

Angioleiomyoma of Nasal Septum: Case Report and Literature Review

Case Report

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Abstract

Introduction: Angioleiomyoma is a benign soft tissue tumor of smooth muscle origin with a vascular component and is an uncommon form of leiomyoma. Angioleiomyoma presenting in the nasal cavity is exceedingly rare and there are 68 reported cases in the literature worldwide. We present a case of angioleiomyoma of the nasal septum and review its diagnosis and treatment.

Study Design: Case report and Literature Review.

Methods: The medical records of a 69-year-old patient with an angioleiomyoma of the nasal septum were reviewed. The PubMed database was searched for literature describing angioleiomyoma of the nasal cavity using the key words “angioleiomyoma” with “nasal cavity,” “nasal septum,” “nose,” or “sinus.”

Results: A 69-year-old female patient presented with progressive right-sided nasal obstruction and epistaxis. Office examination revealed stigmata of recent bleeding and nasal endoscopy revealed a mass arising from the right nasal septum. Computerized tomography with intravenous contrast revealed a 1.3 x 1.1 cm heterogeneously enhancing vascular lesion arising from the right nasal septum. The patient was taken to the operating room for endoscopic resection.

Conclusion: Angioleiomyoma of the nasal septum is a rare and challenging clinical diagnosis that requires detailed histopathologic examination. Literature review suggests a female predilection with possible hormonal influence.

Keywords: Rhinology; Angioleiomyoma; Nose Neoplasms.

Introduction

Angioleiomyoma is a benign soft tissue tumor of smooth muscle origin with a vascular component and is a rare form of leiomyoma. Like other sinonasal mesenchymal tumors, angioleiomyomas are uncommon in the nasal cavity and there are 68 reported cases in the literature worldwide [1-22]. Angioleiomyoma presenting on the nasal cavity is exceedingly rare and the first reported case of sinonasal angioleiomyoma was by Maesaka in 1966 [2]. Literature review reveals 16 reported cases on the nasal septum [1, 4-8, 19, 20, 22-25]. We present a new case of angioleiomyoma of the nasal septum, review its diagnosis, clinical and pathologic characteristics, and management.

Case Presentation

A 69-year-old African American female was referred to our

Otolaryngology clinic with the chief complaints of progressive right-sided nasal obstruction and epistaxis for one year in duration. She had a history of allergic rhinitis and also endorsed sneezing and clear rhinorrhea. The patient is post-menopausal and has a past medical history of hypertension, hyperlipidemia, chronic gastritis, multiple renal cysts, renal artery aneurysm, and hyperaldosteronism. She denied tobacco use. She has no history of head or nasal trauma, sinonasal surgery, or history of cancer.

Upon physical examination, anterior rhinoscopy revealed crusting and stigmata of recent bleeding and nasal endoscopy revealed a smooth, pink, vascular appearing mass arising from the right nasal septum was visualized. The mass was touching the right lateral wall and completely filled the superior aspect of the right nasal vestibule (See Figure 1). A computerized tomography scan with intravenous contrast revealed a 1.3 x 1.1 cm heterogeneously enhancing vascular lesion within the right anterior nasal cavity

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that appeared to arise from the nasal septum (Figure 2). Given the benign appearance of the lesion, further imaging was not obtained.

The patient was taken to the operating room for endoscopic resection. The mass was observed to extend to the superior aspect of the internal nasal valve without involvement of the upper lateral nasal cartilage. The mass was removed en bloc including a wide cuff of normal mucosa circumferentially around the mass before dissecting the mass from the nasal septal cartilage, preserving perichondrium, without disrupting the nasal valve, upper lateral cartilage, or right nasal ala. No septal perforation was noted

intraoperatively or at 3 months post operatively. The specimen was sent for permanent pathology. The gross specimen was 3.0 x 2.0 x 1.2 cm in size and tan and pink in appearance with a smooth and glistening surface. H&E stain revealed a well-circumscribed tumor with proliferation of fascicles of smooth muscle cells and surrounding thick-walled vessels with narrow lumens (Figure 3). The endothelial cells lacked pleomorphism, mitosis, nuclear atypia, or necrosis. Immunohistochemistry was positive for SMA, which revealed the smooth muscle cell component, as well as ERG1 (nuclear stain), which highlighted the endothelial cells lining narrow vessel lumens (Figure 4).

Figure 1. Endoscopic image of the patient's lesion on the right nasal septum.

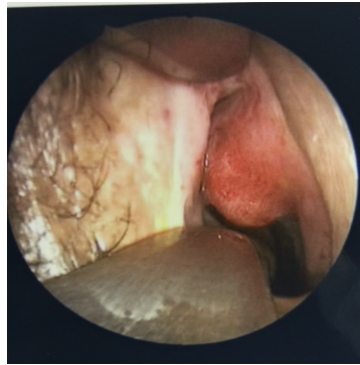


Figure 2. Computerized Tomography images in the coronal plane depicting a nasal cavity mass arising from the right nasal septum.

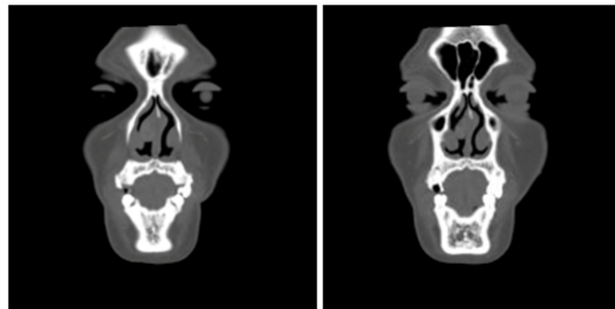


Figure 3. Medium power view (H&E stain) demonstrating the well circumscribed smooth muscle cell proliferation with thick vessel walls (arrows) and narrow lumens.

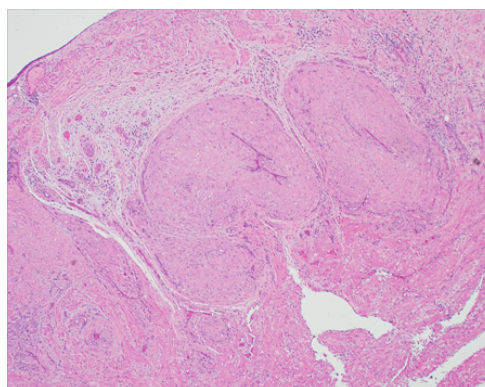
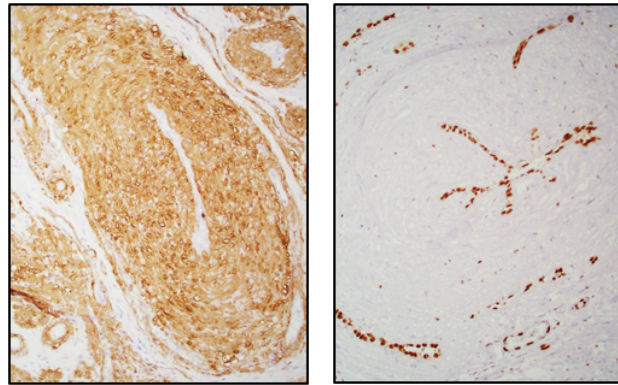


Figure 4. Immunohistochemistry for smooth muscle actin (SMA) showed diffuse immunoreactivity in the smooth muscle cells comprising thick blood vessel walls (Left). Immunohistochemistry for ERG1 (nuclear stain) highlighted the endothelial cells lining the narrow vessel lumens (Right).



Discussion

Sinonasal angioleiomyomas make up less than 1% of leiomyomas in the body [5, 10]. Roughly 10% of these tumors are located in the head and neck region and presenting sites include the nose, auricle, lips, and neck [1, 21]. This slow growing, benign tumor may present as a solitary mass that may or may not be painful. Other typical presenting symptoms may include nasal obstruction, epistaxis, facial pain, recurrent sinusitis, and can be associated with a septal deviation [1]. Angioleiomyoma is not often included in the clinician's differential diagnosis of an obstructive nasal mass. The differential diagnosis includes both benign and malignant tumors of the nasal cavity including inverted papilloma, nasal angiofibroma, hemangioma, hemangiopericytoma, solitary fibrous tumor, desmoid fibromatosis, peripheral nerve sheath tumors, and sinonasal sarcomas including leiomyosarcoma and rhabdomyosarcoma [1, 22].

The rare nature of these tumors is attributed to the paucity of smooth muscle tissue in the region. Smooth muscle in the nasal cavity is present in blood vessel walls as well as the piloreceptor muscles of the anterior vestibule [26]. Angioleiomyoma of the nasal cavity appears to present more commonly on the turbinates and nasal vestibules presumably due to the presence of smooth muscle. The nasal septum is understandably an exceedingly rare location for this tumor.

The World Health Organization classifies leiomyomas into three histologic groups: leiomyoma, angioleiomyoma, and epithelioid leiomyoma [10, 26, 27]. Our patient's specimen was classified as angioleiomyoma by histopathologic examination. The vascular component of this smooth muscle derived tumor may also be classified as solid, venous, or cavernous type based on histological findings [1, 27]; this was not specified in our patient. Immunohistology will reveal expression of alpha smooth muscle actin (SMA) and possibly h-caldesmon, muscle specific actin, desmin, S100, or myoglobin [1, 3].

Literature review reveals that the most common age range for presentation is between 40 and 60 years of age [3, 28, 29]. A female predilection has been suggested in the literature, with reports of a female:male ratio of 3.75:1 [3, 21, 29]. Marioni et al., suggest possible hormone dependence and report a case of a progesterone dependent sinonasal angioleiomyoma [29]. Heet al

reported a case of nasal cavity angioleiomyoma that was partially immunoreactive to progesterone receptor [16]. This phenomenon may account for the female preponderance of reported cases [29]. Other risk factors, prognostic factors, and mortality data has yet to be elucidated given the rarity of these lesions.

The treatment of choice appears to be local excision, which may either be performed transnasally or endoscopically with macroscopic margins or by open approach; pre-operative embolization may be considered for large tumors with abundant vascular supply. KTP laser has been used for bloodless ablation [3, 22]. A lateral rhinotomy approach has been described for a large tumor [24]. One case of a particularly extensive tumor in the nasal cavity required craniofacial resection [12].

Our literature review reveals 16 total cases of angioleiomyoma arising from the nasal septum [1, 4-8, 19, 20, 22-25]. This represents 23.5% of angioleiomyomas of the sinonasal cavities reported worldwide. In the English literature, 8 nasal septum angioleiomyomas were reportedly resected transnasally or endoscopically while 1 tumor was resected using an open (lateral rhinotomy) approach. The remainder of reports did not specify the surgical approach. There were no reports of patients requiring reconstruction or a second stage procedure. Recurrent sinonasal angioleiomyoma has been reported on the middle turbinate [3], however, no recurrent lesions have been reported on the nasal septum. Malignant transformation has yet to be reported in the literature for sinonasal angioleiomyomas. The current patient is doing well 3 months post operatively.

Conclusion

Angioleiomyoma of the nasal septum is an exceedingly rare tumor and is a challenge to clinically diagnose. There are 68 cases that have been reported in the English literature worldwide and 16 cases reported on the nasal septum. Diagnosis requires detailed histopathologic examination and the differential includes a variety of epithelial and mesenchymal derived tumors and malignancies. Definitive treatment can include endoscopic resection as well as laser ablation. The literature suggests a possible hormonal influence and a female predilection; however, definite risk factors and prognostic indicators have yet to be described. Angioleiomyoma is not commonly included in the differential diagnosis for an obstructive nasal mass and even less often

for a mass of the nasal septum. Despite the rare nature of the tumor, case reports are accumulating and may provide further information regarding risk factors, immunoreactivity, recurrence rates, prognosis, and anatomic sites of presentation.

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