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Glomangiopericytoma in a 17-Year-Old Filipino Woman: A Case Report

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ABSTRACT

Objective: To report a rare case of glomangiopericytoma in a teenage Filipino woman and our rationale for a transpalatal approach in its surgical management.

Methods:

Design: Case Report
Setting: Tertiary Government Training Hospital
Patient: One

Results: A 17-year-old woman consulted due to a 10-month history of an intranasal mass. CT scan revealed a well-delineated contrast-enhancing soft tissue density occupying the entire right nasal cavity which extended anteriorly to the right nasal vestibule, posteriorly to the nasopharyngeal area with noted erosions of the medial wall of the right maxillary sinus and part of the bony nasal septum. Excision of mass via transpalatal approach achieved grossly complete removal, with no recurrence observed at one year.

Conclusion: Glomangiopericytoma is a rare neoplasm of the nasal cavity and paranasal sinuses that usually has a borderline low malignant potential. This case reports its occurrence in a young Filipino woman and suggests that the transpalatal approach may be an alternative in carefully selected cases as it allowed apparently complete surgical removal with an initially favorable outcome.

Keywords: *glomangiopericytoma; hemangiopericytoma-like tumor; sinonasal perivascular neoplasms; transpalatal approach*

Glomangiopericytoma is a rare sinonasal tumor of the head and neck accounting for less than 0.5% of all sinonasal tumors.[1,2,3] The clinical presentation is often nonspecific, most commonly including recurrent epistaxis, nasal obstruction and anosmia. Because of these features, the tumor may mimic other vascular or spindle cell neoplasms, complicating timely diagnosis. Most reported cases occur in middle-aged to elderly patients with only a few documented in the pediatric and adolescent population.[4] A search of HERDIN Plus, MEDLINE (PubMed), and Google Scholar using the terms "*glomangiopericytoma Philippines," "hemangiopericytoma-like tumor Philippines," and "sinonasal perivascular tumors Philippines"* yielded only two published case reports, both of which primarily focused on the pathologic features of glomangiopericytoma.[5,6] We report a case of glomangiopericytoma in a young Filipino woman and describe the rationale for selecting a transpalatal approach for its surgical management, an intervention not previously reported in the literature.

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CASE REPORT

A 17-year-old woman with no known comorbidities, presented at *Ospital ng Maynila* Medical Center with a 10-month history of recurrent epistaxis and associated nasal obstruction of the right nasal cavity, and conductive anosmia. This was followed by a gradually enlarging soft, non-painful bulge over the right nasofacial groove with associated progression of nasal obstruction now involving the left nasal cavity. On examination, there was a firm, well circumscribed intranasal mass on the right, with superficial crusting and pin-point bleeding on manipulation. The nasal septum was deviated leftward, obliterating the left nasal cavity due to mass effect. *(Figure 1)*

Transillumination was negative for the right maxillary sinus and a bulge on the right nasofacial groove was appreciated. Otoscopic and ophthalmologic examinations were unremarkable. Computed tomography (CT) imaging revealed a well-delineated contrast-enhancing soft tissue density occupying the entire right nasal cavity which extended anteriorly to the right nasal vestibule and extended posteriorly to the nasopharyngeal area with erosions of the medial wall of the right maxillary sinus and part of the bony nasal septum.



Figure 1. Patient profile showing intranasal mass producing a leftward deviation of the nasal septum and obliteration of the left nasal cavity due to mass effect. Photos published with permission.

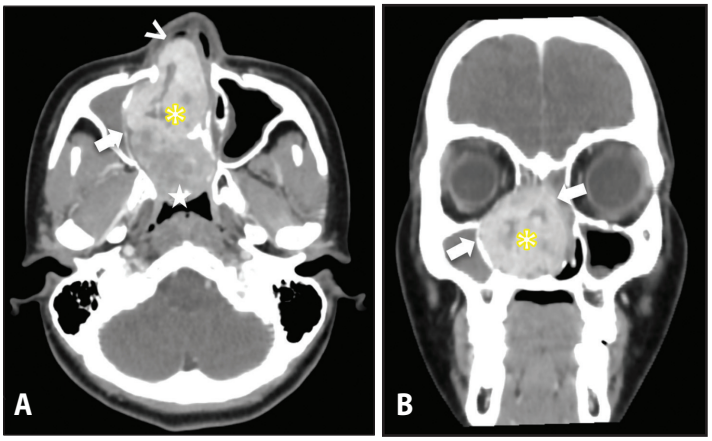


Figure 2. Contrast-enhanced CT scans **A.** Axial and **B.** Coronal sections showing a solitary, well-defined, lobulated, contrast-enhancing mass occupying the right nasal cavity (asterisk) extending anteriorly to the right nasal vestibule (arrowhead) and posteriorly extending to the nasopharyngeal area (star) with erosions of the medial wall of the right maxillary sinus and part of the bony nasal septum (arrows).

The nasal septum was deviated to the contralateral side, already abutting the left lateral nasal wall. There was appreciable thinning and upward displacement of the orbital wall with no erosions seen. The hard palate was likewise intact. *(Figure 2)* Punch biopsy revealed spindle cell neoplasm. Immunohistochemical staining showed immunoreactivity on SMA, negative on CD34, STAT6, Caldesmon, and CK fitting the immunohistochemical profile of a sinonasal glomangiopericytoma. *(Figure 3)*

Endoscopic removal was deemed technically challenging due to the limited surgical corridor and difficulty in achieving complete exposure of the tumor margins. An external facial approach, although feasible, was avoided to minimize morbidity and cosmetic consequences. The patient underwent excision of mass via transpalatal approach. This approach was selected after discussion with the patient and her family, taking into consideration the size of the lesion, its posterior extension to the nasopharynx, and its attachment to the posterior bony septum.

A U-shaped palatal mucoperiosteal flap was elevated to expose the posterior aspect of the hard palate and nasal floor. Limited posterior palatal bone resection using a Kerrison rongeur and osteotome provided direct access to the posterior nasal cavity and nasopharynx. *(Figure 4)* This wide surgical exposure allowed *en bloc* removal of the mass under direct visualization. Intraoperative findings revealed a submucosal, lobulated, friable 8 x 5 x 4 cm mass occupying the entire right nasal fossa, reaching the vestibule anteriorly, the nasopharynx posteriorly, displacing the nasal septum to the left and the medial wall of the maxillary sinus to the right. Attachment was identified at the posterior vomer. Following resection, the palatal flap was repositioned and closed primarily using interrupted sutures. Nasal packing was placed at the end of the procedure and removed on the fifth postoperative day.

Final histopathologic results revealed respiratory epithelium with underlying sheets of spindle-shaped tumor cells with minimal atypia, arranged around prominent thin-walled, staghorn-shaped vessels. These histomorphologic features were consistent with a sinonasal glomangiopericytoma. The patient was discharged in a week after an uneventful post-operative course. Due to the then ongoing COVID-19 pandemic, the patient was unable to travel for physical follow-up; hence, regular virtual follow-up consults were conducted. The patient was asymptomatic with no perceived recurrence of the mass. Repeat contrast-enhanced CT scan was requested but was not available in her province. The patient was then lost to follow-up one-year post-operation. *(Figure 5)*

DISCUSSION

We reported a rare case of glomangiopericytoma in a teenage Filipino woman and our rationale for a transpalatal approach in its

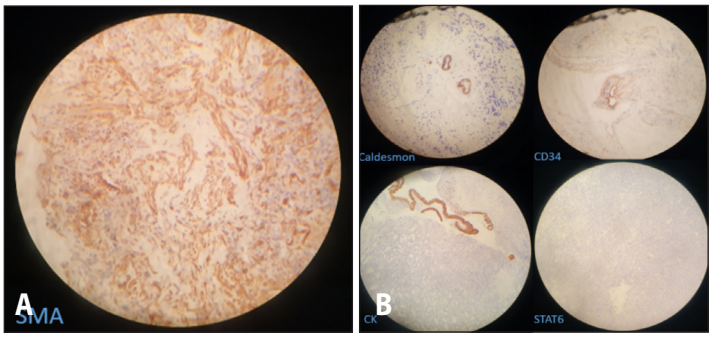


Figure 3. Immunohistochemical stains **A.** Alpha Smooth Muscle Actin (SMA) showing immunoreactivity for actin; and **B.** negative reactivity for Caldesmon, CK, CD34 and STAT6

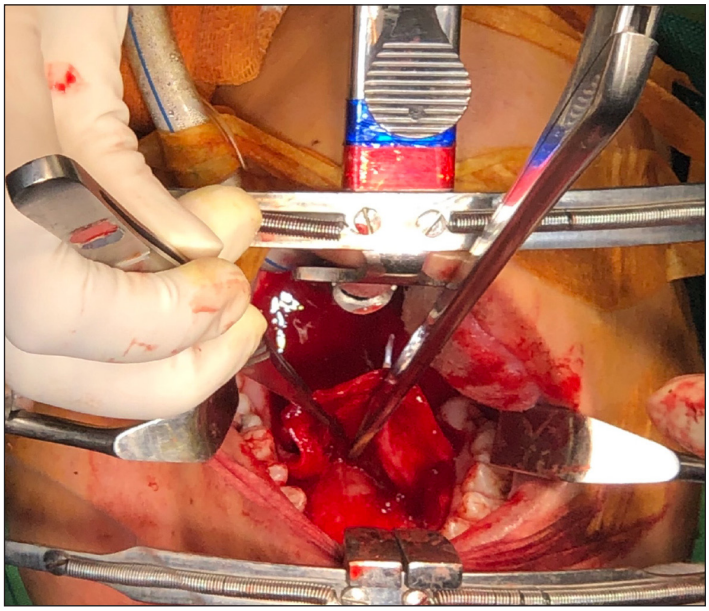


Figure 4. Posterior palatal bone resection performed using a Kerrison rongeur



Figure 5. Patient profile one-year post-operation. Photos published with permission.

surgical management. According to the World Health Organization 2005 classification of head and neck tumors, glomangiopericytoma is a term used to describe sinonasal tumors demonstrating a perivascular myoid differentiation.[7] It has a slight female predilection, usually found in the middle-aged and peaks in the 6th or 7th decade of life, although individuals of any age may be affected.[1,2,4,8] To date, the etiology is still

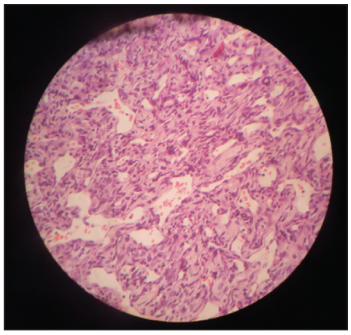


Figure 6. Patient histopathologic slide showing packed spindle cells arranged in short fascicles surrounding a thin-walled blood vessel (Hematoxylin-Eosin, 40x magnification). This histologic architecture is characteristic of glomangiopericytoma.

unknown but several risk factors (such as past trauma, hypertension, pregnancy, and chronic use of corticosteroids) may predispose to its development.[2,3,4,8] Our patient conformed with the usual gender predilection of this tumor– being a young female; but had no risk factors.

As observed in our case, patients with glomangiopericytoma usually present with unilateral obstruction and/or recurrent epistaxis. Rarely, they present with dyspnea, anosmia, pain, headache, visual disturbances, serous otitis media, proptosis and infraorbital anesthesia.[1,2,4,8,9] The nasal septum is the most frequent site of origin, followed by the paranasal sinuses.[9] Grossly, glomangiopericytoma appears as a polypoid, beefy red to greyish pink, soft, fleshy to friable, edematous to hemorrhagic tumor that bleeds upon palpation.[1,2,3] The average size is around 3 cm but may grow up to 8 cm.[3] Our patient presented with the common signs and symptoms of glomangiopericytoma– nasal obstruction and epistaxis. Upon evaluation, the mass also bled minimally on manipulation, measuring 8 cm in its largest diameter, which ranked on the larger side of the usual size range.

The histopathologic findings were consistent with sinonasal glomangiopericytoma, characterized by hematoxylin-eosin staining subepithelial tumor that is circumscribed yet unencapsulated, uniform, elongated to oval, with round to spindle-shaped nuclei and slightly eosinophilic cytoplasm. It is usually surrounded by normal respiratory epithelium and has tightly packed small cells that form short fascicles with storiform, whorled or palisading patterns, with many thin-walled, branching, staghorn vascular channels.[1,2,3] *(Figure 6)* These vessels may be surrounded by a small cuff of cellular fibrosis. Prominent perivascular hyalinization is a feature.[1,8,10] Immunohistochemistry confirmed glomangiopericytoma by SMA positivity and CD34/STAT6 negativity.[1,2,3,8,10,11] These histopathologic characteristics were present in our case.

The accepted gold standard for the management of glomangiopericytoma is resection with tumor-free margins.[12] This may be achieved via endoscopic endonasal removal as the best less-invasive tumor resection method.[1] Other reported options include

medial maxillectomy, combined endoscopic surgery and Caldwell-Luc, combined external and endoscopic approach, and purely external approach.[9] Contrast-enhanced imaging is a basis for preoperative planning for the approach, extent of resection and need for preoperative embolization.[3,13] In cases where complete surgical excision is not possible, adjuvant and palliative radiotherapy/chemotherapy may be considered although this tumor tends to be radioresistant.[3,8] A preoperative biopsy is performed to establish the histopathologic diagnosis and confirm the vascular or perivascular nature of the lesion. This information is critical for surgical planning as it helps determine the safest and most appropriate approach, anticipate intraoperative bleeding, and define the extent of resection required for tumor clearance. By enabling precise preoperative planning, biopsy indirectly minimizes the risk of incomplete excision and subsequent recurrence. [4]

In this case, the patient underwent preoperative imaging and biopsy, and excision of mass via transpalatal approach. To our knowledge, based on an English-language search on HERDIN Plus, MEDLINE (PubMed), and Google Scholar using the terms "glomangiopericytoma," "sinonasal hemangiopericytoma," "sinonasal perivascular tumor," and transpalatal approach," no previous reports have described the use of a transpalatal approach for the management of glomangiopericytoma. Our choice of this approach was based on the tumor's size (8 x 5 x 4 cm), posterior extension, and firm attachment to the bony nasal septum. The transpalatal approach offered a wide, direct surgical field that allowed safe *en bloc* resection with excellent visualization and control of intraoperative bleeding. Compared to endoscopic resection, which is widely regarded as the preferred approach for smaller, accessible lesions due to its minimal invasiveness, the transpalatal approach provided superior access to the posterior nasal cavity and nasopharyngeal region in this patient. Furthermore, it minimized the risk of incomplete resection that may occur when working through a narrow endoscopic corridor with a large, broad-based tumor. When contrasted with external approaches such as lateral rhinotomy or maxillary swing approach, the transpalatal approach was advantageous because it avoided visible facial scarring and osteotomies, while still providing a wide surgical field. Thus, in carefully selected cases where tumor size, location, and attachment make endoscopic removal less optimal, the transpalatal approach remains a safe and effective alternative with low morbidity and favorable cosmetic outcomes.[14]

The final histopathologic results of the excised mass revealed positive tumor involvement on the anterior, posterior and inferior surgical margins. These positive margins corresponded to the anatomic limits of resection as the tumor's attachment was located along the medial aspect. In sinonasal glomangiopericytoma, the turbinate and nasal septum may be affected in isolation.[15] In this case, the tumor completely involved the nasal turbinate, leaving no residual normal respiratory epithelium at the corresponding surgical margins. Thus, the absence of normal respiratory mucosa reflected the anatomic extent and signifies intimate physical contact of the tumor to adjacent structures rather than true invasion or incomplete excision. Given these intraoperative findings, no adjuvant therapy was deemed necessary.

Prognosis following complete resection is highly favorable, with more than 95% of cases showing no recurrence and long-term survival rates remaining excellent.[10,12] While recurrence can still occur in a minority of cases, particularly after incomplete excision, vigilant follow-up remains essential.[1,2,3,4,8]

This report suggests the transpalatal approach as a possible surgical option for large, posteriorly extending sinonasal glomangiopericytomas, especially when endoscopic excision seems difficult. Though limited to a single patient with relatively short follow-up, it provides encouraging early evidence that this approach may achieve apparently complete resection with excellent short-term outcomes.

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