

## Case Report

### A Widely Destructive Leiomyoma of the Nasal Septum – An Unusual Presentation

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#### ABSTRACT

Leiomyoma of nasal cavity is a rare and benign tumor of smooth muscle origin that uncommonly arises from the nasal septum. We present an unusual case of histopathologically diagnosed locally extensive leiomyoma of the nasal septum which was clinically and radiologically misdiagnosed as malignancy of the nasal cavity. This case report emphasizes the rare occurrence of this entity at this site and highlights the need to consider this pre-operatively to avoid its associated significant bleeding and altogether different clinical management of this benign entity.

**Keywords:** Leiomyoma, Nasal Septums

#### Introduction

Leiomyomas are benign tumors of myogenic origin that commonly occurs in uterus, frequently in the walls of the alimentary tract and rarely in the skin. They are very rarely found in head and neck area. In the nasal cavity and paranasal sinuses they contribute about 1% of all benign tumors (1). A review of literature revealed only 29 reports of leiomyomas of the nasal cavity, of which only 8 cases have been reported to be arising from nasal septum (2, 3).

Considering the extreme rarity of this tumor, here we describe the clinical, radiological

and histological features of a case of a widely destructive leiomyoma of the nasal septum arising in a 50 year old female, followed by a brief review of literature pertaining to the histogenesis and histopathology of this rare lesion arising in this location.

#### Case Report

A 50 year old female patient presented to Otorhinolaryngology Outpatient Department with complaints of left sided nasal obstruction, pain and headache since 2 years. No history of epistaxis was given. Ear nose and throat (ENT) examination revealed a polypoid mass in left nasal cavity, with pink, smooth

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and glistening surface, fully occupying the left nasal cavity and also extending up to the nasopharynx. CT scan of the nose and para nasal sinuses revealed a lobulated heterogeneously enhancing soft tissue density mass (approx size 3x2.5x3.8 cm) (Apx Trans x CC) with multiple calcified foci in left nasal cavity, extending into bilateral ethmoid sinuses, left sphenoid sinus with erosion and displacement of nasal septum towards right side with erosion of floor of sphenoid sinus (Fig. 1A & 1B). Bilateral frontal and maxillary sinuses appeared clear without any evidence of thickening, sclerosis or bony abnormality. There was no evidence of any intracranial or intra-orbital extension. The radiological impression given was that of a malignant lesion – a squamous cell carcinoma of the nasal cavity.

Intraoperatively the mass was confirmed to be attached to the partially eroded nasal septum and the whole mass could be excised in pieces. There was significant bleeding when the mass was excised from its attachment and patient had to be transfused 2 units of blood. Reconstructive surgery was done to repair the defect. Post-operatively patient was also given 2 units of fresh frozen plasma in anticipation of consumptive coagulopathy.

### Pathological Findings

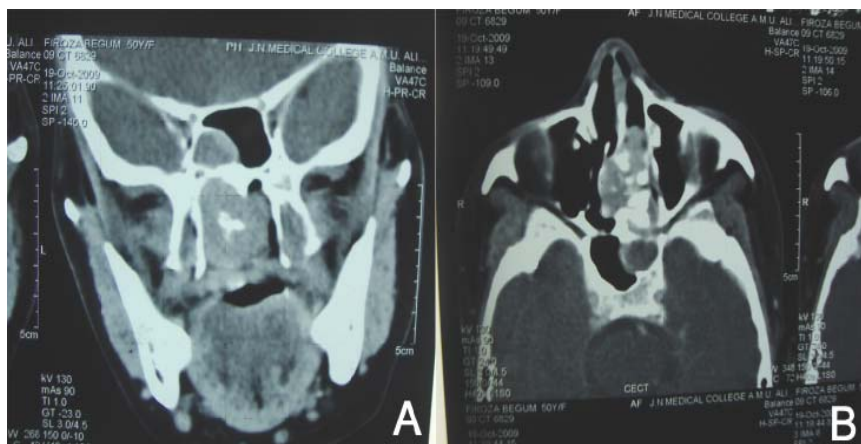
Grossly surgical specimen consisted of 3 soft

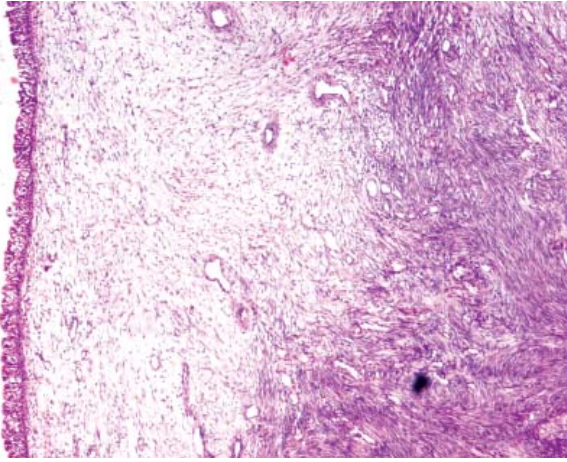
tissue pieces, largest measuring 2.5x1.5x1 cm. Each piece having glistening white surface. Cut surface was smooth and solid without any area of necrosis and hemorrhage. Whole tissue was submitted to paraffin blocking.

Histopathological sections (hematoxylin and eosin) from multiple blocks revealed an intact respiratory epithelium overlying a poorly circumscribed, partially encapsulated spindle cell tumor, comprising of long sweeping and intersecting fascicles of spindle cells having ovoid nuclei with rounded and blunt ends. There was minimal nuclear atypia and infrequent mitotic figures (Fig. 2). Surrounding stroma was mostly collagenous but also showed hyalinization at places. Stroma also showed few thin walled mostly collapsed vascular channels lined by single layer of endothelial cells (Fig. 3).

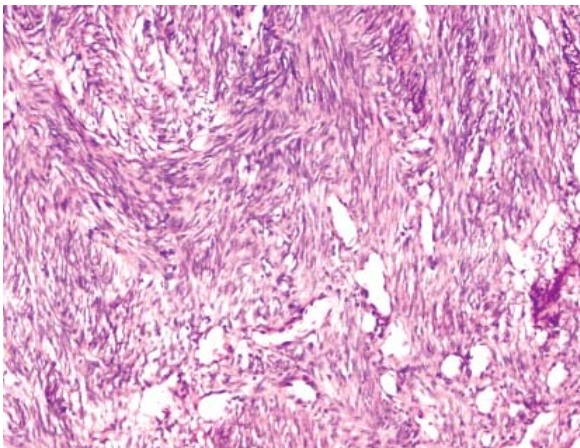
Immunohistochemically the tumor cells were strongly positive for following antibody panels—Dako mouse monoclonal antihuman - smooth muscle actin, (clone1A4), anti-vimentin (clone 3B4) and desmin (clone D33) and were negative for mouse monoclonal antihuman - neurofilament protein (clone 2F11), CD34 (classII, clone QB End10), cytokeratin (clone 34βE12), S100 polyclonal rabbit anti S100 (N1519RTU) and other markers, thus further confirming the histological diagnosis of leiomyoma. On follow up of about 6 months after surgery, no recurrence of the tumor was seen.

**Fig. 1- A:** CT Scan - a lobulated heterogeneously enhancing soft tissue density mass (approx size 3x2.5x3.8 cm) (Apx Trans x CC) in left nasal cavity with peripheral extension. **B:** CECT scan-Sagittal view showing un-circumscribed heterogenous mass in left nasal cavity





**Fig. 2-** Tissue section exhibiting a poorly circumscribed spindle cell tumor. Normal respiratory mucosa is on the left (H&E  $\times 100$ )



**Fig. 3-** Tissue section showing intersecting fascicles of spindle cells without nuclear atypia and mitoses with small thin walled blood vessels (H&E  $\times 200$ )

## Discussion

Maesaka *et al.* first reported a case of an intranasal leiomyoma – an angioleiomyoma in 1966 (4). Since then only 29 cases have been reported from nasal cavity, of which only 8 cases have been reported to be arising from nasal septum, indicating the extreme rarity of this condition. The rarity is partly attributable to the fact that smooth muscle is sparsely present in the nasal cavity, apart from the wall of blood vessels. Three hypotheses

have been proposed to explain the origin of smooth muscle tumors in the nasal cavity (5). Firstly, they may originate from the aberrant undifferentiated mesenchyme, secondly the presence of smooth muscle elements in the walls of the blood vessels may give rise to this tumor and finally they may arise from nasal vestibule as erector pilae muscle and sweat gland (6, 7). Microscopically leiomyomas are classified into two types – vascular and non vascular (8). On histology, the vascular type, which is less common, exhibits double walled vessels indicating that the mass originated in smooth muscle of the veins. Progressive development of smooth muscle tumor from hemangioma to vascular leiomyoma has been postulated (6). These vascular leiomyoma finally develop to solid leiomyoma with gradual reduction in vascularity (7).

Histologically smooth muscle tumors can be classified into three groups namely: Leiomyomas, Angiomyoma (vascular Leiomyoma) and Epithelioid leiomyoma (bizarre leiomyoma and Leiomyoblastoma). Morimoto classified these tumors into three histological types in 1973, solid or capillary, cavernous and venous (9). Tumors of solid type are composed of smooth muscle bundles which surround the vascular channels and are closely packed and intersect with one another. Cavernous type tumors are composed of dilated vascular channels with smaller amount of smooth muscle. Tumors of the venous type have vascular channels with thick muscular walls and smooth muscle bundles are not so compact. The most common type found in the head and neck area is the venous type (9).

Histopathological findings in our study were consistent with a poorly vascularized leiomyoma of solid type without any mitotic figures. It appears that the absence of mitosis

is the most useful histological indicator of benign lesions (2). Rarely local pressure of a leiomyoma may result in bony erosion and local tumor extension clinically mimicking a malignant growth which can only be sorted out by subsequent histopathological examination. Similar observation was made in our case.

The histologic differential diagnosis for these lesions includes hemangioma, nasal angiofibroma, myopericytoma, fibromyoma, leiomyoblastoma, hemangiopericytoma, angiomylipoma, angiosarcoma and leiomyosarcoma. In hemangiomas the intervascular stroma does not have smooth muscle bundles as in leiomyoma. Angiofibromas are tumors with proliferated thin walled staghorn vascular channels in a stroma containing round to stellate to spindle shaped fibroblasts. Myopericytoma 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. A leiomyosarcoma is a malignant smooth muscle tumor with nuclear atypia and mitosis. Malignant variants and recurrences of the leiomyoma have been reported, however they are rare. The most of the tumors were classified as benign, none showing mitoses or presence of atypia to suggest possible malignant behavior (10).

Leiomyomas exhibit no characteristic radiological findings, but CT or MRI is helpful in determining the extent of tumor invasion and in planning treatment. The leiomyomas that develop in nasal cavity are often of vascular origin hence a digital subtraction angiography may be done in anticipation of intraoperative bleeding.

The most satisfactory treatment for these lesions is complete excision with a surrounding

rim of normal mucosa. KTP532 laser assisted transnasal endoscopic approach provides complete removal of the disease and definitive treatment in small tumors confined to the nasal cavity (2, 3).

Thus to conclude, leiomyoma of the nasal septum is a rare benign neoplasm. Our was an unusual case as it was a widely destructive and extensive lesion which led the radiologist and ENT surgeons to form an erroneous impression of a malignancy and consequently patient had suffered significant bleeding intra-operatively and it was only after histopathological examination, the diagnosis of a solid leiomyoma of the nasal septum was made. Therefore, this case report prompts us to be aware of this rare entity and all measures should be accomplished to minimize intra-operative bleeding in anticipation of a vascular leiomyoma. Further a long term follow up is required to see for any recurrence.

### Acknowledgements

The authors declare that there is no conflict of interests.

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