

Histopathological Insights into Angioleiomyoma of the Nasal Cavity

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Abstract

Angioleiomyoma (ALM) is a rare soft-tissue tumour with an overall prevalence of fewer than 100 cases reported so far. ALMs are well-defined, painless, slow-growing lesions, making preoperative differentiation from leiomyomas or other mesenchymal tumors difficult. We present here a case of a 24-year-old female with an exophytic, cystic swelling in the nasal cavity clinically suspected to be a nasolabial cyst. Surgical excision is the primary treatment of ALM, with a low recurrence rate and only one reported case of malignant transformation to this date. This case highlights the importance of considering this rare benign lesion in the list of differential diagnoses for a fairly common complaint of sinonasal swelling and the definitive role of histopathology along with IHC for diagnosing such tumors.

Keywords: Angioleiomyoma, Sinonasal, Vascular leiomyoma.

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INTRODUCTION

Leiomyoma is a common benign smooth muscle tumour reported in several locations in the body. The common locations are the female genital tract (95%), cutaneous (3%) and the gastrointestinal tract (1.5%) as well.¹ ALM however, are rarer and account for 0.2 % of all head and neck benign tumours and 2% of total cases of tumours of the sinonasal tract.² These are well-defined, typically painless and slow-growing lesions, a feature similar to leiomyomas. Typically these are <2 cm in diameter, covered by normal mucosa, and are sometimes bluish or purple.³ They are solitary, usually occurring in the subcutis location and common in the head and neck region.¹ Other locations of ALMs are extremities and around main large vessels. Trauma-induced venous stasis, genetic factors, vascular malformations, and hormonal effects with a focus on oestrogen are postulated as potential etiological factors.⁴ Some studies have documented the aetiology of ALM related to oestrogen as is seen in uterine leiomyomas.⁵ The overall male-to-female ratio is 0.7:1 with variation in the M: F incidence.² The most common areas of occurrence are auricle, nose, lip, and neck. In the nasal cavity, locations of ALM include the inferior turbinate (being the most common), septum, and nasal vestibule.⁶

Case Report

A 24-year-old female patient presented to the ENT OPD on 19th March 2024 with swelling in the left nasolabial region for about 6 months which rapidly increased in size in the last month. On clinical examination swelling was exophytic, cystic, soft to firm in consistency, painless with no other signs of inflammation. Overlying skin was unremarkable and swelling was measuring about 1.5x1.5 cm in diameter. The patient had no other symptoms and signs of local infection or any other co-morbidity. A clinical diagnosis of Nasolabial cyst was made. However, NCCT & CECT-PNS reported an elliptical, well-defined, homogeneous and expansile enhancing iso-dense mass in the lateral wall of the left nasal cavity with no adjacent bony erosion or destruction suggestive of? Soft tissue neoplastic lesion. A surgical excision was performed and the specimen was sent for histopathological examination to the Department of Pathology. Gross examination showed a single capsulated soft tissue piece measuring 2x1.5x0.6cm. (Fig. 1) On the cut, grey-white homogenous whorl-like areas are seen with few haemorrhagic foci. No cyst was seen grossly. Microscopic examination revealed a capsulated cellular lesion comprising

of smooth muscle cells showing bland elongated blunt-ended nuclei with eosinophilic cytoplasm-forming smooth muscle bundles showing interlacing patterns with collagen fibres in between and the presence of several thin-walled congested vascular channels between the fascicles. Few large vascular channels of varying calibre, surrounded by spindle-shaped cells showing dilated and slit-like vascular spaces. At places the whorled bundles of muscle fibres were seen fused with the vessel walls. Histopathological diagnosis of Angioleiomyoma was made and IHC was advised for confirmation. (Fig. 2a, b, c, d). The tumor shows IHC positivity for desmin, SMA, CD34 and CD31. Therefore a final diagnosis of AML was reported. Patient was followed up for 6 months and was doing good.



Fig. 1: Gross specimen of angioleiomyoma measuring 3 cm×2 cm×2cm

DISCUSSION

The 2020 5th edition of World Health Organization (WHO) Classification of Soft Tissue Tumours for the first time considered Angioleiomyoma (ALM) as a separate entity from leiomyoma.⁷ ALM is included in the group of pericytic tumours, which share a perivascular growth pattern, a variable contractile phenotype, and represent 0.2% of all head and neck benign tumours.² The first sinonasal leiomyoma case was reported in 1966.⁸ Nasal ALM shows a female to male ratio of 2:1 and occur commonly in the fourth to sixth decade. Sinonasal is a female predominant location of ALM.⁹ The solid type is significantly more common in women, while venous and cavernous types are male predominant, and the venous type

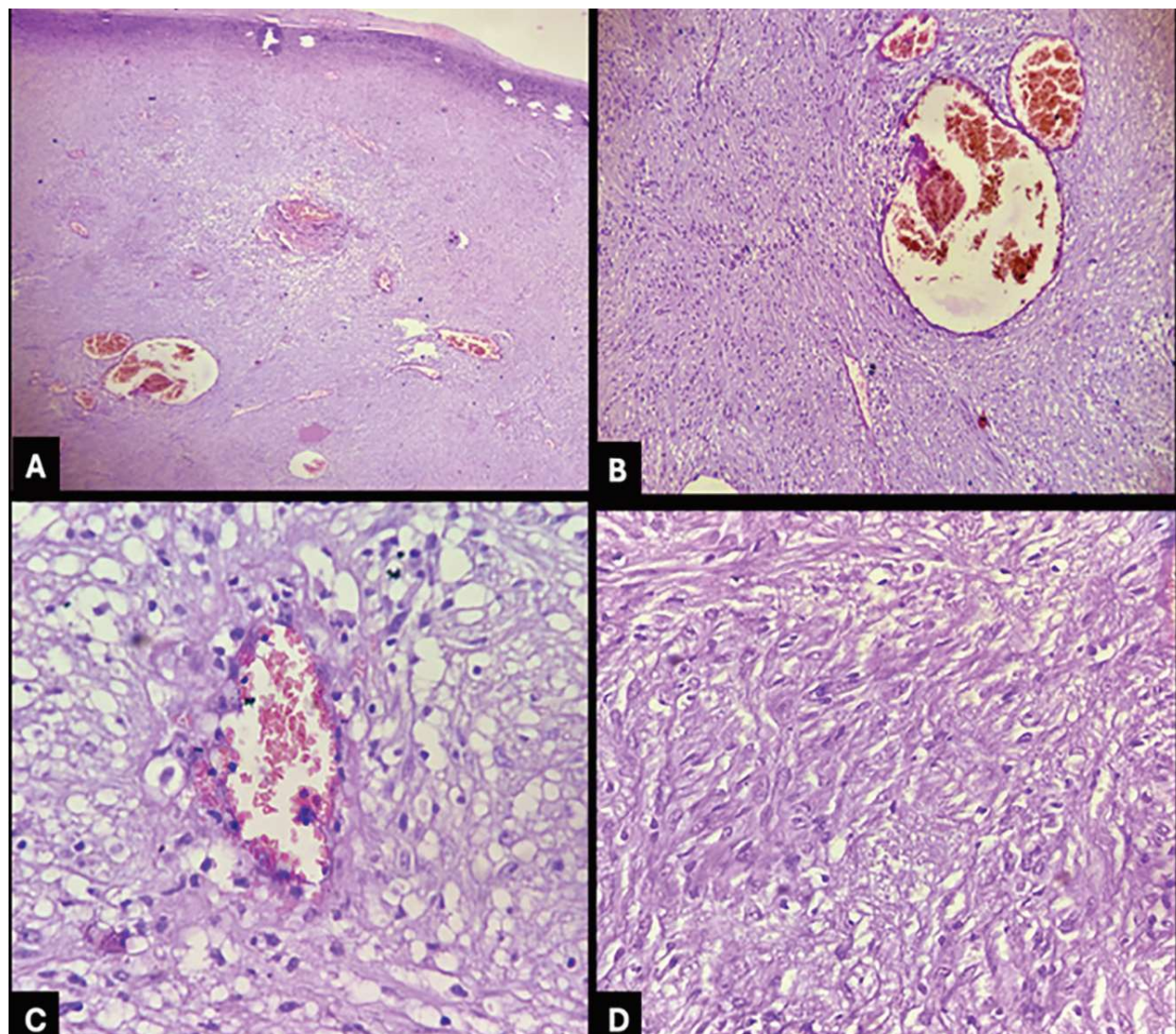


Fig. 2: Microphotographs of the tumor show cellular lesion comprising of smooth muscle cells showing interlacing patterns with collagen fibres in between and presence of several thin walled dilated congested vascular channels between the fascicles. (H and E stain a. 10X; b. 10X; c. 40x; d. 40X)

is reported to be more often involved in the head and neck region.²

It presents as a Solitary, slow-growing, mobile, firm and half of ALMs are painful cutaneous mass.¹⁰ Classic clinical presentation are nasal obstruction (seen in 56.25%), epistaxis (present in 56.25%), facial pain (in 25%), and headache (seen in 25%), few patients presenting with nasal discharge.⁸ These symptoms were caused by mass effect related airflow changes that caused nasal crusting and nasal desiccation.⁹ Hachisuga *et al* have described pain as burning with rapid worsening with light contact or exposure to cold which is thought to be due to active contraction of smooth muscle that results in local ischaemia.^{2,10}

Imaging with computed tomography (CT) is

widely used as an adjunct technique to diagnose the extent of the tumour growth⁶ Due to their nonspecific appearance on imaging ALM may be misdiagnosed as nasal polyp, fibroma, papilloma, haemangioma, or angiofibroma. The lesion in our case was initially suspected to be a nasolabial cyst based on clinical examination.^{6,8}

Grossly ALM typically is tan-pink, fleshy with a smooth surface. Histopathological evaluation is gold standard.^{6,8} Classical histopathologic features of ALM include well-defined fascicles or whorls of mature smooth muscle cells surrounding blood vessel lumina, cigar-shaped nuclei with blunt ends, and absence of nuclear atypia and mitosis. Severe atypia, pleomorphism with infiltrative borders, high mitotic count, and tumour cell necrosis

would suggest leiomyosarcoma.¹⁰ Morimoto *et al.* proposed a classification system, that divided ALM into solid, cavernous, and venous types which is now adopted and documented in the WHO classification of the same.¹¹ The differential diagnosis for ALM has a wide spectrum from ganglionic cyst, gouty tophus, tophi, glomus tumour, lipoma, hemangioma, foreign-body granuloma, superficial acral fibromyxoma, giant cell tumour of bone to desmoid tumour, neurofibroma, schwannoma, sarcoma and leiomyosarcoma. However, hemangioma, angiofibroma, myopericytoma, fibromyoma form the closest differentials of ALM.¹⁶ H & E along with immunohistochemistry can help in the definitive diagnosis of such lesion. IHC staining with actin, desmin, or myosin, for smooth muscle, and factor VIII or CD31 for vascular endothelium, can help differentiate from other similar lesion.^{9,10}

Though vascular in nature, excessive bleeding during surgical excision is not common. Complete surgical resection is advised and curative as recurrence after complete removal is extremely rare, regardless of the subtype.¹⁰ Heren *et al.* reported single case of malignant transformation so far to our knowledge.¹²

CONCLUSION

This case highlights the importance of histopathology in diagnosing such relatively uncommon tumor of nasal cavity. Clinicians should remember that ALM can be a differential diagnosis in the sinonasal area, especially in middle aged women. This benign tumor has excellent prognosis after surgical removal along with resolution of symptoms.

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Conflicts of interest

There are no conflicts of interest

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