JK SCIENCE pISSN: 0972-1177

CASE REPORT

Glomangiopericytoma—A Rare Sino-nasal Tumour

Shravanthi Mantra Prithviraj, Shyam Sudhakar Sudarsan, Dinesh Ram Ravi, *Sarah Grace Priyadarshini

Abstract

Haemangiopericytoma, an uncommon vascular tumor originating from perivascular cells, encompasses a sinonasal variant known as glomangiopericytoma (GPC). A 75-year-old female presented with complaints of recurrent episodes of bleeding from the left nasal cavity for 8 months, along with a history of recurrent episodes of headaches and left nasal obstruction for 6 months. DNE was done and showed a polypoidal vascular mass filling the left nasal cavity. CECT PNS was done, showing signs of sinonasal malignancy. Biopsy showed features of low-grade spindle cell carcinoma. Debulking of the tumour followed by FESS with medial maxillectomy was done successfully, with currently no signs of recurrence.

Key Words : Haemangiopericytoma,Vascular Tumor,Glomangiopericytoma

Introduction

Haemangiopericytoma, an uncommon vascular tumor originating from perivascular cells, encompasses a sinonasal variant known as glomangiopericytoma (GPC).[1]

This specific type of haemangiopericytoma typically exhibits low malignant potential, presenting as a rare tumor within the respiratory mucosa, with a prevalence of less than 0.5% among all sinonasal tumors. [2]

The most frequently reported symptoms encompass nasal obstruction, recurrent episodes of epistaxis, and non-specific manifestations like sinusitis and headaches. [3]

In this case report, we present a detailed examination of a patient diagnosed with glomangiopericytoma originating from the nasal cavity and paranasal sinuses, shedding light on the clinical manifestations, diagnostic challenges, and management strategies.

Case Report

A female in her 70's presented with complaints of recurrent episodes of bleeding from the left nasal cavity for 8 months. There was a history of recurrent episodes of headaches and left nasal obstruction for 6 months. There was no history of post-nasal drip or nasal discharge. No other significant ENT specific complaints. DNE was done and showed a polypoidal vascular mass filling the left nasal cavity. (*Fig 1*)

CECT-PNS was doneand showed fairly defined

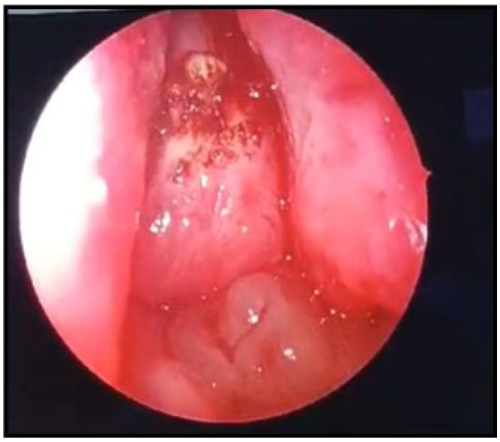


Fig 1: Diagnostic Nasal Endoscopy Showed a Polypoidal Vascular Mass Filling the Left Nasal Cavity.

From : Department of ENT, and *Pathology, Saveetha Medical College and Hospital, Saveetha Institute of Medical and Technical Sciences (SIMATS), Chennai, India.

Correspondence to:Dr Shyam Sudhakar Sudarsan Associate Professor, Department of ENT, Saveetha Medical College and Hospital, Saveetha Institute of Medical and Technical Sciences (SIMATS), Chennai, India.

Manuscript Recieved: 21.3.24; **Revision Accepted** 15.6.2024

Published online First: 10,Oct 2024

Open Access at : http://journal.jkscience.org

Cite this article as: Prithviraj SM, Sudarsan SS, Ravi DR,Priyadarshini SG. Glomangiopericytoma — A Rare Sino-nasal Tumour. JK Science 2024; 26(4):250-3

heterogeneous soft tissue dense focus seen in the left nasal cavitymedially limited by the nasal septum and perpendicular plate of ethmoid bone with bowing of nasal septum to the right, laterally - seen abutting and causing thinning, bony remodelling of the medial wall of the left maxillary sinus, posteriorly - seen extending beyond the left choana and causing narrowing of the left side nasopharyngeal airway, inferiorly seen limited by the bony hard palate, however no evidence of erosive changes, superiorly - seen extending into the left ethmoid air cells, and causing thinning of the ethmoidal septa with no evidence of bony erosive changes and anteriorly - not extending into the anterior nasal cavity. (***Fig 2, 3***)

A biopsy was done from the mass which showed features suggestive ofLeft sinonasal mass- Spindle cell carcinoma/Vascular neoplasm. The patient then underwent debulking of the tumor, followed by FESS with medial maxillectomy, and the tissue was sent for histopathological examination.

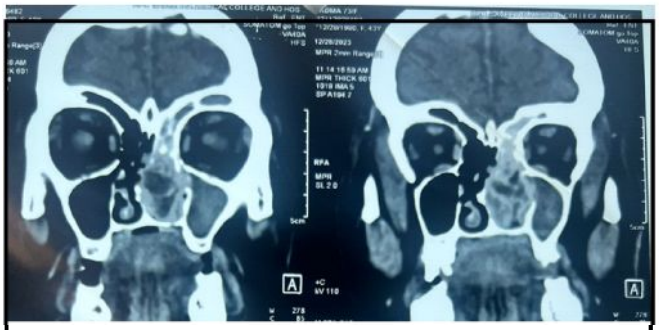


Figure 2

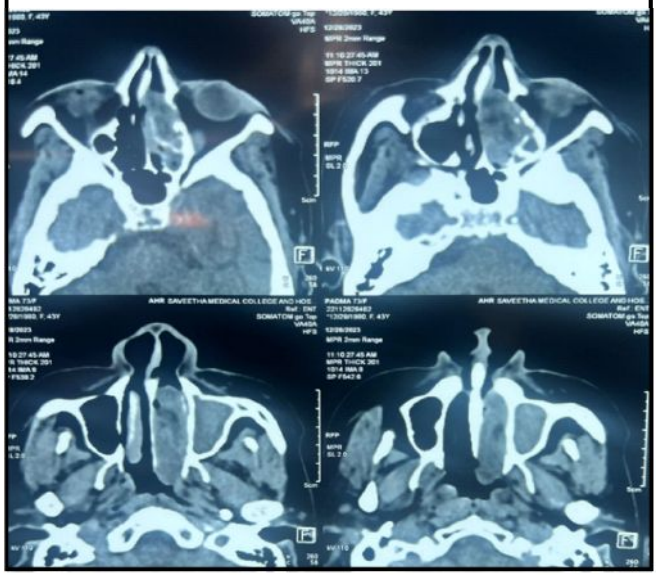


Fig 3: CECT-PNS showed fairly defined heterogeneous soft tissue dense focus seen in the left nasal cavity medially limited by the nasal septum and perpendicular plate of ethmoid bone with bowing of nasal septum to the right.

HPEshowed sections of multiple fragments of tissue lined by ciliated pseudostratified columnar epithelium with underlying stroma showing oedema and minimal lymphoplasmacytic infiltrates. (***Fig 4a & 4b***)

The underlying tissue shows a neoplasm composed of fairly uniform to mildly pleomorphic cells with scant to moderate eosinophilic cytoplasm and oval to elongated spindle nuclei with numerous capillaries interspersed within the tumor. Focal areas show minimal perivascular hyalinization around a few blood vessels. The tumor cells extend superficially up to the subepithelial stroma with a clear Grenz zone and extend into the deeper adipose tissue. Focal areas show spicules of bone. No mitosis or necrosis was noted.

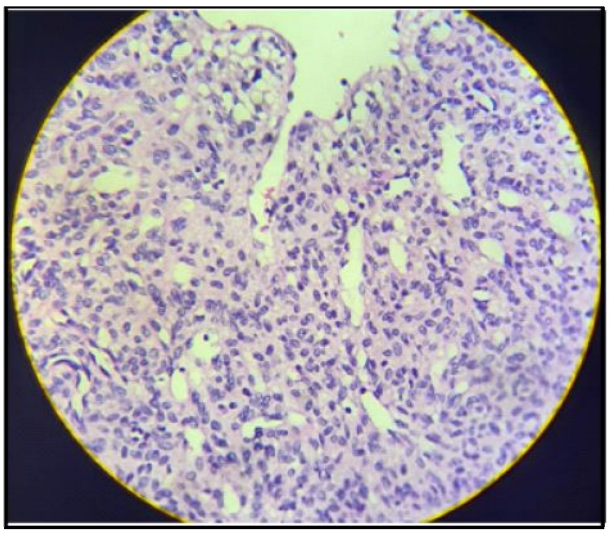
Advised Immunohistochemistry (Ki67, pan-cytokeratin, CD34, SMA, and Beta-catenin) for further evaluation and categorization, which showed 1) SMA - Positive. Percentage of cells with positivity: 50 % Average intensity of staining: Strong Pattern of staining: Cytoplasmic. (***Fig 5a)*** (2) CD34 - Negative in tumor cells. (***Fig 5b***) (3) Ki67 labeling index - Percentage of cells with positivity in tumor cells: <5%. Average intensity of staining: Strong. Pattern of staining: Nuclear. (4) Beta-catenin - Negative for nuclear staining. (5) Cyclin D1 - Focally (15-20 %) moderate positivity in tumor cells.

On 3 months follow-up, the patient is asymptomatic at present and has been relieved of her symptoms. Repeat diagnostic nasal endoscopy showed no signs of recurrence.

Discussion

GPC specifically arises from pericytes surrounding capillaries and accounts for a minute fraction of sinonasal tumors, making up less than 0.5% of cases. Classified as a borderline and low-malignant-potential tumor by the World Health Organization in 2005, GPC poses a unique clinical challenge due to its rarity and distinctive characteristics.[4]

GPCs with low malignant potential are typically confined to the nasal cavity, with infrequent extension into the paranasal sinuses and skull base. In cases where paranasal sinus involvement occurs, the ethmoid and sphenoid sinuses are the most commonly affected. The clinical symptoms arise from the mass effect and include headache, nasal congestion, epistaxis, and sinusitis. [5,6]



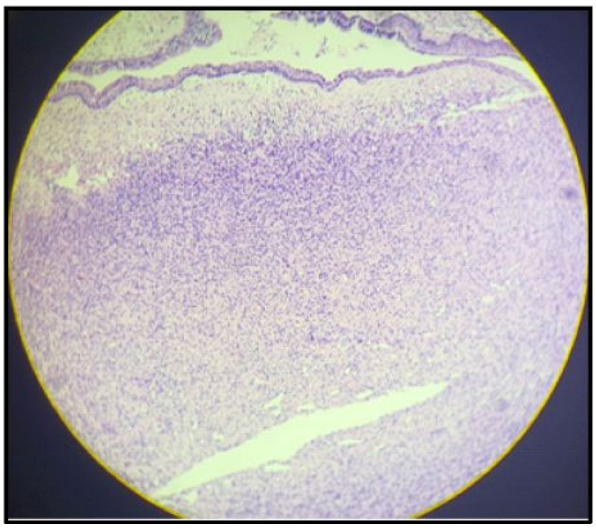
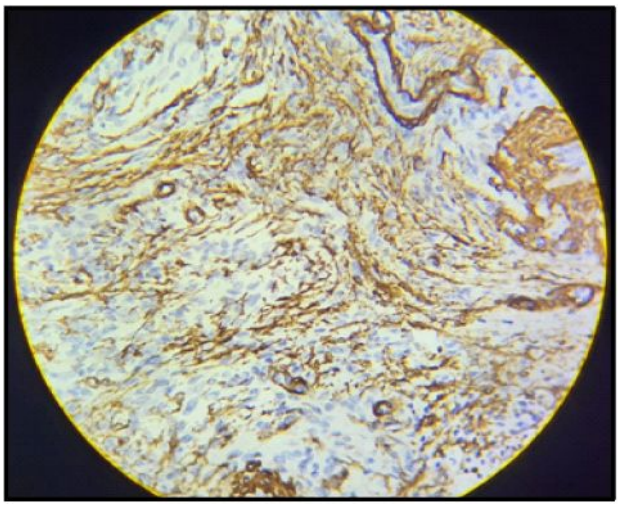


Fig 4a and 4b- HPE showed sections of multiple fragments of tissue lined by ciliated pseudostratified columnar epithelium with underlying stroma showing oedema and minimal lymphoplasmacytic infiltrates.



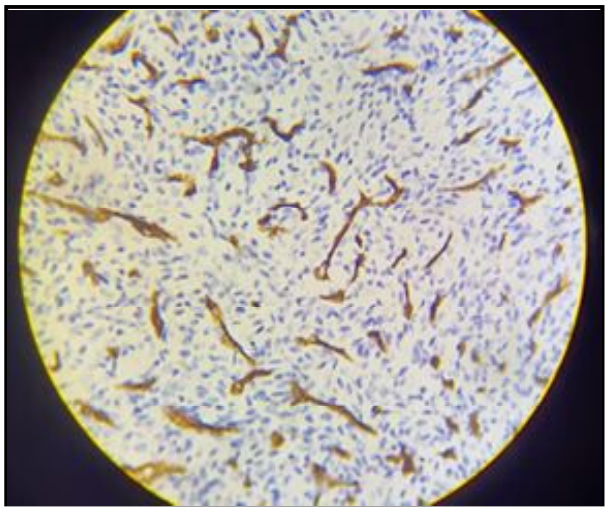


Fig 5a and 5b- Immunohistochemistryshowed SMA - Positive. Percentage of cells with positivity: 50 % Average intensity of staining: Strong Pattern of staining: Cytoplasmic. (Fig 5a) and CD34 - Negative in tumor cells. (Fig 5b)

The potential risk factors, including trauma, corticosteroid use, hypertension, and pregnancy, warrant further investigation to elucidate the underlying etiology[5]

The differential diagnosis for a sino-nasal mass includes glomus tumors, solitary fibrous tumors, lobular capillary haemangiomas, angiofibromas, myoepitheliomas, leiomyomas, olfactory neuroblastomas and rarely leiomyosarcomas and fibrosarcomas.[6]

Investigations include Nasal Endoscopy, CT, and MRI Imaging to assess the tumor extent, size and characteristics of the tumor, followed by Histopathological examination to confirm the diagnosis.The diagnostic marker of GPC includes nuclear staining for beta-catenin [3,6].

In immunohistochemical analysis, these cells exhibit positive staining for actin, vimentin, and beta-catenin, while displaying negative staining for desmin, keratin, and S10. [7]

The treatment of choice for GPC is complete surgical resection, as most GPCs are relatively resistant to chemotherapy and radiotherapy.[8,9] Though claims to be

of low malignant potential, GPC tends to recur.[10]

While intranasal endoscopic treatment is more efficient than more traditional open approaches, the presence of highly vascularized or sizable tumors can pose challenges. To address this, preoperative embolization may be considered to reduce bleeding volume during surgery, particularly in cases involving large or highly vascular tumors.

Postoperative care necessitates ongoing surveillance, involving periodic nasal endoscopy, to monitor for potential local tumor recurrence. Additionally, the inclusion of MRI and CT scans at regular intervals is advised as part of the long-term management strategy. [8,10]

Conclusion

Glomangiopericytoma are rare mesenchymal tumour arising from the nasal cavity and paranasal sinuses. Though claims to be low malignant potensial, GPC tends to recur Angiography is strongly recommended to identify the primary vascular feeder of the tumor, aiming to minimize intraoperative bleeding. Additionally, histopathological and immunohistochemical analyses play a crucial role in determining the tumor's characteristics, aiding in the selection of an appropriate treatment strategy. The treatment of choice involves complete surgical resection, followed by regular clinical monitoring and extended follow-up periods to detect and address any potential recurrence.

References

1. Ghaloo SK, Dhanani R, Pasha HA, Wasif M, Fatima S, Ikram M. Glomangiopericytoma: A rare tumour of sinonasal cavity. J Pak Med Assoc 2020 ;70(12(B)):2469-71.
2. Misio³ek M, Namyslowski G, Scierski W, Czecior E, Lisowska G, Lange D. Klebczako-obloniaknosaizatokprzynosowych—opisprzypadku [Sinonasal glomangiopericytoma—case report]. Otolaryngol Pol 2007;61(6):987-9.
3. Kono M, Bandoh N, Matsuoka R, Goto T, Akahane T, Kato Y, Nakano H, Yamaguchi T, Harabuchi Y, Nishihara H. Glomangiopericytoma of the Nasal Cavity with CTNNB1 p.S37C Mutation: A Case Report and Literature Review. Head Neck Pathol 2019 ;13(3):298-303.
4. Higashi K, Nakaya K, Watanabe M, Ikeda R, Suzuki T, Oshima T, Kobayashi T. Glomangiopericytoma of the nasal cavity. Auris Nasus Larynx 2011 ;38(3):415-7.
5. Moussaoui ZNE, Najjar ZA, Diab N, Saker Z, Choukr H, Aoude AK, et al. Clinical and histopathological findings of a rare sinonasal glomangiopericytoma. Autops Case Rep 2023 10;13:e2023424.
6. Kazi AA, McDougal EM, Howell JB, Schuman TA, Nord RS. Glomangiopericytoma: a case series with review of literature. Braz J Otorhinolaryngol 2022 ;88(5):817-20.
7. Sheikh S, Sarwar F, Khan NU, Khan MS. Endonasal endoscopic laser-assisted resection of septal glomangiopericytoma. BMJ Case Rep 2018 ;2018:bcr2017223752.
8. Al-Jobory YM, Pan Z, Manes RP, Omay SB, Ikuta I. Sinonasal Glomangiopericytoma: Review of Imaging Appearance and Clinical Management Update for a Rare Sinonasal Neoplasm. Yale J Biol Med 2021 ;94(4):593-7.
9. Almarri FK, Alnatheer AM, Abuhaimed MK, Albathi AA, Alqahtani AQ, Tatwani T. A rare case of glomangiopericytoma in the nasal cavity: A case report in light of recent literature. Ann Med Surg (Lond) 2022 28;77:103685.
10. Gordon AJ, Papazian MR, Chow M, Patel A, Placantonakis DG, Lieberman S, Givi B. Sinonasal Glomangiopericytoma with Prolonged Postsurgical Follow-Up. J Neurol Surg Rep 2022 ;83(3):e87-e89.