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Managing Sinonasal Hemangiopericytomas: A Case Study and Comprehensive Review

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Abstract

Introduction: Hemangiopericytomas are rare vascular tumors arising from pericytes surrounding capillaries, posing diagnostic challenges due to their size and clinical presentation. Diagnosis relies on radiological and histopathological assessments, with surgery as the primary treatment for nasosinusal hemangiopericytomas, which exhibit distinct characteristics compared to other locations.

Case Report: We present a case of a 35-year-old female, Mrs. NK, with no significant medical history, presenting with chronic nasal obstruction, left anterior rhinorrhea, and episodes of epistaxis. Complications included grade II exophthalmos and ipsilateral eye strabismus, alongside progressive left jugal swelling. Imaging revealed complete left maxillary sinus occlusion, leading to a nasal endonasal biopsy confirming sinonasal hemangiopericytoma. Despite initial excision, recurrence prompted further surgical intervention.

Discussion: Hemangiopericytomas, originating from mesenchymal cells with pericytic differentiation, are rare tumors primarily affecting adults aged 30-50 years. Clinical manifestations vary widely, emphasizing the importance of early detection and accurate diagnostic tools such as contrast-enhanced CT and MRI. Surgical excision remains the gold standard, supported by immunohistochemical markers and considerations for adjuvant therapies in extensive cases.

Conclusion: Sinonasal hemangiopericytomas represent a distinct clinical entity with unique diagnostic and therapeutic challenges. Although they demonstrate lower aggressiveness and improved outcomes with complete surgical resection, recurrence rates underscore ongoing management complexities and the need for comprehensive treatment strategies. This abstract highlights the clinical course, diagnostic approach, and therapeutic considerations in managing sinonasal hemangiopericytomas, emphasizing the importance of multidisciplinary collaboration and long-term follow-up in optimizing patient outcomes.

Keywords: Hemangiopericytoma; Sinonasal; Immunohistochemistry

Introduction

Hemangiopericytomas are uncommon vascular tumors that arise from pericytes surrounding capillaries [1,2]. Clinical diagnosis remains challenging due to the tumor's size and extent. Diagnosis relies on a combination of radiological findings and histopathological evidence. Surgery is the preferred treatment for nasosinusal hemangiopericytomas. Nasal localization exhibits specific characteristics that differentiate it from other hemangiopericytomas, showing lower aggressiveness but higher recurrence rates. Here, we present a case of a large hemangiopericytoma in the right nasal cavity managed at our institution.

Case Report

Mrs. N, K., aged 35, with no significant medical or surgical history, presented with chronic nasal obstruction associated with left anterior rhinorrhea and episodes of epistaxis. The symptoms complicated 3 months prior to admission with grade II exophthalmos and ipsilateral eye strabismus, all evolving in the context of progressively worsening left jugal swelling. A contrast-enhanced CT scan revealed complete filling of the left maxillary sinus, distending its walls, predominantly confined to the ostium which appeared enlarged (Figure 1). The patient underwent a nasal endonasal biopsy under general anesthesia. Histopathological examination revealed small to medium-sized round cells with abundant eosinophilic cytoplasm, occasionally exhibiting myxoid features, arranged around vessels characterized by thickened arteriolar muscular walls and turgid endothelium. Immunohistochemical analysis showed positivity for vimentin and negativity for CD34, HMB45, and PS1000, confirming the diagnosis of sinonasal hemangiopericytoma (Figure 2).

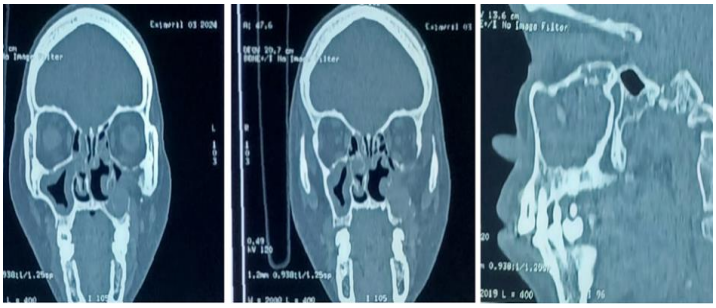

Figure 1: *A contrast-enhanced CT scan revealed complete filling of the left maxillary sinus.*

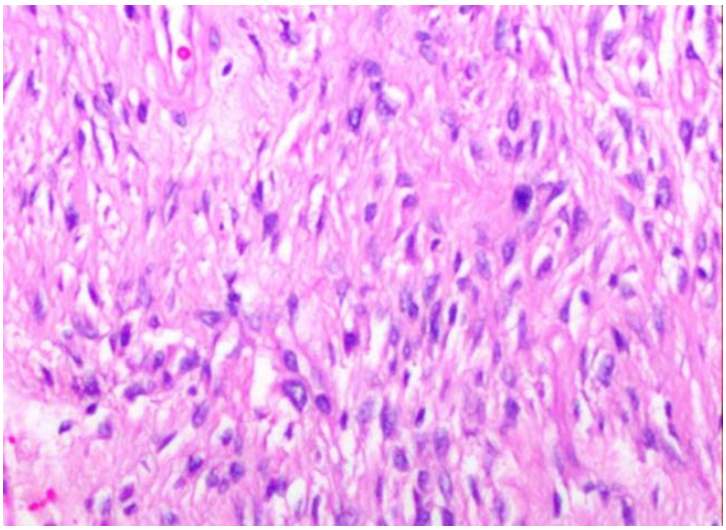

Figure 2: *Microscopy examination showing small to medium-sized round cells with abundant eosinophilic cytoplasm, occasionally exhibiting myxoid features.*

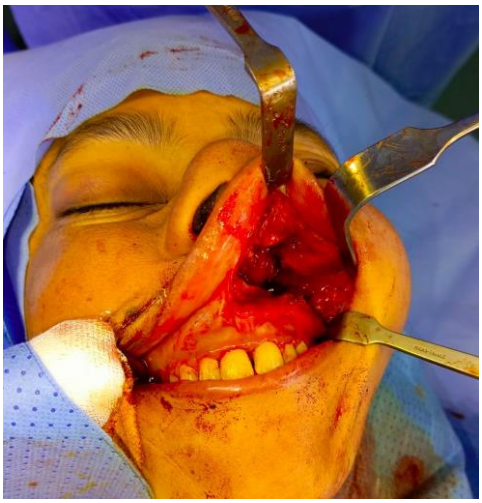

Figure 3: *Superior vestibular r approach.*

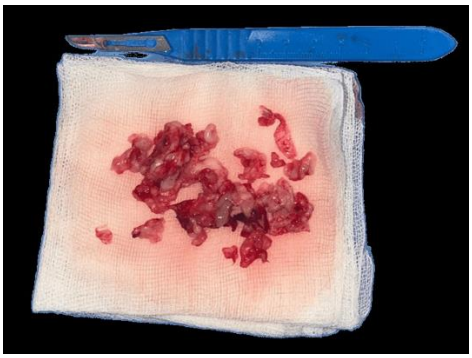

Figure 4: *Image of the surgical specimen after wide excision.*

The patient adamantly declined the lateral nasal wall approach, thus a wide excision of the tumor was performed via a superior vestibular approach (Figure 3) under general anesthesia. However, at the 6-month follow-up, the patient reported recurrence of left nasal obstruction. A follow-up CT scan was performed, revealing re-filling of the left sinus cavity indicative of potential recurrence. Following multidisciplinary team consultation, the patient underwent another wide excision (Figure 4) via the same surgical approach. The patient was subsequently reviewed in consultation and followed up with rhinocavoscopy, showing no detectable lesions at a 10-month follow-up.

Discussion

Hemangiopericytomas are categorized as vascular tumors originating from mesenchymal cells with pericytic differentiation [1,2]. These tumors are rare, constituting approximately 1% of vascular-origin tumors [3]. First described in 1942 by Stout and Murray [4], about 15% of hemangiopericytomas occur in the head and neck region [5]. Nasosinusal hemangiopericytomas typically manifest most frequently between the ages of 30 and 50 years, with an approximate 1:1 sex ratio [6]. Classified by the World Health Organization (WHO) among soft tissue tumors with low to

intermediate malignant potential [7], no specific etiological factors have been identified, except for a history of facial trauma reported in some cases [8]. Clinical presentation varies based on the tumor's location and stage. Some patients may remain asymptomatic until advanced stages, highlighting the importance of early diagnosis. Increasing tumor size can lead to symptoms such as nasal obstruction, epistaxis, and ophthalmic manifestations like exophthalmos, diplopia, blindness, and cranial nerve impairment [9]. Diagnosis relies on a combination of radiological findings and histopathological evidence. Initial evaluation typically involves contrast-enhanced CT scanning of the nasosinusal region to accurately characterize local findings, followed by MRI for further differentiation from inflammatory conditions. Angiography may be necessary given the tumor's vascular nature, potentially for preoperative embolization [10]. Definitive diagnosis requires histological examination, which can be challenging. Immunohistochemistry utilizes a panel of markers, where positive cytokeratin staining rules out hemangiopericytoma (HPC). Vimentin is typically positive in 98% of cases, while markers like actin and factor XIIIA may also show positivity; however, CD34 and PS100 are typically negative [11,12]. Surgery remains the treatment of choice for nasosinusal hemangiopericytomas. The endoscopic approach is recommended as first-line due to lower morbidity, except in cases of extensive local involvement where an external approach may be preferred [13].The role of radiotherapy and chemotherapy in sinonasal hemangiopericytomas remains controversial, with chemotherapy showing limited efficacy [14]. External beam radiation therapy is recommended for extensive tumors [15,16]. However, sinonasal hemangiopericytomas exhibit recurrence rates ranging from 7% to 50%, typically occurring 6 to 7 years after initial treatment [17].

Conclusion

Sinonasal hemangiopericytomas are recognized as a distinct entity, exhibiting less aggressiveness compared to those found in other locations and showing improved survival rates when completely surgically removed. Diagnostic challenges persist due to nonspecific clinical features, necessitating detailed architectural studies to exclude various differential diagnoses before confirming this histological type.

Conflict of Interest

The authors declare no conflicts of interest.

Authors' Contributions

All authors participated in the patient's care and in the writing of the article. All authors have read and approved the final version of the manuscript.

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Ethical approval

The authors declare that ethical approval was not required.

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