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Endoscopic Treatment of Sinonasal Glomangiopericytoma: A Case Report in Light of the Literature

Yutaro Saito,* Nobuo Ohta,* Sachiko Konosu-Fukaya,† Fumi Shoji,* Takahiro Suzuki,* Naoya Noguchi,* Risako Kakuta,* Ryoukichi Ikeda,* Muneharu Yamazaki,* Yusuke Kusano,* Yusuke Ishida,‡ Masafumi Satake,* Kazue Ise,† Yuriko Kagaya,§ Ryo Tamura,§ Kazuhiro Murakami,† and Yasuhiro Nakamura†

**Division of Otolaryngology, Tohoku Medical and Pharmaceutical University, Sendai 981-0905, Japan, †Division of Pathology, Tohoku Medical and Pharmaceutical University, Sendai 981-0905, Japan, ‡Division of Anatomy and Cell Biology, Tohoku Medical and Pharmaceutical University, Sendai 981-0905, Japan, and §Division of Radiology, Tohoku Medical and Pharmaceutical University, Sendai 981-0905, Japan*

ABSTRACT

A 71-year-old Japanese male patient presented with a rare case of Glomangiopericytoma (GPC) of the left nasal with obstruction. Complete resection with endoscopic surgery was performed. Immunohistochemical staining for smooth muscle actin, β catenin, cyclin D1, vimentin, and factor 13 were helpful in establishing a definitive diagnosis. Extranasal treatment has been traditionally performed for successful management. However, recent advances in endoscopic treatment have enabled complete endoscopic resection of GPC, minimizing morbidity and facilitating subsequent surveillance for recurrence. Endoscopic management should be considered in suitable cases.

Key words endoscopic surgery; glomangiopericytoma; nasal obstruction

Glomangiopericytoma (GPC) is an extremely rare sinonasal perivascular tumor characterized by low-grade malignant tumor with a tendency of local recurrence. GPC was firstly reported as hemangiopericytoma in 1942, but the World Health Organization classified it as a distinct entity in 2005.[1, 2] Clinically, GPC is associated with nasal obstruction and severe nasal bleeding. Histologically, GPC is made up of short, bland spindle cells forming fascicular, storiform, or whorled patterns with frequent perivascular hyalinization showing immunoreactivity for smooth muscle actin and strong nuclear β-catenin. The mainstay of treatment is complete surgical extirpation to avoid recurrence. We report the case of a 71-year-old Japanese man who presented with left nasal obstruction. Complete resection with endoscopic surgery was performed. Diagnostic and therapeutic options are discussed in the light of the current literature.

PATIENT REPORT

A 71-year-old Japanese man was referred to our otolaryngology department for evaluation of nasal obstruction for several months. Endoscopic examination revealed a soft, gray, elastic mass in the left nasal cavity (Fig. 1). Preoperative coronal computed tomography (CT) scan of the paranasal sinuses showed a low-density, homogeneous lesion occupying the left nasal cavity (Fig. 2). Preoperative coronal and axial magnetic resonance imaging (MRI) showed that the tumor had low signal intensity in the T1-weighted image and low to iso signal intensity in the T2-weighted image (Fig. 2). CT and MRI indicated no evidence of intracranial extension or bony defect of the skull base. In case of malignancy, preoperative pathological examination was not performed to

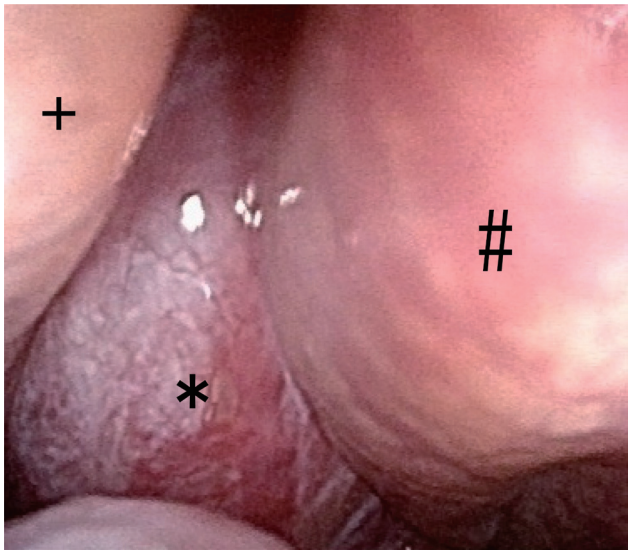

Fig. 1. Preoperative endoscopic photograph of the left nasal cavity occupied by the mass. (* tumor, # middle turbinate, + nasal septum)

Corresponding author: Nobuo Ohta, MD, PhD
noohta@hosp.tohoku-mpu.ac.jp
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Abbreviations: CT, computed tomography; GPC, glomangiopericytoma; MRI, magnetic resonance imaging; WHO, World Health Organization

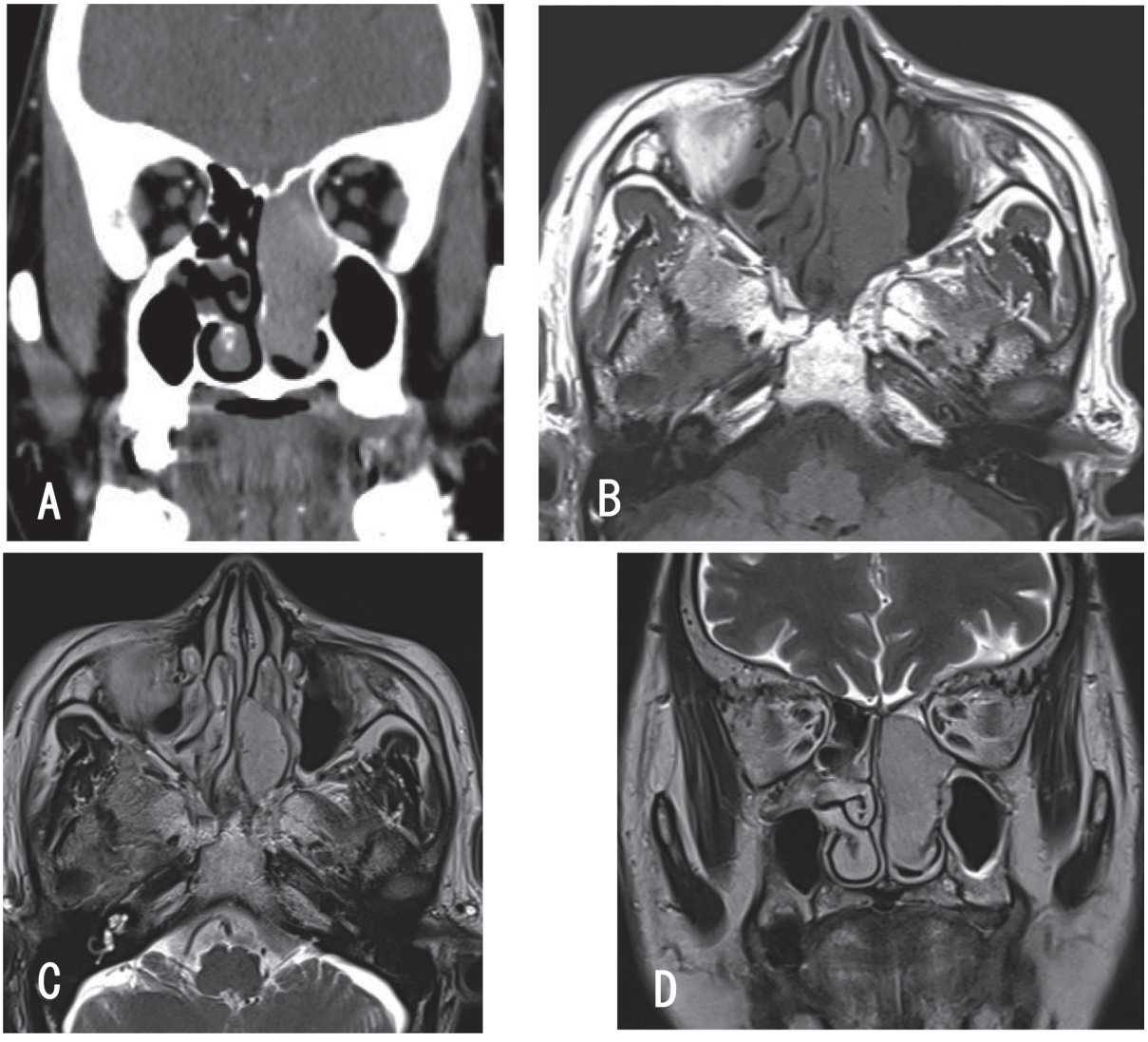

Fig. 2. Preoperative coronal CT scan of the paranasal sinuses showed a low-density, homogeneous lesion occupying the left nasal cavity (**A**). Preoperative coronal and axial MRI showed that the tumor had low signal intensity in the T1-weighted image and low to iso signal intensity in the T2-weighted image (**B**–**D**). CT, computed tomography; MRI, magnetic resonance imaging.

avoid tumor dissemination. The patient was admitted to our department for definitive diagnosis and treatment. Total endoscopic resection of the tumor was performed under general anesthesia. The tumor was removed with the use of 0-degree-of-view, 4-mm-diameter rigid endoscopes. Histological examination during the operation suggested that the tumor was a vascular tumor. Since the tumor adhered to the posterior middle turbinate, the inferior turbinate was firstly partially resected to obtain a clearer and well-defined visualization of the tumor. Next, the upper portion of the middle turbinate was also resected to access the tumor easily. Finally, the tumor was removed *en bloc* with the middle turbinate. The volume of bleeding during surgery was 50 mL. Histologically, it consisted of a uniform proliferation of ovoid to spindle-shaped cells with partially surrounding hyalinization and slightly branching vascular structures (Fig. 3). The tumor cells were positive for smooth muscle actin, nuclear β catenin, vimentin, factor 13, and cyclin D1 (Fig. 3). The definitive diagnosis was GPC. The patient's condition was successfully managed with excision of the tumor, and he remains well with no evidence of recurrence 13 months after treatment.

DISCUSSION

GPC is a sinonasal perivascular tumor with low malignant potential,[1, 2] and had been designated as an hemangiopericytoma-like tumor or sinonasal hemangiopericytoma. Its similarity with a glomus tumor, the term of GPC was preferred by the WHO in 2005.[1, 2] Most

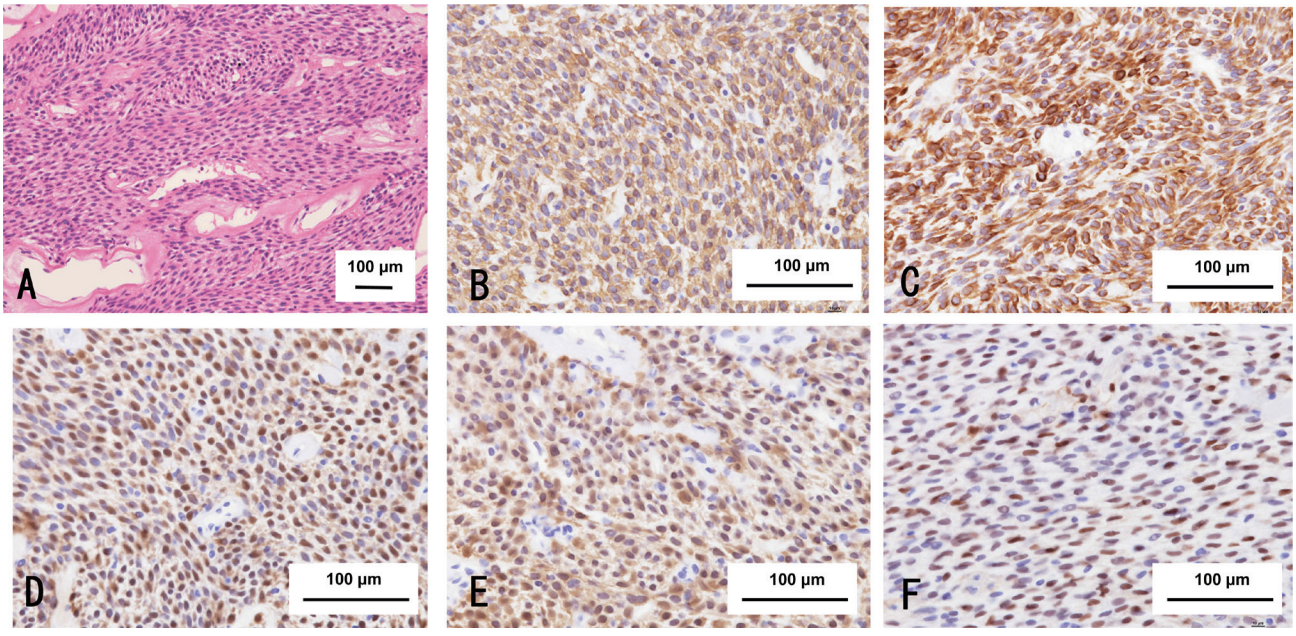

Fig. 3. Pathological examination showed (**A**) uniform proliferation of oval to short spindle-shaped cells with fibrous edematous tissue, and hyalinization (hematoxylin and eosin staining, Bar = 100 μm). (**B**) From the same area, cone-shaped cells positive for anti–smooth muscle actin antibody can be seen (Bar = 100 μm). (**C**) Cone-shaped cells also positive for anti-vimentin antibody can be seen (Bar = 100 μm). (**D**–**F**) Cone-shaped cells are also positive for (**D**) beta-catenin, (**E**) factor 13a, and (**F**) cyclin D1 (Bar = 100 μm).

GPC are diagnosed in the sixth and seventh decades of age.[1, 2] There is no recognized familial tendency and sexual predilection.[1, 2] High vascularization caused by trauma, hypertension, pregnancy, or usage of corticosteroids might be involved in its pathogenesis, however, its etiology is still not clear.[1, 2]

The diagnosis of GPC is facilitated by CT or MRI, but a definitive diagnosis is often obtained only after biopsy or complete resection.[2–6] For differential diagnosis, vascular and other tumors such as solitary fibrous tumors, lobular capillary hemangiomas, angiofibromas, and glomus tumors should be kept in mind.[7–9] Interestingly, the expressions of cyclin D1 and β catenin were observed in this case. Cyclin D1 is considered to dysregulate the cell cycle contributing neoplastic transformation.[7–9] Overexpression of β catenin encoded by CTNNB1 gene might play important role in tumorigenesis and tumor progression.[7–9] Aberrant expression of CTNNB1 can lead to cyclin D1 overexpression.[7–9] β catenin, a cadherin-associated membrane protein, is involved in the regulation of cell-to-cell adhesion and gene transcription as a nuclear protein.[7–9] Taken together, activating β catenin and overexpression of cyclin D1 have a central role in pathogenesis of GPC. Recent reports demonstrated that strong nuclear accumulation of β catenin was observed in GPC and not in Glomus tumor.[7–9] Therefore, nuclear expression of β catenin can be useful in distinguishing GPC and glomus tumor.[4, 6–9]

The treatment of choice is complete surgical extirpation to avoid recurrence. The recurrence rate is reported to be 7% to 20%; recurrence is thought to result from incomplete excision.[4, 6, 7] However, endoscopic treatment has been attempted for tumor resection.[4, 5] Intranasal endoscopic treatment has some advantages over extranasal treatment. Endoscopic treatment allows clear visualization of each wall of the nasal cavity, and a precise procedure can be performed with minimum invasion of intact tissues. Accordingly, in juvenile cases, endoscopic treatment enables complete removal of the mass with neither postoperative facial deformity or subsequent abnormal development of the nose and paranasal sinuses.[5] Although intranasal endoscopic treatment can be effective in most cases, anatomical variants such as severe septal deviation, large tumors or highly vascularized tumors can make it difficult.[4, 6–8] In preset cases, CT and MRI indicated no evidence of intracranial extension or bony defect of the skull base and intranasal endoscopic treatment was preferred to extranasal treatment. Preoperative embolization before resection of GPC has been recommended to reduce the volume of bleeding during surgery.[4, 6–8]

In conclusion, although GPC is extremely rare, it should be considered in the differential diagnosis of masses in the nasal cavity. Immunohistochemical

staining for smooth muscle actin, β catenin, cyclin D1, vimentin, and factor 13 are helpful in establishing a definitive diagnosis. Endoscopic management should be considered in suitable cases.

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The authors declare no conflict of interest.

REFERENCES

1 Stout AP, Murray MR. Hemangiopericytoma: a vascular tumor featuring Zimmermann's pericytes. Ann Surg. 1942;116:26-33. PMID: 17858068, DOI: 10.1097/00000658-194207000-00004

2 Barnes L. World Health Organization Classification of Tumors: pathology and genetics of head and neck tumors. Lyon France: IARC Press; 2005. p. 43-5.

3 Higashi K, Nakaya K, Watanabe M, Ikeda R, Suzuki T, Oshima T, et al. Glomangiopericytoma of the nasal cavity. Auris Nasus Larynx. 2011;38:415-7. PMID: 21093999, DOI: 10.1016/j.anl.2010.08.009

4 Lasota J, Felisiak-Golabek A, Aly FZ, Wang ZF, Thompson LDR, Miettinen M. Nuclear expression and gain-of-function β-catenin mutation in glomangiopericytoma (sinonasal-type hemangiopericytoma): insight into pathogenesis and a diagnostic marker. Mod Pathol. 2015;28:715-20. PMID: 25431235, DOI: 10.1038/modpathol.2014.161

5 Tessema B, Eloy JA, Folbe AJ, Anstead AS, Mirani NM, Jourdy DN, et al. Endoscopic management of sinonasal hemangiopericytoma. Otolaryngol Head Neck Surg. 2012;146:483-6. PMID: 22049019, DOI: 10.1177/0194599811427660

6 Anzai T, Saito T, Tsuyama S, Toh M, Ikeda K, Ito S. A case of glomangiopericytoma at the nasal septum. Head Neck Pathol. 2018;12:572-5. PMID: 29168109, DOI: 10.1007/s12105-017-0870-6

7 Psoma E, Karkos PD, Dova S, Gavriilidis M, Markou K, Kouskouras C, et al. Sinonasal glomangiopericytoma treated with preoperative embolisation and endoscopic sinus surgery. Ecancermedicalscience. 2016;10:692. PMID: 27994648, DOI: 10.3332/ecancer.2016.692

8 Park ES, Kim J, Jun SY. Characteristics and prognosis of glomangiopericytomas: A systematic review. Head Neck. 2017;39:1897-909. PMID: 28497509, DOI: 10.1002/hed.24818

9 Asimakopoulos P, Syed MI, Andrews T, Syed S, Williams A. Sinonasal glomangiopericytoma: is anything new? Ear Nose Throat J. 2016;95:E1-6. PMID: 26930335, DOI: 10.1177/014556131609500202