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Nasal smooth muscle tumor of uncertain malignant potential: A case report

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ABSTRACT

Smooth muscle tumors that cannot be histologically classified as leiomyomas or leiomyosarcomas are defined as smooth muscle tumors of uncertain malignant potential. The location of these tumors in the nose is very rare, and the appropriate surgical extent to manage these neoplasms has not been adequately defined.

Here we describe the case of a 16-year-old female adolescent who consulted due to a vascular-like tumor in the right nasal cavity who was successfully treated with intranasal surgery. The histological diagnosis was smooth muscle tumor of uncertain malignant potential.

Given that these neoplasms are rare, the uncommon location in the nose, and the lack of evidence indicating the extent of surgery, it is relevant to describe and discuss this clinical case.

Keywords: *nasal tumor; smooth muscle tumor; uncertain malignant potential; nasal cavity; endoscopic surgery through natural orifices.*

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INTRODUCTION

Smooth muscle tumors that cannot be classified as benign or malignant based on their histopathological diagnosis are defined as smooth muscle tumors of uncertain malignant potential (STUMP). STUMP are mesenchymal neoplasms of the smooth muscle that comprise leiomyomas —and their subtypes— and leiomyosarcomas.

The system used to classify uterine smooth muscle tumors proposed by Bell et al.[1] differentiates conventional leiomyosarcomas —which have at least 2 of the following 3 features: diffuse cytologic atypia, tumor cell necrosis, and ≥10 mitoses per 10 high-power fields (HPF)— from leiomyomas —which are defined as neoplasms without tumor cell necrosis and ≤4 mitoses per 10 HPF. Variants include mitotically active leiomyomas, which have between 5 and 19 mitoses per 10 HPF, and atypical leiomyomas, which have cytologic atypia, <10 mitoses per 10 HPF, but no tumor cell necrosis. Tumors that do not meet these definitions are classified as STUMP.

The fact that these neoplasms are uncommon and their rare location in the nasal cavity, in addition to the difficulty in establishing an adequate management, denote the relevance of describing this case and discussing this condition.

CASE REPORT

A 16-year-old female adolescent consulted due to a tender tumor located in the right nasal ala present for the past 6 months. Erythema and telangiectasis were observed in the skin overlying the lesion.

By anterior rhinoscopy, a tumor was observed protruding into the anterior nasal cavity covered by normal mucosa.

A nasal endoscopy confirmed the anterior location of the neoplasm.

A hypoechoic tumor with abundant blood flow inside (more than 10 vessels per cm^2) was diagnosed by ultrasound (*Figure 1*).

A magnetic resonance imaging (MRI) with contrast of the facial bones showed a solid lesion with intermediate signal intensity in T2 sequence and low signal intensity in T1. A very high enhancement was noted after the administration of intravenous contrast. The lesion measured 18 mm × 22 mm × 21 mm and was compatible with a vascular lesion (possible malformation), so a frozen biopsy was not performed.

Surgical treatment under general anesthesia was indicated; interstitial laser coagulation of the tumor was performed using a diode laser in contact mode at 6 watts with bleomycin injection at 2 mg in a 1 mg/mL dilution. The surgery was performed intranasally under endoscopic vision.

The patient had a good clinical course and was discharged 24 hours later. No tumor shrinkage was observed during the control visits for 2 months.

A new endoscopic endonasal surgery was indicated to collect material for histopathological examination.

An electrocautery incision (Colorado tip) was performed in the area where the tumor showed the greatest intranasal bulging; the tumor was dissected and completely resected, preserving the cartilage and overlying skin of the nasal ala *(Figure 2*).

Fibrillar absorbable hemostatic material was placed in the cavity resulting from tumor resection and an absorbable compression material was placed inside a glove finger to be used as an anterior tamponade.

No frozen biopsy was done.

There was no significant intra- or post-operative bleeding.

The patient was discharged from the hospital the following day and the tamponade was removed 48 hours after the procedure.

The deferred histopathological examination of the surgical specimen (3.2 cm × 2 cm × 1.3 cm) reported tumor proliferation consisting of cells with elongated, irregular nuclei, forming intertwined bundles in variable directions, with no evidence of necrosis. STUMP diagnosis was confirmed by immunohistochemistry (*Figure 3*).

The resection was marginal.

Clinical, endoscopic, and MRI controls showed no evidence of tumor recurrence during the 18-month follow-up (*Figure 4)*.

DISCUSSION

Mesenchymal smooth muscle tumors of uncertain malignant potential were defined by the World Health Organization as those tumors that cannot be histologically classified as benign or malignant, accounting for an intermediate grade between leiomyomas and sarcomas.

Smooth muscle tumors located in the upper airway are extremely rare, accounting for less than 2.5% of mesenchymal neoplasms.[2,3] Fu et al.[3] described 2 leiomyomas and 6 leiomyosarcomas in a study of 256 non-epithelial neoplasms of the nasal cavity, paranasal sinuses, and nasopharynx.

Figure 1. Ultrasound of left nasal ala: hypoechoic tumor with abundant blood flow inside (A). Magnetic resonance imaging: solid lesion with intermediate signal intensity in T2 and low signal intensity in T1, with very high enhancement after intravenous contrast administration (B, C)

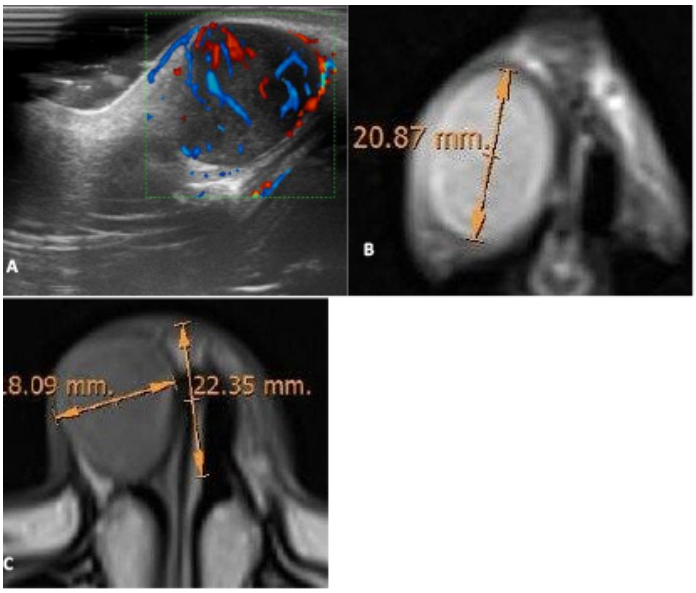


Figure 2. Tumor in right nasal ala, with erythema and telangiectasis in the overlying skin (A). Intranasal tumor resection with preservation of cartilage and skin of the nasal ala (B, C)

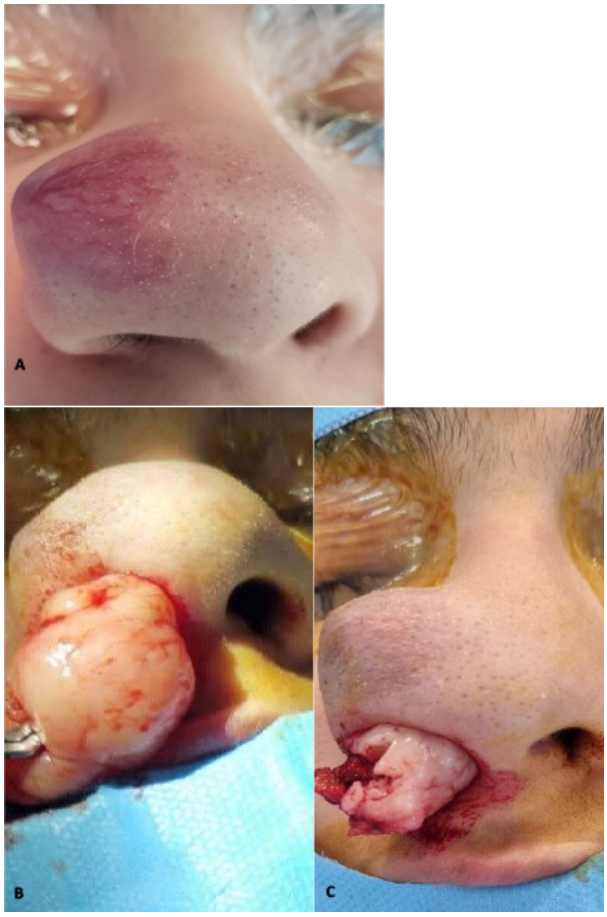
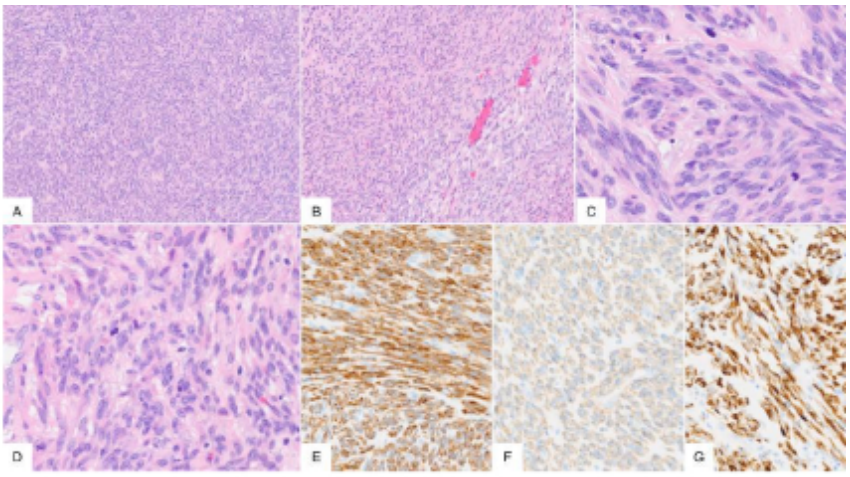
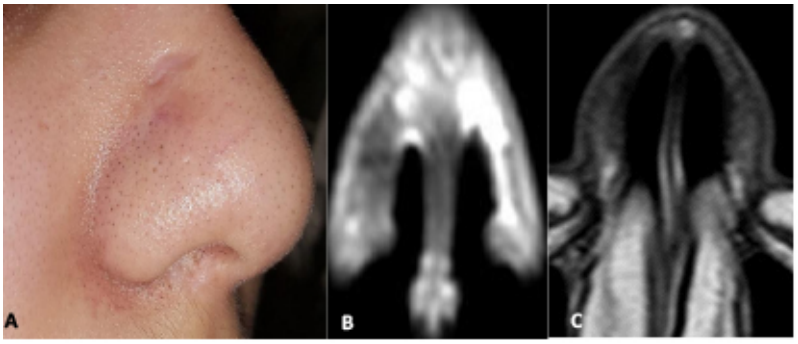


Figure 3. Smooth muscle tumor of uncertain malignant potential (STUMP)



A and B: Tumor proliferation consisting of cells with eosinophilic cytoplasm and elongated, mildly irregular nuclei, forming intertwined bundles in variable directions (hematoxylin and eosin staining; 10X).
C and D: 4–5 mitoses per 10 high-power fields (hematoxylin and eosin staining; 40X).
The immunohistochemistry found that tumor cells were positive for calponin (E), smooth muscle actin (F), and desmin (G).

Figure 4. Post-operative image of patient at 18 months (A) and post-operative magnetic resonance imaging (B, C)



Most of the descriptions found in the bibliography refer to tumors located in the uterus.

The pathological diagnosis of benign and malignant lesions in relation to uterine smooth muscle tumors is based on the combination of several criteria: cytologic atypia, mitotic index, and presence or absence of tumor cell necrosis.[4]

The combination of a mitotic index of 10 mitoses per 10 HPF, diffuse cytologic atypia, and coagulative necrosis is associated with metastasis. The presence of necrosis is an important finding, since it is present in 80% of leiomyosarcomas. In the absence of tumor necrosis, the factor that determines the potential in neoplasms with focal, multifocal, or diffuse atypia is the mitotic index, where the most aggressive tumors are those with ≥10 mitoses per 10 HPF. Bell et al.[1] established that tumors with this degree of atypia and <10 mitoses per 10 HPF are atypical leiomyomas with low risk of recurrence, given that they described only 1/46 cases of recurrence.

The immunohistochemical diagnosis is very useful to differentiate between benign and malignant neoplasms, together with the histopathological examination.

Most of these tumors may originate in the smooth muscle present in the walls of the vessels of the nasal cavity. A study reported a series of 12 smooth muscle tumors located in the nose; the distribution by sector was as follows: 8 in nasal cavity, 2 in paranasal sinuses, and 2 in nasal cavity and paranasal sinuses. Their histology corresponded to leiomyomas (7/12), STUMP (2/12), and low-grade leiomyosarcomas (3/12).[5]

These neoplasms are managed surgically, in an attempt to achieve complete resection.[6]

Given the infrequent nasal location, no factors have been determined to have an incidence in STUMP prognosis.

The extent of surgery in tumors with uterine location has not been clearly established. No significant differences have been seen in prognosis or survival by performing lumpectomy or hysterectomy with double adnexectomy.[2]

In the case described here, resection was complete but marginal; resection with free margins would have required a partial right rhinectomy with reconstruction.

Due to the uncertain potential of the neoplasm, the age of the patient, and the lack of studies suggesting a better prognosis with more aggressive surgeries, we opted to perform an endonasal surgery instead of extending the surgical margins (rhinectomy) in another procedure.

Smooth muscle tumors of uncertain malignant potential located in the nasal cavity or paranasal sinuses are very rare, and even more so in children. The diagnosis is based on histological findings and it is essential to consider the number of mitoses, the degree of atypia, and tumor necrosis, together with immunohistochemistry.

The best management is complete surgical resection; however, depending on the nasal or sinusal location of the tumor, the functional or aesthetic defect that may be caused by wide resection with free margins should be considered because the impact on prognosis is not known with certainty. ■

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