Angiomyolipoma of the nasal cavity

Clinical Case

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Abstract

Angiomyolipoma is a hamartomatous lesion most frequently found in the kidneys and liver, presenting as solitary or multiple lesions and sometimes associated with tuberous sclerosis.
Fewer than 20 cases have been reported in the nasal cavity in the literature.
This case concerns a 75-year-old female patient who presented with recurrent epistaxis and unilateral nasal obstruction with a one-year history.
A lesion was identified in the right nasal vestibule. Surgical excision and biopsy confirmed a mesenchymal neoplasm with prominent vascular components, smooth muscle, and mature adipose tissue, consistent with nasal angiomyolipoma. Nasal angiomyolipomas are rare and may present diagnostic challenges due to nonspecific symptoms. Histopathology and surgical excision remain essential for diagnosis and treatment.

Keywords: angiomyolipoma; nasal cavity;

Introduction

Mesenchymal neoplasms, including angiomyolipoma, are extremely rare in the nasal cavity.[1] Angiomyolipoma is a hamartomatous lesion which is most frequently found in the kidney, followed by the liver, and may present as a solitary or multiple lesion, either isolated or associated with the tuberous sclerosis complex.[2,3] Nasal cavity angiomyolipoma is characterized by the presence of smooth muscle cells that lack epithelioid morphology, mature adipose tissue, and thick-walled blood vessels. Unlike angiomyolipomas found in other sites, nasal cavity angiomyolipoma does not have a melanocytic immunophenotype.[1]
Given its rarity in this location, fewer than twenty cases have been reported in the literature.[4] Because of its low prevalence and nonspecific presentation, this neoplasm represents a diagnostic challenge and requires a high index of clinical suspicion.

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Case report

A 75-year-old woman presented to the outpatient otorhinolaryngology clinic in December 2024 with a one-year history of recurrent, self-limited episodes of epistaxis and right-sided nasal obstruction. She also reported progressive growth of a lesion inside the right nasal vestibule. Anterior rhinoscopy/nasal endoscopy revealed a sessile, nonulcerated lesion with a hemorrhagic appearance measuring approximately 3 cm in its greatest dimension. The lesion originated from the lateral wall of the nasal vestibule and extended posteriorly to the head of the ipsilateral inferior turbinate. The patient denied other nasal symptoms, such as rhinorrhea, hyposmia, or facial pressure.

Her medical history included hypertension and dyslipidemia, managed with amlodipine (5 mg) and olmesartan (20 mg) combined with simvastatin (40 mg) daily.

Computed tomography (CT) of the nose and paranasal sinuses (PNS) revealed a 3.0 × 2.5 cm hypervascular soft-tissue lesion arising from the lateral wall of the right nasal vestibule, with no evidence of invasion of the adjacent structures (Figure 1). The patient underwent a complete excisional biopsy of the unilateral lesion under general anesthesia in the operating room after considering her age, the sessile nature of the tumor, and the need for a comprehensive assessment of its extension. Histopathological examination demonstrated a mesenchymal neoplasm with a prominent vascular component composed of thick-walled muscular vessels. Immunohistochemical analysis revealed positivity for muscle-specific actin (MSA), calponin, CD31, and CD34. The stroma contained bundles of smooth muscle fibers and areas of mature adipose tissue, which were negative for HMB-45, consistent with angiomyolipoma of the nasal cavity (Figure 2). The postoperative course was uneventful. At six months post-surgery, the patient remained asymptomatic with complete resolution of the nasal symptoms

Figure 1
Computed tomography CT (axial section) of the nose and paranasal sinuses (PNS) showing a lesion with soft tissue density in the right nasal vestibule, measuring 3.0 × 2.5 cm, and originating from the lateral wall.

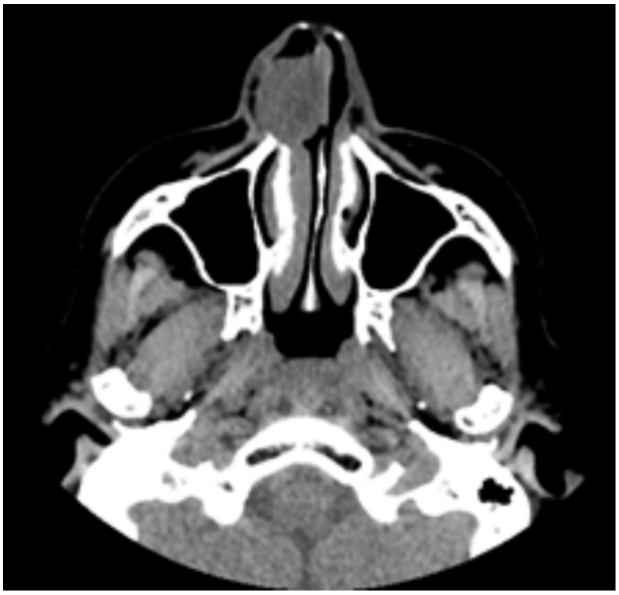
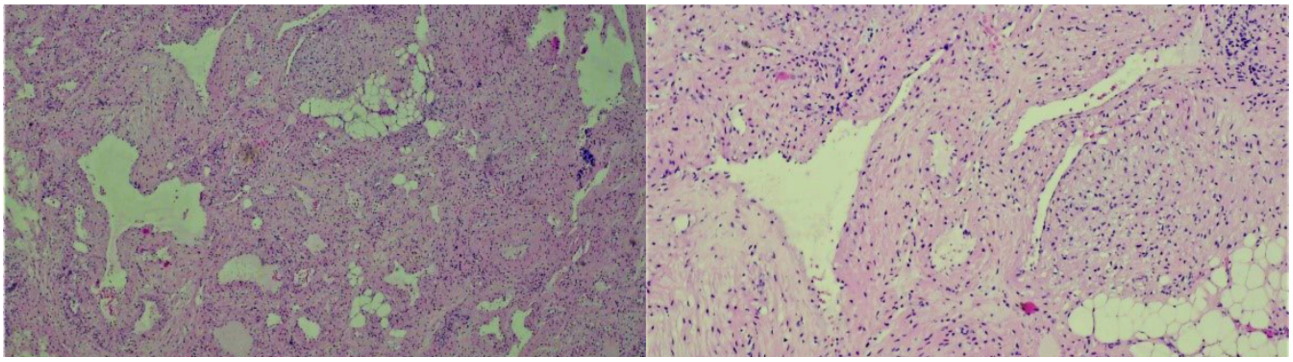


Figure 2
Biopsy of the lesion shows a mesenchymal neoplasm with a prominent vascular component, characterized by thick-walled muscular vessels that are positive for MSA, calponin, CD31, and CD34. The stroma contains bundles of smooth muscle with some mature adipose tissue.



and no evidence of local recurrence. She continues to undergo periodic follow-up at our institution.

Discussion

Vascular tumors of the nasal cavity and PNS are relatively rare, with angiofibroma being the most common tumor. Other rare types described in the literature include hemangiopericytoma, angioleiomyoma, and capillary and cavernous hemangiomas.[4]

Angiomyolipoma is a rare hamartomatous vascular tumor located in the nasal cavity and vestibule, with only a few reported cases. This entity can be classified into two subtypes: mucocutaneous, arising in the skin, nasal cavity, oropharynx, and larynx; and hepatorenal, corresponding to perivascular epithelial tumors.[4,5] Histopathologically, the mucocutaneous subtype consists of spindle-shaped smooth muscle cells without epithelioid morphology, whereas the hepatorenal subtype typically demonstrates epithelioid smooth muscle cells positive for melanocytic markers (HMB-45). Most angiomyolipomas are benign; however, malignant transformation has been documented in the hepatorenal subtype due to its epithelioid structure, and thus it carries a potential for malignancy.[1,3,5] Mucocutaneous angiomyolipomas occur predominantly in older men and, unlike the hepatorenal type, show no correlation with the tuberous sclerosis complex, which is associated with approximately 20% of renal and 15% of hepatic angiomyolipomas.[4] This report describes the case of an older female patient with a mucocutaneous angiomyolipoma originating in the nasal cavity, which was negative for melanocytic markers and without epithelioid morphology. Because of their rarity and nonspecific clinical presentation, nasal cavity angiomyolipomas can be easily misdiagnosed.

Conclusion

When assessing a benign-appearing lesion in the nasal cavity, angiomyolipoma should be considered among the differential diagnoses alongside more common entities such as nasal polyps, juvenile angiofibroma, squamous papilloma, and other hamartomatous neoplasms. Complete surgical excision establishes the histological diagnosis and serves as the definitive treatment.

Conflicts of interest

The authors declare that they have no conflict of interest regarding this article.

Data confidentiality

The authors declare that they followed the protocols of their work in publishing patient data.

Human and animal protection

The authors declare that the procedures followed are in accordance with the regulations established by the directors of the Commission for Clinical Research and Ethics and in accordance with the Declaration of Helsinki of the World Medical Association. Privacy policy, informed consent and Ethics committee authorisation. The authors declare that they have obtained signed consent from the participants and that they have local ethical approval to carry out this work.

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Scientific data availability

There are no publicly available datasets related to this work.

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