Head and Neck Pathology (2025) 19:101
https://doi.org/10.1007/s12105-025-01810-1

IMAGE

Glomangiopericytoma of the Infraorbital Region

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Received: 16 May 2025 / Accepted: 3 June 2025 / Published online: 9 August 2025

Abstract

Background Glomangiopericytomas are rare spindle cell neoplasms that typically arise within the sinonasal tract and exhibit borderline to low malignant potential. While unusual, occurrences of this tumor in other regions of the head and neck have been reported, such as the deep neck spaces, larynx, tongue, and middle ear.

Case Presentation A 44-year-old male presented with a one-year history of left infraorbital swelling, enlarging over two months and occasionally pulsating with discomfort. Cross-sectional imaging revealed a 1.5 cm x 0.8 cm x 1.7 cm homogeneously-enhancing mass in the left medial infraorbital region eroding the adjacent maxillary sinus wall, suggestive of an infraorbital nerve schwannoma or hemangioma. The tumor was excised via a lateral rhinotomy approach with anterior maxillectomy. The final pathological diagnosis was a glomangiopericytoma.

Conclusion Although typically localized to the sinonasal tract, glomangiopericytomas may arise in unusual head and neck locations such as the infraorbital region. It is important to distinguish these tumors from related mesenchymal tumors such as solitary fibrous tumors and myopericytomas due to differences in their biology and malignant potential.

Keywords Glomangiopericytoma · Infraorbital tumor · Spindle cell tumor · Mesenchymal tumor

Case Report

A 44-year-old male with underlying ischemic heart disease presented with a one-year history of left infraorbital swelling, enlarging over two months and occasionally pulsating with discomfort. There were no nasal symptoms. Examination revealed a vague, pulsatile, soft-to-firm left infraorbital mass measuring 1 cm x 2 cm (Fig. 1A and B). Other physical examination findings, including nasoendoscopy, and blood investigations were unremarkable. Contrasted tomography revealed a 1.5 cm x 0.8 cm x 1.7 cm homogenously enhancing well-defined mass in the left medial infraorbital region eroding the adjacent maxillary sinus wall (Fig. 2A). The mass had intermediate signal on T1-weighted magnetic resonance imaging (MRI) and heterogeneously high signal on T2-weighted MRI with gadolinium enhancement (Fig. 2B). The possible differential diagnoses at that time included infraorbital nerve schwannoma or hemangioma.

As the mass was pulsatile, an in-office biopsy was not performed. It was surgically excised via a lateral rhinotomy approach with anterior maxillectomy. The tumor appeared lobulated, reddish, and well-encapsulated, overlying the left facial artery (Fig. 2C and D). Histopathological examination of the tumor revealed monomorphic spindle cells arranged in fascicles and whorls, associated with thin branching staghorn-like and medium-sized vessels with perivascular hyalinization (Fig. 3A). The spindle cells had elongated hyperchromatic nuclei with inconspicuous nucleoli and scanty cytoplasm (Fig. 3B). The mitotic figures were rare and there was no necrosis.

On immunohistochemistry, the tumor cells were positive for markers of perivascular myoid differentiation (CD99, Cyclin D1, smooth muscle actin, and β-catenin), and negative for markers of vascular (CD34 and ERG), epithelial

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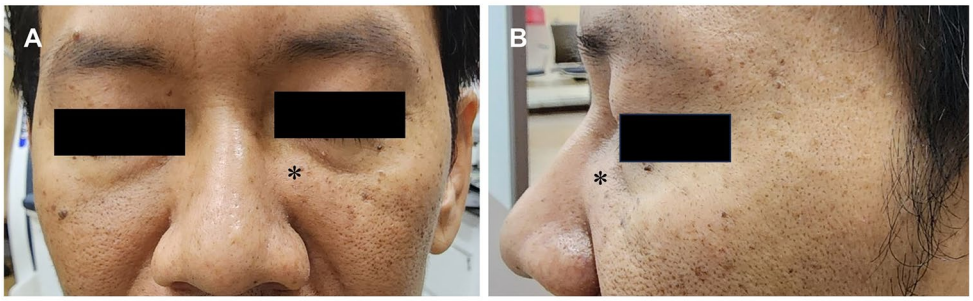

Fig. 1 Clinical photograph of the case showing the left infraorbital mass (black asterisk) (**A.** Frontal view, **B.** Lateral view)

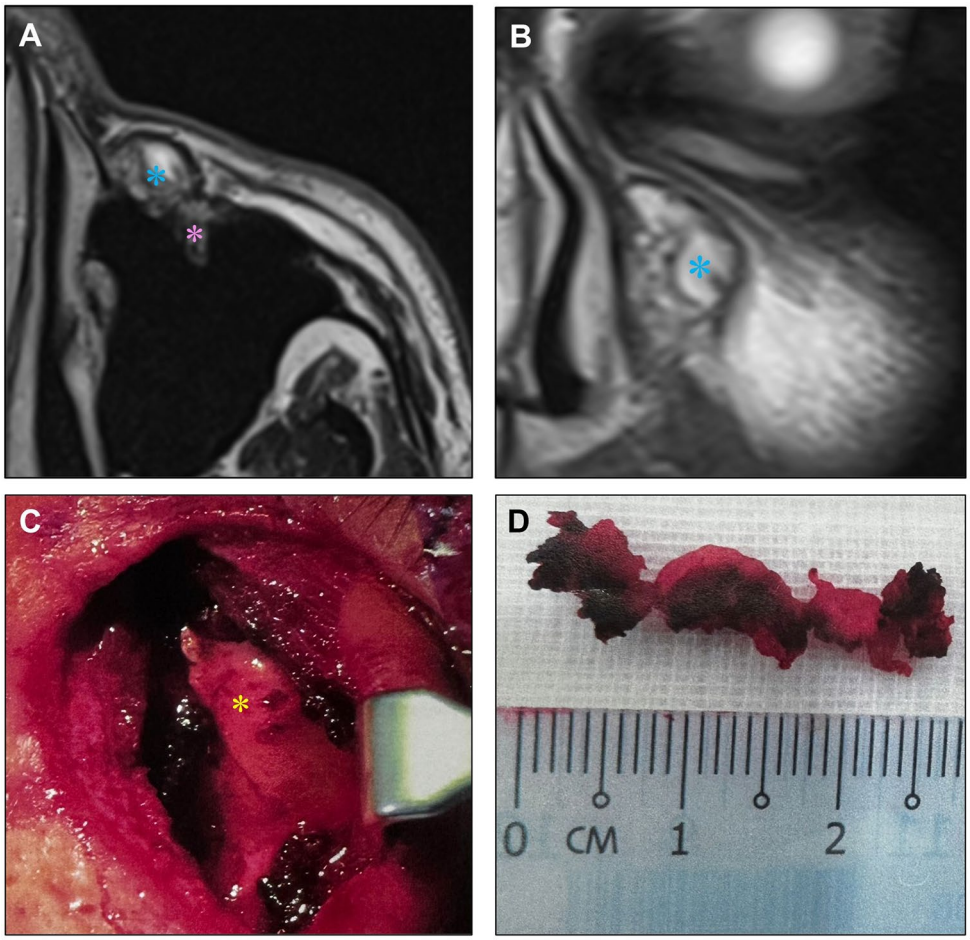

Fig. 2 **(A)** Axial view of the CT scan showing the left infraorbital tumor (blue asterisk) with erosion of the adjacent maxillary sinus wall (pink asterisk). **(B)** Coronal view of the T2-weighted MRI scan showing heterogeneous intensity and gadolinium enhancement of the tumor (blue asterisk). **(C)** Intraoperative photograph of the infraorbital tumor in situ (yellow asterisk). **(D)** Intraoperative photograph of the excised infraorbital tumor specimen

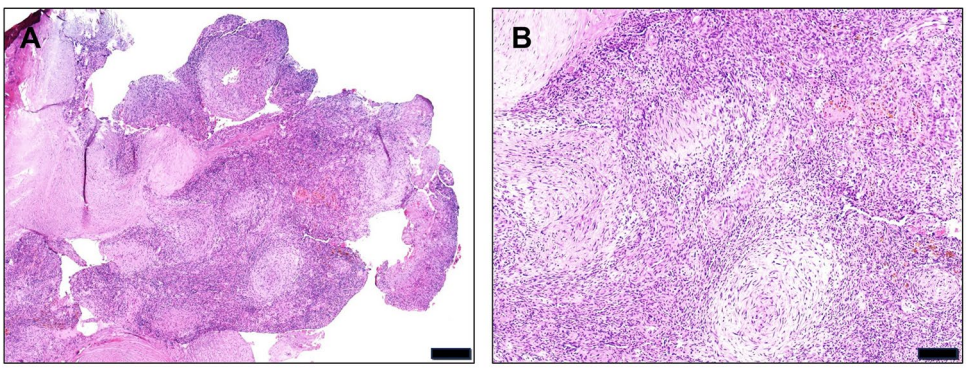

Fig. 3 **(A)** Hematoxylin and eosin (H&E) stain of the tumor showing spindle cells arranged in fascicles and whorls. There were thin branching staghorn-like medium-sized vessels with perivascular hyalinization (original magnification x 4). **(B)** The spindle cells had elongated hyperchromatic nuclei with inconspicuous nucleoli and scanty cytoplasm. Mitotic figures were hardly seen and there was no necrosis (original magnification x 10). Scale bar: 500 µm

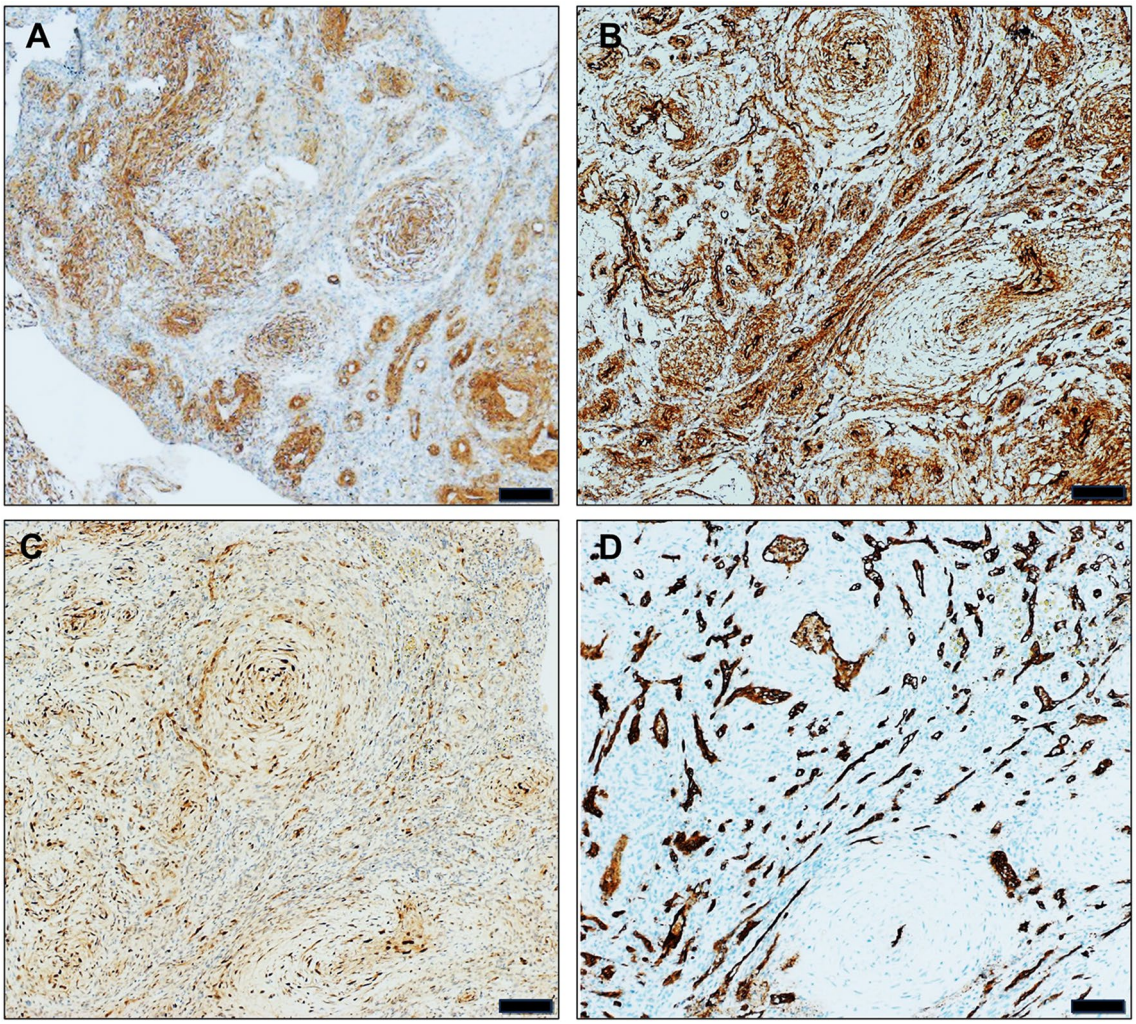

Fig. 4 Immunohistochemical staining of the tumor: **(A)** The spindle cells had positive immunoreaction with smooth muscle actin, suggestive of perivascular myoid differentiation (original magnification x 10). **(B)** Positive nuclear immunoreaction for β-catenin, a key feature of GPC signifying activation of WNT signaling pathway (original magnification x 10). **(C)** Positive immunoreaction for cyclin D1, a cell cycle regulator that is overexpressed following WNT signaling (original magnification x 10). **(D)** CD34, a marker of vascular endothelial cells, stained the thin branching staghorn-like medium-sized vessels within the tumor but not the spindle cells (original magnification x 10). Scale bars: 500 μm

(EMA), and Schwann (S100) cell differentiation, establishing the diagnosis of glomangiopericytoma (Figs. 4A– D). The patient remained well on subsequent follow-up with no evidence of recurrence.

Glomangiopericytoma (GPC) is a rare spindle cell neoplasm with perivascular myoid phenotype and borderline to low malignant potential [1]. In contrast to mesenchymal tumors such as myopericytoma (MPC) and solitary fibrous tumor (SFT) which can occur throughout the body, GPCs arise primarily in the sinonasal tract causing nasal obstruction, sinusitis, headache, and epistaxis. Nonetheless, this tumor has also been reported in other head and neck sites such as the deep neck spaces, larynx, and middle ear [2, 3]. A hallmark of GPC is their overexpression of cyclin D1 and nuclear expression of β-catenin secondary to *CTNNB1* mutations and WNT pathway alterations [1].

In our case, the tumor may have arisen from perivascular myoid cells of the infraorbital facial artery. Although certain perivascular myoid tumors such as MPCs have been reported in the facial soft tissue, they are distinct from GPCs, being negative for β-catenin [4]. Intriguingly, GPC-like areas have been detected in other perivascular myoid tumors such as glomus tumors, myofibromas, and angioleiomyomas [5]. It is plausible that these tumors are part of a spectrum of perivascular cell differentiation along the smooth muscle, pericyte, or glomus cell lineages.

It is important to recognize that GPCs can arise outside the sinonasal tract and to distinguish them from MPCs and SFTs. GPCs have borderline or low malignant potential

whereas MPCs are usually benign and SFTs have variable prognoses depending on their clinical and histological features [1]. Long term-outcomes of a completely resected GPC is good with an estimated 5-year survival exceeding 90% [1].

Acknowledgements We thank the patient for permission to publish this information. We are also grateful to LabLink Medical Laboratory, Kuala Lumpur, Malaysia for preparing and scanning the histopathological and immunohistochemical slides.

Author Contributions Writing– Original Draft: Shen-Han Lee; Writing– Review & Editing: Shen-Han Lee, Mohamad Azizul Fitri Khalid, Amirozi Ahmad, Noorjehan Omar, Ramiza Ramza Ramli; Resources: Mohamad Azizul Fitri Khalid, Amirozi Ahmad, Noorjehan Omar; Visualization: Shen-Han Lee, Noorjehan Omar; Conceptualization: Shen-Han Lee, Mohamad Azizul Fitri Khalid, Ramiza Ramza Ramli; Supervision: Ramiza Ramza Ramli.

Funding This work received no specific grant from any funding agency in the public, commercial, or non-profit sectors.

Data Availability No datasets were generated or analysed during the current study.

Declarations

Ethical Approval Institutional review board approval was not required as this manuscript describes a single anonymous retrospective case report. The study was performed in accordance with the ethical standards of the 1964 Helsinki Declaration and its later amendments, or comparable ethical standards.

Patient Consent Written informed consent was obtained from the patient for publication of the details of their clinical details and clinical images.

Competing Interests The authors declare no competing interests.

References

1. Thompson LDR, Jun SY, Lai CK (2023) Sinonasal glomangiopericytoma. In: WHO Classification of Tumours Editorial Board, ed. Head and Neck Tumours (WHO Classification of Tumours Series), IARC Press, Lyon, 9(5):90-92
2. Suh CH, Lee JH, Lee MK et al (2020) CT and MRI findings of glomangiopericytoma in the head and neck: case series study and systematic review. AJNR Am J Neuroradiol 41(1):155. https://doi.org/10.3174/ajnr.A6336
3. Liu GS, Berry GJ, Soltys SG, Blevins NH (2024) Glomangiopericytoma presenting as a middle ear mass. Laryngoscope 134(3):1426–1430. https://doi.org/10.1002/lary.30987
4. Kim EK, Lee JH, Kim SY, Kim GM (2013) Myopericytoma of the facial cheek. Ann Dermatol 25(1):122–124. https://doi.org/10.5021/AD.2013.25.1.122
5. Granter SR, Badizadegan K, Fletcher CDM (1998) Myofibromatosis in adults, glomangiopericytoma, and myopericytoma: a spectrum of tumors showing perivascular myoid differentiation. Am J Surg Pathol 22(5):513–525. https://doi.org/10.1097/00000478-199805000-00001