CT and MRI Findings of Glomangiopericytoma in the Head and Neck: Case Series Study and Systematic Review

C.H. Suh, J.H. Lee, M.K. Lee, S.J. Cho, S.R. Chung, Y.J. Choi, and J.H. Baek

ABSTRACT

SUMMARY: Glomangiopericytoma is a rare sinonasal mesenchymal tumor of borderline or low malignant potential. We reviewed the CT and MR imaging findings of head and neck glomangiopericytoma via a retrospective case series study and systematic review. Our study revealed that glomangiopericytoma is a well-defined lobulated avidly enhancing soft-tissue mass with erosive bony remodeling that is most commonly found in the sinonasal cavity. Typically, it is hyperintense on T2-weighted images with vascular signal voids, has a high mean ADC value, and a wash-in and washout pattern on dynamic contrast-enhanced MR imaging. Although the CT findings are nonspecific, typical MR imaging findings, including those on the ADC map and dynamic contrast-enhanced MR imaging, may be helpful for differentiating glomangiopericytomas from other hypervascular tumors in the head and neck.

ABBREVIATION: DCE = dynamic contrast-enhanced

Glomangiopericytoma is a rare sinonasal mesenchymal tumor demonstrating a perivascular myoid phenotype and is one of the sinonasal vascular spindle cell tumors, such as spindle cell hemangioma, kaposiform hemangioendothelioma, and Kaposi sarcoma.¹⁻³

Glomangiopericytoma, which accounts for <0.5% of all sinonasal tumors, was categorized as a borderline or low-malignant-potential tumor by the World Health Organization in 2005.¹ A previous study showed that 18 of 104 patients (17.3%) had a local recurrence and were managed by additional surgery.⁴ Glomangiopericytoma was formerly called “sinonasal hemangiopericytoma” because of a hemangiopericytomatous pattern.

To date, a number of case reports have been published describing the CT or MR imaging findings of glomangiopericytoma in the head and neck. Although these previous reports demonstrated that glomangiopericytoma is usually a well-defined and avidly enhancing soft-tissue mass with erosive remodeling mainly in the sinonasal cavity, there has been no report systematically

reviewing the CT and MR imaging findings of glomangiopericytoma in the head and neck. In addition, details on the findings on advanced MR imaging methods including DWI and dynamic contrast-enhanced (DCE)-MR imaging have not yet been published.

Therefore, the purpose of our study was to review the CT and MR imaging findings of glomangiopericytoma in the head and neck, including findings on DWI and DCE-MR imaging, via a retrospective study of a case series and a systematic review.

Case Series

Our study was approved by the institutional review board of Asan Medical Center, and the requirement for informed consent was waived due to the retrospective nature of the study. The pathology reports for January 2010 to January 2019 were electronically searched using “glomangiopericytoma,” “sinonasal hemangiopericytoma,” “sinonasal-type hemangiopericytoma,” “hemangiopericytoma-like tumor,” and “hemangiopericytoma of sinonasal origin” as keywords. Patients who underwent preoperative CT and/or MR imaging were enrolled. Clinical and demographic data including age, sex, and symptoms were acquired from the electronic medical record. The imaging findings including location, size, border, adjacent bone change, CT density, enhancement pattern, MR imaging signal intensity on T2WI, ADC value of the whole tumor, the presence of vascular signal voids, and DCE-MR imaging patterns were independently reviewed by 2 board-certified neuroradiologists (J.H.L., with

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From the Department of Radiology and Research Institute of Radiology, University of Ulsan College of Medicine, Asan Medical Center, Seoul, Republic of Korea.

Please address correspondence to Jeong Hyun Lee, MD, PhD, Department of Radiology and Research Institute of Radiology, University of Ulsan College of Medicine, Asan Medical Center, 86 Asanbyeongwon-Gil, Songpa-Gu, Seoul 05505, Republic of Korea; e-mail: jeonghlee@amc.seoul.kr

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FIG 1. A 79-year-old woman with sinonasal glomangiopericytoma. A, Nonenhanced CT shows a midline bilobed soft-tissue mass (*long arrow*) overriding the permeated nasal septum with erosive remodeling on the bony wall of the left ethmoidal sinus (*short arrows*). On MR imaging, the mass (*arrow*) shows slight hyperintensity on T2WI (B) and strong and homogeneous enhancement on fat-suppressed contrast-enhanced T1WI (C). D, On the ADC map, the mass shows a high mean ADC value ($1.47 \times 10^{-3} \text{ mm}^2/\text{s}$) (*arrows*). E and F, On DCE-MR imaging, the mass reveals wash-in and washout patterns on the initial and final area under the curve maps, which were obtained by trapezoidal integration of the normalized signal intensity curve during the initial 90 seconds after the onset of contrast enhancement and during the last 90 seconds using the same enhancing voxels.

18 years of experience in head and neck neuroradiology, and C.H.S., with 6 years of experience in neuroradiology). We placed ROIs for the entire tumor to obtain a mean ADC value of the whole tumor.

Six patients satisfied the inclusion criteria (3 men and 3 women; mean age, 67 years; range, 48–79 years; On-line Table 1). Four patients had symptoms, and 2 patients were identified incidentally on imaging studies. Following initial detection, all patients underwent an operation. One patient had preoperative embolization of the tumor. Two patients had minor bleeding during resection, but the other 3 had no bleeding complication, despite no preoperative embolization. Four glomangiopericytomas were located in the sinonasal cavity; 1, in the masticator/buccal spaces, and 1, in the prevertebral space. The maximum diameters ranged from 2.9 to 5 cm (median, 3.3 cm).

All patients underwent CT, which was performed using ≥ 16 -detector row CT scanners with excellent image quality. Intravenous iodine contrast agent was used in all except 1 patient. All glomangiopericytomas were well-defined round or lobulated contoured soft-tissue masses (Fig 1). Five tumors were in direct contact with the adjacent bone, which showed erosive remodeling, while 1 tumor was surrounded by soft tissue. On contrast-enhanced CT, 2 tumors revealed avid and homogeneous enhancement patterns that were slightly lower than the adjacent vessels (Fig 2), whereas 3 tumors demonstrated heterogeneous enhancement.

Four of 6 patients underwent MR imaging, which included DWI in 1 patient and both DWI and DCE-MR imaging in 2 patients. All MR imaging was performed using a 3T scanner with excellent image quality. Detailed information about our own MR imaging parameters was the same as that published in previous articles by our group.^{5,6}

On MR imaging, 2 glomangiopericytomas showed homogeneous hyperintensity on T2WI, 1 glomangiopericytoma showed intermediate signal intensity, and 1 glomangiopericytoma showed heterogeneous intermediate-to-high signal intensity. All glomangiopericytomas revealed vascular signal voids on T2WI. The 3 patients for whom ADC maps were available demonstrated high mean ADC values ranging from 1.27 to $2.09 \times 10^{-3} \text{ mm}^2/\text{s}$ (On-line Table 1 and Figs 1 and 2). On contrast-enhanced MR imaging, 3 glomangiopericytomas in the sinonasal cavity or masticator space revealed avid and homogeneous enhancement, whereas 1 glomangiopericytoma in the prevertebral space revealed heterogeneous enhancement. On DCE-MR imaging, 2 glomangiopericytomas revealed a rapid wash-in and washout pattern (Fig 1).

For the systematic review, a systematic search of PubMed and EMBASE was performed using the following search term: ((glomangiopericytoma) OR (“sinonasal hemangiopericytoma”) OR (“sinonasal-type hemangiopericytoma”) OR (“hemangiopericytoma-like tumor”) OR (“hemangiopericytoma of sinonasal origin”)) AND ((“computed tomography”) OR (CT) OR (“magnetic

FIG 2. A 72-year-old man with glomangiopericytoma in the prevertebral space. A, Contrast-enhanced CT shows a well-defined heterogeneously enhancing mass in the right prevertebral space. The mass demonstrates iso- to hyperintensity on T2WI (B) and has heterogeneous enhancement on fat-suppressed contrast-enhanced T1WI (C). Note several vascular signal voids (arrows) at the periphery of the mass. D, On the ADC map, the mass shows a high mean ADC value ($2.09 \times 10^{-3} \text{ mm}^2/\text{s}$). E and F, On DCE-MR imaging, the mass reveals rapid wash-in and washout patterns on the initial and final area under the curve maps, which were obtained by trapezoidal integration of the normalized signal intensity curve during the initial 90 seconds after the onset of contrast enhancement and during the last 90 seconds using the same enhancing voxels.

resonance imaging”) OR (“MR imaging”) OR (MRI)). The literature search was not limited by any filters or published language. The search was updated until August 15, 2019. The literature search, study selection, and data extraction were independently performed by 2 board-certified neuroradiologists (J.H.L. and C.H.S.).

This systematic search originally yielded 59 articles (38 from PubMed and 21 from EMBASE). After removal of duplicate articles and those that were not in the field of interest, 11 case reports with 14 cases were left for inclusion in this systematic review (On-line Table 2).⁷⁻¹⁷ Eleven of 14 glomangiopericytomas were located in the sinonasal cavity and had maximum diameters ranging from 2.9 to 4 cm. On CT, 11 glomangiopericytomas were well-defined lobulated contoured soft-tissue masses and 7 showed erosive remodeling of adjacent bone. On contrast-enhanced CT, 6 glomangiopericytomas revealed strong and homogeneous enhancement patterns, while 2 glomangiopericytomas demonstrated heterogeneous enhancement. On MR imaging, 3 glomangiopericytomas showed hyperintensity on T2WI. These CT and MR imaging findings were similar to our findings.

DISCUSSION

Both our case series and the systematic review confirmed that glomangiopericytomas are well-defined round or lobulated,

contoured soft-tissue masses that frequently show erosive bone remodeling, especially in the sinonasal cavity. Our study also revealed that glomangiopericytomas usually show avid and homogeneous enhancement on contrast-enhanced CT or MR imaging, intermediate-to-high signal intensity on T2WI, high ADC values, and rapid wash-in and washout patterns on DCE-MR imaging. They are also accompanied by vascular signal voids on T2WI. Although the individual imaging findings might be nonspecific, a combination of these findings on CT and MR imaging, especially when DWI and DCE-MR imaging are included, may help to suggest glomangiopericytoma of borderline malignant potential, which is important in preoperative planning of the surgical approach or extent of resection.

In this study, we report on the CT and MR imaging (including DWI and DCE-MR imaging) findings of glomangiopericytoma. While the lesions are imaged in a manner similar to that of other vascular tumors, the combination of findings may help differentiate glomangiopericytoma from other hypervascular tumors. High ADC values indicate low cellularity, which reflects the low malignant potential of the tumor. Therefore, high ADC values may be a key imaging finding for differentiating glomangiopericytoma from malignant sinonasal cancer.¹⁸ The signal voids on T2WI and avid enhancement suggest a vascular tumor. Surgeons may request preoperative embolization in this category of lesion before endoscopic resection.⁹

Given the submucosal nature of glomangiopericytomas, they can be differentiated from more common mucosal tumors such as squamous cell carcinomas or inverted papillomas. The differential diagnosis for a submucosal tumor is broad and can include solitary fibrous tumor, lobular capillary hemangioma, or nasopharyngeal angiofibroma. Differentiating these entities when they occur in the head and neck can be challenging because of similar imaging findings. However, the distinction is clinically important because glomangiopericytoma has borderline or low malignant potential, unlike some of the other disease entities. Solitary fibrous tumor, formerly known as hemangiopericytoma, consists of a cellular spindle cell proliferation effacing submucosal structures with staghorn-type vessels,¹⁹ with the tumors being characterized by chromosomal translocation resulting in the formation of a *NAB2-STAT6* fusion gene; thus nuclear staining of *STAT6* is a reference standard method for diagnosing solitary fibrous tumors.²⁰ Glomangiopericytoma is most commonly confused with solitary fibrous tumor because solitary fibrous tumor is also a well-defined tumor with avid enhancement and a washout time-intensity curve pattern on DCE-MR imaging.²¹ However, solitary fibrous tumor in the sinonasal cavity shows iso- to hypointensity on T2WI because of its fibrous component.²² Although it is difficult to differentiate glomangiopericytoma from solitary fibrous tumor on CT, these signal intensity differences on T2WI may be helpful.

Lobular capillary hemangioma is another potential differential diagnosis; it has a lobular architecture with staghorn-type vessels in the interlobular stroma.²³ Although lobular capillary hemangioma is also known to be a well-defined avidly-enhancing mass with hyperintensity on T2WI and a washout time-intensity curve pattern, bony changes are not usually seen on CT or MR imaging.^{24,25} Nasopharyngeal angiofibroma has abundant stromal collagen and a prominent vascular stroma. Although angiofibroma is a similar hypervascular tumor, the typical demographic data, including age and sex and the characteristic location (ie, lateral nasopharyngeal mucosa at the sphenopalatine foramen), can help to differentiate angiofibroma from glomangiopericytoma. When glomangiopericytoma occurs outside the sinonasal cavity, neurogenic tumors can be one of the major differential diagnoses. However, neurogenic tumor can be differentiated by its location, typical shape, and heterogeneous hypoenhancement.

The main limitations of our study are the small number of enrolled cases and its retrospective nature, inevitable given the rare occurrence of glomangiopericytoma in the head and neck. We have attempted to overcome these limitations by performing a systematic review, and the results are in accord with ours. We were able to include 3 cases with advanced MR imaging techniques, and these techniques may be of assistance in generating a more focused differential diagnosis. Tissue sampling remains the standard, however. We believe that our results could provide an insight into further investigation of glomangiopericytoma with advanced MR imaging techniques.

CONCLUSIONS

Our case series and the systematic review revealed that glomangiopericytoma is a well-defined lobulated avidly-enhancing soft-tissue mass with erosive bony remodeling usually occurring in

the sinonasal cavity. It typically shows hyperintense signal on T2WI with vascular signal voids, a high mean ADC value, and a wash-in and washout pattern on DCE-MR imaging. Although the CT findings are nonspecific, typical MR imaging findings, including the ADC value and DCE-MR imaging pattern, can be helpful for differentiating glomangiopericytomas from other hypervascular tumors in the head and neck, especially in the sinonasal cavity.

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