

Subcutaneous Angioleiomyoma of the Nasal Tip. Report of a Rare Case

MARIA G. ONESTI¹, MICHELE MARUCCIA¹, SARA CARELLA¹,
ANTONIO ROSSI¹, GIUSEPPE SODA² and NICOLÒ SCUDERI¹

¹Department of Plastic and Reconstructive and Aesthetic Surgery, Sapienza University of Rome, Rome, Italy;

²Department of Anatomic Pathology, Sapienza University of Rome, Rome, Italy

Abstract. *Angioleiomyoma is a rare benign tumor of smooth muscle origin. It has been reported in many anatomical sites, but it is very rare in the nose. This article describes the case of a 46-year-old woman, with a subcutaneous vascular leiomyoma of the nasal tip. She reported a small painless mass. The skin which was covering it became strongly erythematous at high temperatures, with an annoying increase of the sensitivity. There are no specific imaging techniques capable of characterizing the vascular leiomyoma, however sonography represents an excellent modality for initial evaluation of this kind of lesion. We chose excision with an open rhinoplasty approach for treatment of the lesion. Obvious and disfiguring scars do not remain with this surgical approach. The patient had immediate and complete relief of her symptoms, the aesthetic result is very good and her satisfaction is complete.*

Angioleiomyoma, or vascular leiomyoma, is a rare benign soft-tissue tumor which consists of a mixture of well-differentiated smooth muscle cells and thick-walled vessels. Common sites are the uterus (95%), skin (3%) and the gastrointestinal tract (1.5%) (1), although all body sites may be potentially affected. Angioleiomyomas constitute fewer than 1% of all benign tumors of the nose and paranasal sinuses (2). This rarity is partially due to the scarcity of smooth muscle in the nasal cavity, except for the vessel walls. Even more rarely described, is its subcutaneous localization in the nose. This type of lesion occurs more frequently in females than in males, and pregnancy may increase the severity of the pain it causes (3). Clinically, this tumor appears as a small (<20 mm, seldomly above this size)

(4), freely-movable, subcutaneous nodule, causing pain in approximately 60% of patients (5). We present a rare case of a patient who had an angioleiomyoma of the nasal tip. We describe the clinical and histopathological findings, and surgical treatment of this lesion, which is rarely described in the literature.

Case Report

In March 2012 a woman of 46 years of age was brought to our attention at the Department of Plastic Surgery of Umberto I Polyclinic of Rome, affected by a subcutaneous lesion of the nasal tip. The patient reported that the lesion had appeared about 5 years earlier. One year previously, she reported a progressive increase in the size of the mass and a color change. The skin covering the mass became strongly erythematous at high temperatures, with an annoying increase of its sensitivity. Palpation revealed a firm, elastic, mobile mass, located between the triangular cartilage and the left crus. The patient underwent an ultrasound examination, which confirmed the presence of a round nodule, located in the subcutaneous adipose tissue. The mass had a relatively homogeneous hypoechoic echo texture, with a small amount of posterior acoustic enhancement (Figure 1). No internal calcification was present, and evaluation by color Doppler revealed diffuse arterial hypervascularity. The patient, with an uncertain preoperative diagnosis, underwent surgical excision under local anesthesia. We chose an excision with the open rhinoplasty approach. This approach is an easy and comfortable technique for the excision of subcutaneous masses on the nasal tip and dorsum. An incision was performed at the level of the columella and extended inside the nostrils. Dissection of the dorsum of the nose was performed and the reddish lesion, which was situated between the cartilages, was then evident (Figure 2). At the time of surgery, the mass was easily separated from the underlying upper lateral cartilages and from the overlying skin and was easily removed. It was a hard, round-shaped tumor, measuring approximately 1.45×1.28 cm in diameter,

Correspondence to: Michele Maruccia, MD, Sapienza University of Rome, Via Mongiana n.28, 00126, Roma, Italy. Tel: +39 3397765379, e-mail: marucciam@gmail.com

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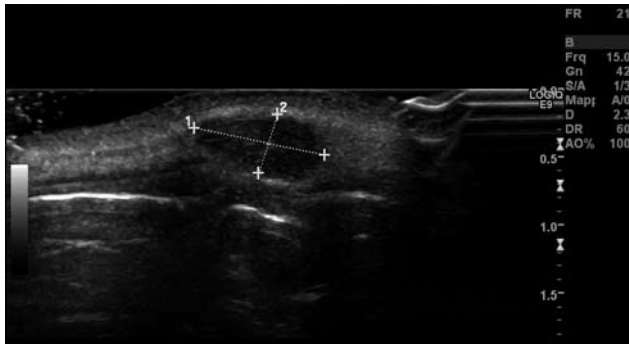


Figure 1. Ultrasound examination showed a homogeneous, hypoechoic-echo textured lesion, in the subcutaneous soft tissue with a small amount of posterior acoustic enhancement.

not adherent to the deep or superficial tissues. We performed a complete excision with careful hemostasis. The histopathological examination described a well-circumscribed, non-capsulated nodular tumor made of smooth muscle bundles, closely compacted and intersecting with one another, and surrounded by thick fibrous tissue (Figure 3). The smooth muscle bundles had uniform spindle cells, with eosinophilic cytoplasm and cylindrical nuclei with blunt ends. The proliferation surrounded blood vessels and slit-like vascular channels. Immunohistochemistry for smooth muscle actin (SMA) and desmin, highlighted the presence of muscular fibers, therefore confirming the diagnosis of a vascular leiomyoma. The postoperative course was uneventful and the patient had immediate and complete relief of her symptoms. Obvious and disfiguring scars do not remain with this surgical approach and the cosmetic result is very good (Figure 4). The patient's satisfaction is complete.

Discussion

Vascular leiomyoma is a tumor rarely reported in literature consisting of a mixture of well-differentiated smooth muscle cells and thick-walled vessels. Superficial leiomyomas are usually confined to the subcutaneous tissue of the skin, only rarely occurring in the deep soft tissues (6). They arise from structures that contain smooth muscle, such as the *arrectores pilorum*, the *tunica dartos* and the media of large and small blood vessels. They grow slowly and may persist for a long time. Histological classification of tumors by the World Health Organization (WHO) (7), divided leiomyomas into three groups: leiomyoma, angioleiomyoma (vascular leiomyoma or angioleiomoma) and epithelioid leiomyoma (bizarre leiomyoma and leiomyoblastoma). Angioleiomyomas may derive either from the surface or deep tissue. In both cases, the neoplasia seems to develop from the vessels of smooth muscle. Stout (8) published the first comprehensive review of this rare lesion in

1937, and it has been well-characterized in the literature ever since (9-11). The first report of an intranasal leiomyoma was by Maesaka *et al.* in 1966, concerning an angioleiomyoma (12). Between 1966 and 2003 only 15 cases of leiomyomas of the nasal cavity were described, according to Campelo *et al.*'s review (13). As regard to the subcutaneous tissue of the nose, in the literature there are very few cases of this histological type. Wang *et al.* (14) reported 21 cases of angioleiomyoma of the head and neck, between 1988 and 2001, out of which only three were of the external skin of the nose.

Three theories have been put forward for the origin of angioleiomyomas: from aberrant undifferentiated mesenchyme, from smooth muscle in the wall of blood vessels, or from both (15). Angioleiomyoma of the nasal cavity and paranasal sinus is an extremely rare benign neoplasm which is more common in females (3.75:1 female:male ratio) (3).

These tumors can affect any age, but are more common in people between 30 and 60 years of age (10). Due to their rarity, leiomyomas are usually misdiagnosed clinically and their presence is identified only after histological examination. Although they are benign tumours, and no malignant progression has been reported, the presence of benign areas of leiomyoma within leiomyosarcoma specimens raises this possibility, therefore treatment is recommended. Characteristic fusiform cells with a round central nucleus arranged in fascicles suggest smooth muscle differentiation. Special stains, such as phosphotungstic acid-hematoxylin, aniline blue, and Masson's trichrome, are helpful in differentiating muscle from collagen. Immunohistochemical stains are also helpful in establishing a diagnosis.

Differential diagnosis of an angioleiomyoma should include myopericytoma, hemangioma, fibromyoma and leiomyosarcoma (3). Myopericytomas are composed of thin-walled vascular channels with whorled rounded to ovoid myopericytes which are positive for SMA and negative or only focally-positive for desmin. This case was diagnosed as an angioleiomyoma since the immunocytochemistry revealed a strongly positive response for SMA and desmin with the histological fascicular pattern of spindle-shaped cells suggestive of leiomyoma. In hemangiomas, the intervacular stroma does not have smooth muscle bundles as found in an angioleiomyoma. According to the WHO classification of head and neck tumors, angioleiomyomas are vascular leiomyomas and leiomyoblastoma is considered as a synonym. A leiomyosarcoma is a malignant smooth muscle tumor with nuclear atypia and mitosis. An occasional mitotic figure can occur in angioleiomyoma. Angiofibromas are tumors with proliferated thin-walled staghorn vascular channels in a stroma containing round to stellate to spindle-shaped fibroblasts.

Hormone receptors may play a role in the tumorigenesis of extrauterine smooth muscle tumors, like the uterine



Figure 2. Intra-operative image of the angioleiomyoma.

counterparts. Marioni *et al*. (3) documented a case of a nasal vascular leiomyoma which was positive for progesterone (PR) and negative for oestrogen (ER) by immunohistochemistry. However, Kim *et al*. (16) reported a case of a nasal vascular leiomyoma in a female patient with lack of immunoreactivity to both ER and PR. In the present case, the immunohistochemical study demonstrated that neither ER nor PR was expressed in the tumor. Sex hormone dependence is still controversial regarding nasal vascular leiomyomas. More studies are needed to clarify the influence and mechanism of action of sex hormones on this tumor type.

There are no specific imaging techniques capable of characterizing the vascular leiomyoma, however sonography represents an excellent modality for initial evaluation of this kind of lesion. In this case, sonography was essential for describing the location and size of the tumor. Treatment for these tumors is based on local resection, and there are no reports of recurrence after total excision (17, 18); Bloom *et al*. (17) reported a case of spontaneous tumor recurrence after incomplete surgical excision. This event shows potential recurrence after incomplete resection and the need for complete excision in order to guarantee a definitive treatment. The development of infection in a subcutaneous lesion may suggest the diagnosis of a sebaceous cyst (19), however this does not exclude tumors, and the clinical diagnosis should be verified by histopathological examination of the excised specimen. Although extremely rare in the head and neck, leiomyomas should be included in the differential diagnosis of well-circumscribed and encapsulated lesions.

In conclusion, although angioleiomyoma is an infrequent soft-tissue tumor, it does have a typical, but non-specific presentation. Excision of the lesion enables a histopathological diagnosis, and offers a complete resolution of symptoms to the patient. The open rhinoplasty approach is an easy and comfortable technique for the excision of subcutaneous masses on the nasal dorsum or tip. It is also

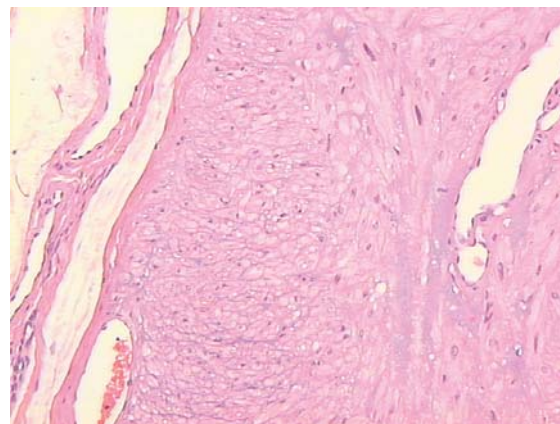


Figure 3. Histopathological examination: Clean presence of closely compacted, intersecting smooth muscle bundles surrounding slit-like vascular channels. No evidence of necrosis, pleomorphism, mitoses or nuclear atypia. Hematoxylin-eosin staining, $\times 100$.



Figure 4. Postoperative outcome: The aesthetic result is very good.

important to note that an external surgical approach could create visible or pathological scars and problems of vascularization, since the skin of the nasal tip is exposed to various tension forces and it has terminal vascularization.

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