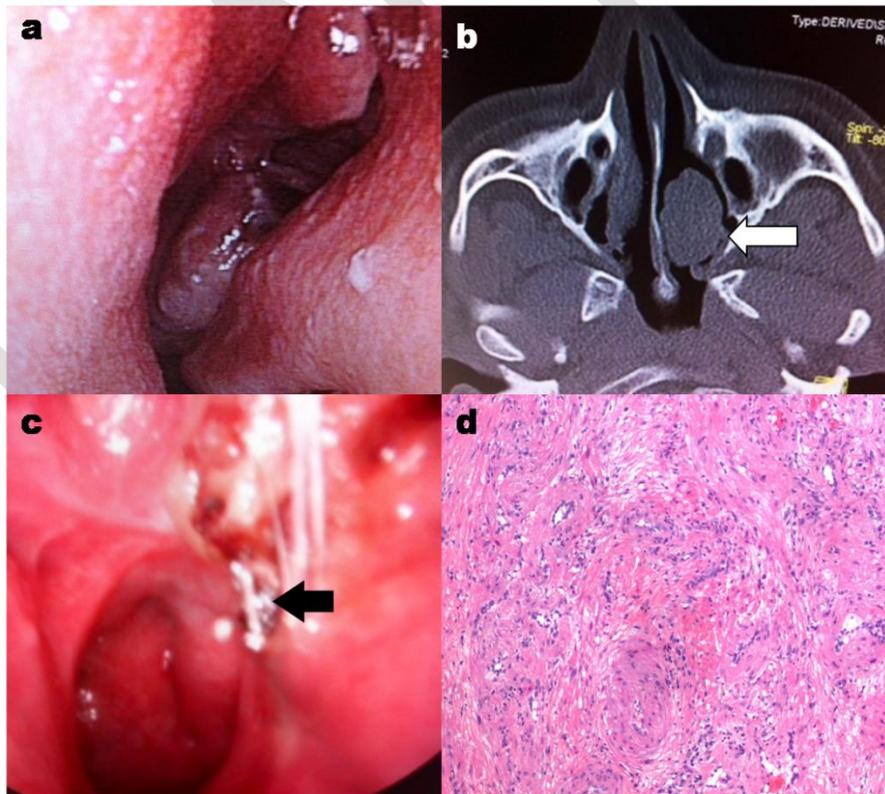


Figure 1.- a.- Nasal endoscopic view showing a brownish mass occupying the back of the left nasal cavity; b.- CT scan in an axial section revealing a well circumscribed, multilobulated soft tissue density mass (white

arrow) originating from the left middle turbinate; c.- Endoscopic view of the resection area without tumor.
d.- Pathologic examination shows blood vessels with thick muscular walls interposed with bundles of smooth muscle (H&E 150 x)

He was consequently scheduled for a surgical resection with a previous tumoral embolization. A complete resection of the mass through an endoscopic approach was performed [Figure 1c]. During the operation there was minimal bleeding. Postoperative evolution was satisfactory. Conclusive anatomopathological results confirmed a multivascular tumor with numerous smooth muscle layers, scarce fibrosis and fatty tissue [Figure 1d]. Intact mucosal surface surrounded the mass. Immunohistochemistry revealed positive Desmin and Smooth Muscle Actin-SMA (Desmin+/ alpha-SMA+) as well as CD34 positive and S100 negative markers suggesting an Angioleiomyoma of the left middle turbinate. No evidence of recurrence has been noticed in a 3 year follow-up.

DISCUSSION:

Angioleiomyoma is an unusual type of benign neoplasm of slow growth. A female prevalence, between the 4th and the 6th decade has been demonstrated in the literature [1, 3]. The most common sites in which they might be found are the uterus (95%), skin (3%), and gastrointestinal tract (1.5%). [1, 3, 4] The sinonasal tract is an exceptionally rare location for this type of neoplasm, being in these cases the inferior conchae mainly affected. Other possible locations involve the paranasal sinuses, the nasal septum, middle and upper turbinates, the nasal floor, lateral nasal wall and the vestibule [3]. One of the biggest case series (562 cases) was reported by Hachisuga et al, finding only 48 cases (8%) of angioleiomyoma in the head and neck region, only 5 of them being in the nasal cavity. Angioleiomyomas of the head and neck account for 9.5–12.5 % of this type of tumors and only 1 % are found in the sinonasal tract. Intracranial angioleiomyomas have also been described usually located on the extraneural axis, the sella, posterior fossa, and the skull. [5, 6, 7]

There are three hypotheses about the origin of smooth muscle tumors in the nasal cavity, which are: aberrant undifferentiated mesenchyma, smooth muscle elements in blood vessel walls and piloerector muscles or a combination of both. The scantiness of smooth muscle in the nasal cavity might explain the rareness of these tumors. [3, 6] Some authors have described the possible influence of sexual hormones and of an Epstein-Barr virus infection. [3, 7] Marioni et al and Tseng et al [3, 7, 8] suggested the tumor growth might be hormone-dependant due to a female predominance, immunopositivity for progesterone receptors and negativity for estrogen receptors and the increased pain during pregnancy or the menstrual cycle as well as the predominant symptom of nasal bleeding in these patients. This hypothesis needs further clarifying. [3, 7, 8]

Macroscopically they are usually described as a painless brownish, solid sessile or polypoid well circumscribed but non-encapsulated elastic mass, such as the one found in our patient, causing symptoms such as epistaxis, headache, nasal discharge and unilateral nasal obstruction. [3, 4, 6] If a cutaneous component is involved, local and facial pain might be described. Differential diagnosis must be made with other nasal tumors such as: nasal angiofibroma, hemangioma, inverted papilloma, malignant lymphoma, fibromyoma, leiomyoblastoma, hemangiopericytoma, angiosarcoma among others. [6]

Radiologic and cytologic findings are inconclusive and doubtful [1, 3] The CT scan and magnetic resonance may help correlate anatomical structures and planes, surgical margins, bony erosions, and characterize the intranasal mass. [2, 6] This leads to a necessary complete surgical excision for a definite diagnosis. The surgical approach depends on the size, location, the extension of tumor and the surgeon's experience. The endoscopic approach in most cases, like in our case, can be performed successfully due to most of the lesions being limited to the sinonasal cavity. [3, 4] Preoperative selective embolization should be considered for large hypervascular lesions or if located high among the nasal cavity. [9] Depending on the extension and location of the lesion laser or a transpalatine approach, Caldwell-Luc, lateral rhinotomy, external ethmoidotomy with medial maxillectomy, open rhinoplasty or a craniofacial resection may be required. [1, 3, 6, 10-12].

The World Health Organization classified leiomyomas in three groups: leiomyoma (non-vascular), angioleiomyoma (vascular leiomyoma), and epithelioid leiomyoma [2, 6]. On the other hand, they can also be classified as being submucosal or soft tissue/cutaneous. A possible fatty component has been described among angioleiomyomas from all sites (2.8 % of cases). Angioleiomyoma with adipocytic differentiation is found with a higher frequency in sinonasal submucosa, affect males with a higher prevalence and occur at a higher age [2].

The angioleiomyoma is characterized by a well defined proliferation of mesenchymal tapered cells with eosinophilic cytoplasm and elongated basophilic nuclei that show tapered endings (cigar-like shape nuclei). The vascular spaces which are lined by a single layer of endothelial cells are constant features in angioleiomyoma. [3, 4, 6] Immunohistochemical staining such as Smooth Muscle Actin, Desmin, Myoglobin, S-100 protein, and Vimentin can be of additional value. [3, 5, 6]

The prognosis of sinonasal leiomyoma is favorable. No malignant transformation has been described and recurrence is extremely rare if complete resection has been performed.

CONCLUSION:

An Angioleiomyoma of the nasal cavity is a rare benign tumor. Clinical and radiological findings are inconclusive for diagnosis but may provide important information about the mass and surrounding structures. The only way to make the diagnosis is surgical excision with histologic examination. Complete surgical excision is the treatment of choice and it is curative. It is important to know this type of tumors to distinguish them from other potentially recurring or locally aggressive neoplasms which may require different therapeutically approaches.

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