Subcutaneous Nasal Angioleiomyoma: Case of a Rare Tumor and Review of the Literature

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Introduction

Leiomyomas are smooth muscle tumors categorized as either vascular or non-vascular lesions. Vascular leiomyomas, or angioleiomyomas (ALM), are rare, benign tumors arising in the deep dermis and subcutaneous tissue. They are derived from smooth muscle often in association with the media of vessel walls. 1 This neoplasm most commonly occurs in women between the fifth and sixth decade, and may arise anywhere in the skin. ALMs are typically well-circumscribed, slow-growing tumors less than 2 centimeters in size.2 The majority of these tumors arise on the extremities and characteristically exhibit pain exacerbated by cold temperature. Such pain is postulated to originate from either nerve involvement or contraction of the tumor’s smooth muscle and resulting ischemia.3,4 An estimated 10% of these tumors arise in the head and neck where they are generally painless and predominantly in males.

Case Report

Intrasal ALMs are rare, comprising only 1% of all benign tumors of the nasal cavity and less than 1% of all angioleiomyomas.1-5 We present an interesting case of a 46-year-old woman with a 5-year history of a left nasal mass. The mass progressively grew to 2 cm in diameter, resulting in nasal deformity, left-sided nasal obstruction, and unremitting pain, especially when exposed to cold. Her pre-operative Nasal Obstruction Symptom Evaluation (NOSE) questionnaire was 100/100 for the left side only. Examination showed a tender, mobile mass with overlying telangectasias at the lateral nasal sidewall extending to the medial cheek (Fig. 1). Within the nasal cavity, the tumor had no mucosal component and was visualized abutting the septum.

Magnetic resonance imaging (MRI) with contrast was performed, revealing a well-circumscribed, contrast-enhancing, benign-appearing subcutaneous mass with no extension and hyperintensity on T2 and FLAIR sequences. The mass was excised via an external rhinoplasty approach and appeared as a hypervascular, well-circumscribed, red mass limited to the subcutaneous tissues (Fig. 2).

Pathology

Evaluation of the tumor revealed it was grossly well-circumscribed and vascular. On H&E staining, it consisted of well-formed, vascular channels with small lumina and thick walls (Fig. 4). Trichrome staining demonstrated vascular smooth muscle fibers forming an inner, orderly circumferential layer and outer fibers swirling away, merging with less orderly intervening smooth muscle (Fig. 5). The smooth muscle origin of the fibers was demonstrated by their strong staining with smooth muscle antigen and desmin (Fig. 6).

Figure 1  Figure 2  Figure 3  Figure 4  Figure 5  Figure 6

Discussion

Only 13 cases of ALMs have been reported in the external nose involving the nasal dorsum and tip, none of which presented with obstruction or pain. In this way, the tumor described behaved more similarly to intranasal tumors than those located on other areas of the head and neck. This is the first report of an ALM presenting more similarly to intranasal tumors, yet located in subcutaneous tissue and more amenable to external rhinoplasty for excision.6

Our case appears is unique resulting from additional efforts for reconstruction. Due to the size of the lesion resulting in depression of the upper lateral cartilage, reconstruction with auricular cartilage grafting was required to restore patency of the nasal airway and symmetric appearance of the nose.

Diagnosis of ALM prior to excision is historically difficult, with cytology from fine needle aspiration and imaging studies only providing relatively nonspecific findings.4,5 Immunohistochemical staining for actin, myosin, desmin and CD31 can help rule out other tumors or malignancies that should be included in the differential diagnosis, such as: myopericytoma, hemangioma, angiolipoma, angiomylipoma, angiomyosarcoma, and leiomyosarcoma. MRI features of a mass consistent with ALM include: well-circumscribed, hyperintense on T2, and contrast enhancing.3

Conclusions

- Angioleiomyomas are rare, benign tumors arising in the deep dermis and subcutaneous tissue. They are derived from smooth muscle often in association with the media of vessel walls.
- Intrasal ALMs are quite unusual, comprising only 1% of all benign tumors of the nasal cavity and less than 1% of all ALMs.
- The histologic pattern of ALM is of spindle shaped cells consisting of a mixture of well-differentiated smooth muscle and thick-walled vessels.
- The tumor is benign and recurrence rates are extremely low, ranging from 0%-4.1,4,5
- Approach considerations were based on complete access to tumor, ability to perform framework reconstruction for functional deficits, and only minimally visible scarring. The external rhinoplasty approach fulfills all these criteria and provides excellent aesthetic outcomes.

References