***Case report***

# Sinonasal Glomangiopericytoma: A Case Report and Literature Review

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**Abstract:**
Glomangiopericytoma is a rare vascular tumor arising from the pericytes surrounding capillaries, and accounts for less than 0.5% of all sinonasal tumors. The aim of this study is to describe our recent experience of sinonasal glomangiopericytoma and thoroughly review our collected data on the basis of age/sex, location, associated diseases, clinical symptoms, endoscopic findings, image findings, pathology, and treatment. We found 21 articles (19 case reports and 2 review articles) relevant to our search since 2005. The mean age was 58.05 years, ranging from 19 years to 86 years with a female to male ratio of 3:1 (15:5). The most frequent site of origin was the nasal septum followed by the paranasal sinuses. The most common symptom was nasal obstruction, followed by epistaxis, headache, facial pain or pressure, and anosmia in decreasing order. Computed tomography was the most common imaging technique used to evaluate sinonasal glomangiopericytoma which the most common mentioned CT finding was soft tissue density mass with enhancement. The most common treatment modality was complete surgical excision through transnasal endoscopic surgery.

**Keywords:** Glomaniopericytoma; Sinonasal; Symptoms; Computed tomography; Histopathology; Treatment

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## Introduction

Glomangiopericytoma is a rare vascular tumor arising from the pericytes surrounding capillaries, and accounts for less than 0.5% of all sinonasal tumors.[1]According to the World HealthOrganization 2005 classification of head and neck tumors, glomangiopericytoma isarecent proposed term describing the sinonasal tumors demonstratinga perivascular myoid differentiation andincludes the tumordescribed as sinonasal type hemangiopericytoma, andhemangiopericytoma-like tumor.[2]However, Asimakopoulos et al recently suggested that glomangiopericytomas and sinonasal hemangiopericytomas sometimes behave in a different clinical manner even if they have histologically similar characteristics and so, more case reports are needed to verify that point.[3]So, as we experienced a case of sinonasal glomangiopericytoma completely removed by endoscopic surgery, we tried to review all the literatures reported since 2005 and elucidate the clinical, radiological, pathological and therapeutic features of sinonasal glomnagiopericytoma.

## Case Report

A 68-year-old previously man with hypertension was referred to our department for proper evaluation and management of left-sided massive epistaxis which was stopped by totally occlusive nasal pack at local clinic. He denied a history of nasal trauma and surgery. Under the provisional diagnosis of posterior epistaxis, we tried to find the bleeding point and control the epistaxis. After packing removal, nasal endoscopy revealed a reddish broad-based bulging mass between the nasal septum and the middle turbinate (Fig. 1A & B).

The mass was smooth and friable in consistency and easily bled on touch.The mass originating from the nasal septum was completely removed with 5 mm safety margin and the periosteum through transnasal endoscopic approach. Histopathological results including immnohistochemical staining for smooth muscle actin were consistent with sinonasal glomangiopericytoma **(Fig. 1C & D).** His postoperative course was uneventful, and he had no evidence of a recurrence at the 2-year follow up.

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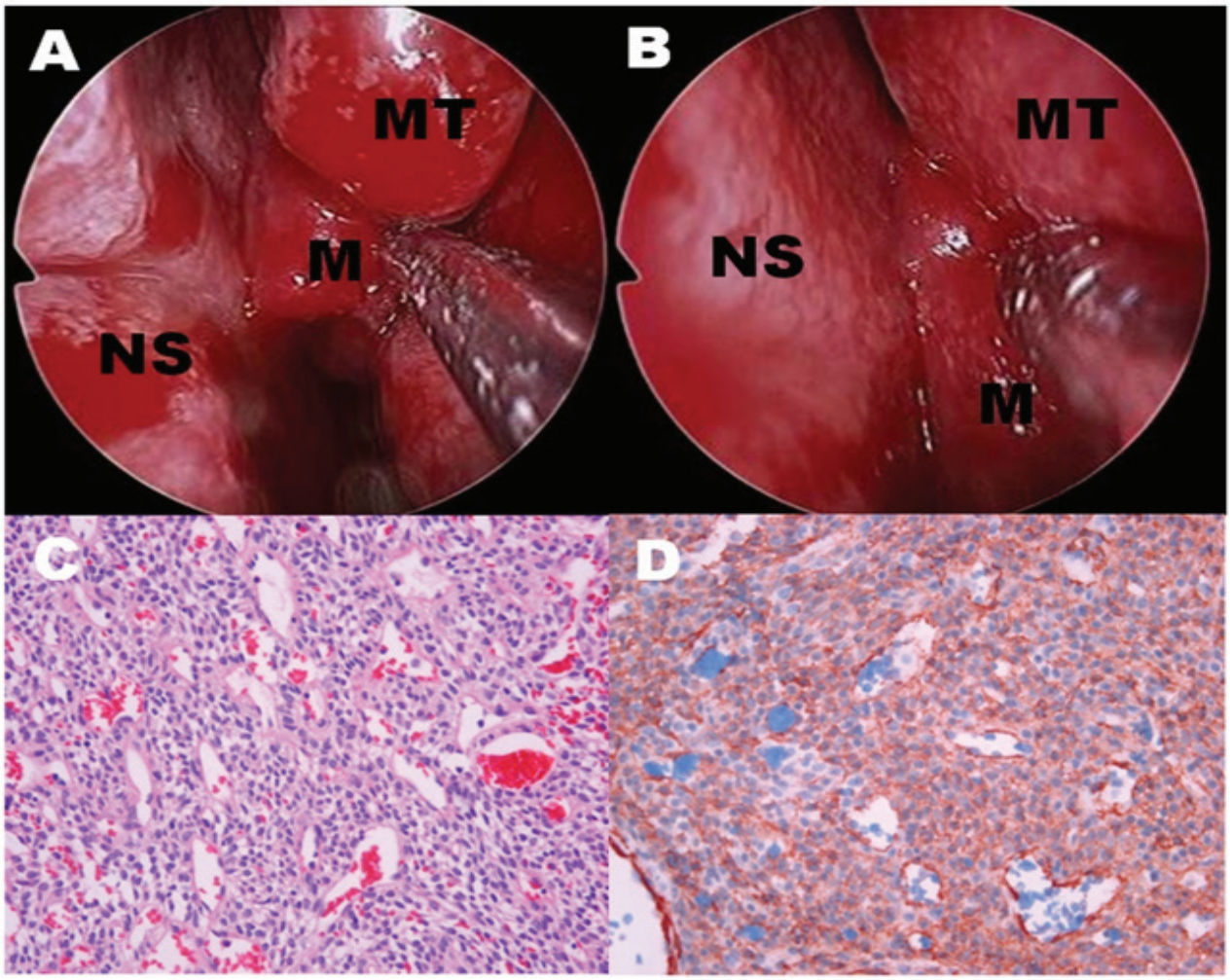


Figure 1. Nasal endoscopy revealed a reddish broad-based bulging mass between the nasal septum and the middle turbinate (A). The mass originating from the nasal septum (B) was smooth and friable in consistency and easily bled on touch. (NS: nasal septum, MT: middle turbinate, M: mass). Histopathological results including immnohistochemical staining for smooth muscle actin were consistent with sinonasal glomangiopericytoma (C & D, x200).

**Search Strategy of Literatures**

In order to review all of the available English-language literature, we searched the MEDLINE database using key words "Glomangiopericytoma" and "Sinonasal". We searched papers written in English only since 2005. Also, we only included the reports about sinonasal glomangiopericytoma and excluded the reports about sinonasal hemangiopericytoma and hemangiopericytoma-like tumor. We found 21articles (19 case reports and 2 review articles) relevant to our search since 2005. Except for two review articles, we thoroughly reviewed these articles (20 cases including our case) on the basis of age/sex, location, associated diseases, clinical symptoms, endoscopic findings, image findings, pathology, and treatment.[1,3-20]

**Discussion**

The mean age was 58.05 years, ranging from 19 years to 86 years with a female to male ratio of 3:1 (15:5) which was statistically more common in woman (Table 1).


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**Table 1.Summary of Patient Demographics and Presenting symptoms**

| Number of subjects | N=20 |
|---|---|
| **Age in years, mean (range)** | 58.05 (19 to 86) |
| Female (mean) | 57.87 |
| Male (mean) | 58.6 |
| **Sex, female/male (ratio)** | 15/5 (3 : 1) |
| **Location** | |
| Unilateral (Right : Left) | 20 (10 : 10) |
| **Origin Site** | |
| 10 | Nasal Septum |
| 4 | Paranasal sinuses |
| 1 | Sphenoethmoidal recess |
| 1 | Olfactory fissure |
| 1 | Inferior turbinate |
| 1 | Alar nose |
| 2 | NOS* |
| **Presenting symptoms (up to 5 in order of frequency** | |
| Nasal obstruction | 13 |
| Epistaxis | 12 |
| Headache | 4 |
| Facial pressure or pain | 3 |
| Olfaction change | 3 |

All 20 patients had unilateral lesion with no side preponderance. The most frequent site of sinonasal glomangiopericytoma origin was the nasal septum (10 patients), followed by the paranasal sinuses (4patients). Clinical symptoms of sinonasal glomangiopericytoma are non-specific and very variable depending on the size and location.

Of the presenting symptoms as shown in table 1, the most common symptom was nasal obstruction, seen in 13 patients, followed by epistaxis (12patients), headache (4 patients), facial pain or pressure (3 patients) and anosmia (3 patients) in decreasing order of frequency. The clinical feature of our case was the character of epistaxis. Usually, epistaxis is self-limited and easily controlled by nasal packing unlike our patient whose epistaxis was so massive that the complete nasal packing in the nasal cavity was necessary.

On the basis of endoscopic findings or gross appearances on specimen of 19 patients, the common finding of sinonasal glomangiopericytoma was a polypoid mass in shape (11 patients). The color of tumor varied from violaceous to gray and the most common color of tumor was reddish (7 patients). The consistency of tumor was easily friable that bled easily with minimal manipulation (5 patients).

The most common imaging technique used to evaluate sinonasal glomangiopericytoma was CT, used in 16 patients. In 6 patients, MRI was performed for further evaluation and the differential diagnosis.[4,6,8,16,17,19] The most common mentioned CT finding was soft tissue density mass, seen in 10 patients and enhancement (heterogeneous, diffuse or obvious) after contrast administration, seen in 3 patients. Bone destruction was observed in 3 patients, and bony erosion was observed in 2 patients and no bony change was observed in 4 patients. On the basis of our review of MRI findings in 6 patients, sinonasal glomangiopericytomawas hypointense to isointense signal on T1-weighted images, isointense to hyperintense signal on T2-weighted images and demonstrated enhancement on gadolinium enhanced MRI. Other image studies performed for evaluation were F-18 FDG PET/CT in 1 patient[6] and MR angiography in 1 patient.[8]F-18 FDG PET/CT showed the mass to have uniformly low-grade hypermetabolism (SUVmax = 2.8).[6]However, there was no any finding of MR angiography which was not minutely described.[8]

Histopathological findings of 19 patients were described in detail. Three common findings were as follows; 1) covered with normal respiratory epithelium, 2) the presence of numerous thin-walled, branching staghorn vessels with perivascular hyalinization, and 3) the vessels surrounded by oval to spindle-shaped tumor cells.

Until now, so many various antibodies for immunohistochemical stains have been used for definitive diagnosis of sinonasal glomangiopericytoma. On the basis of17 patients' data (including our case) for immunohistochemical studies[1,4,5,9-20], various antibodies for immunohistochemical stains were


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used. Immunohistochemistry for vimentin, smooth muscle actin (SMA), muscle specific actin, CD34, desmin, S-100, cytokeratin including pankeratin, CD99, CD31, Bcl-2and Beta-Catenin was performed in at least 2 or more cases. Immunohistochemical staining was scored as negative or positive, but percentage of positive cells was not estimated except Ki-67 marker (2% nuclear positivityin one patient). The tumor cells were positive for vimentin (10/10, 100%), smooth muscle actin (14/15, 93.3%), muscle-specific actin (5/5, 100%), bcl-2 (2/2, 100%) and beta-catenin (2/2, 100%). Also, the tumor cells were negative for keratin (0/5) and desmin (0/4). However, CD34 (2/11, 18.2%),S-100 (1/7, 14.3%), CD99 (1/3, 33.3%) and CD31 (1/5, 20%)showed variable positivity. Although few in number, the tumor cells had immunoreactivity with cyclin D1 (1/1), progesterone receptor (1/1), CD146 (1/1), WT1 (1/1), mTOR (1/1) and EGFR (1/1). Recent Lasota's study[21] found that sinonasal glomangiopericytomas immunohistochemically showed strong expression and nuclear accumulation of β-catenin. Also, they suggest mutational activation of β-catenin and associated cyclin D1 overexpression may be central events in the pathogenesis of glomangiopericytoma and nuclear accumulation of β-catenin can be a diagnostic marker for glomangiopericytoma.

Except 4 patients whose treatment methods were not described in detail[6,7,10,20], data on 16 patients (including our case) were collected for treatment modalities.[1,3,4,5,8,9,11-19]The most common treatment modality was complete surgical excision through transnasal endoscopic surgery, used in 12 patients. The help of microdebrider and image-guided navigation system made it easier to remove the tumor completely in a few cases. Depending on the location and size of tumor, other surgical methods were medial maxillectomy in 1 patient[5], combined endoscopic surgery and Caldwell-Luc approach in 1 patient[9], combined external approach and endoscopic approach in 1 patient[12], and only external approach in 1 patient[15]. The prognosis of sinonasal glomangiopericytoma was excellent without recurrences even if the tumor was not completely removedin 3 patients because of bleeding and the location close to the artery.[9,13,14]Although we should consider the size and location of the tumor, we suggest the best surgical approach for sinonasal glomangiopericytoma is trananasal endoscopic surgery with the help of recently developed instruments. Also, long-term follow-up should be necessary since most of the recurrences may occur within the first 5 years after surgery. Further study is needed to elucidate the association between incomplete excision and tumor recurrence.

**Conclusions**

Based on our comprehensive review of sinonasal glomangiopericytoma, we found some clinical characteristics. First, sinonasal glomangiopericytoma was statistically more common in woman. Second, the most frequent site of origin was the nasal septum followed by the paranasal sinuses. Third, the most common symptom was nasal obstruction, followed by epistaxis, headache, facial pain or pressure, and anosmia in decreasing order of frequency. Fourth, computed tomography was the most common imaging technique used to evaluate sinonasal glomangiopericytoma which the most common mentioned CT finding was soft tissue density mass with enhancement. Last, the common treatment modality was complete surgical excision through transnasal endoscopic surgery.

**Ethical clearance**: prior the submission, this case report was approved by local ethics committee.

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**Author's contribution:**

Data gathering and idea owner of this study: Hyun Jin Min, Kyung Soo Kim

Study design: Hyun Jin Min, Kyung Soo Kim

Data gathering: Hyun Jin Min, Kyung Soo Kim

Writing and submitting manuscript: Hyun Jin Min, Kyung Soo Kim

Editing and approval of final draft: Hyun Jin Min, Kyung Soo Kim

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