YALE JOURNAL OF BIOLOGY AND MEDICINE 94 (2021), pp.593-597.

CASE REPORT

YALE JOURNAL OF BIOLOGY AND MEDICINE
YJBM
CELEBRATING 90 YEARS 1928–2018

Sinonasal Glomangiopericytoma: Review of Imaging Appearance and Clinical Management Update for a Rare Sinonasal Neoplasm

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Introduction: Glomangiopericytoma (GPC) is a rare tumor in the nasal cavity or paranasal sinuses with low malignant potential. Initially deemed a hemangiopericytoma, in 2005 it was classified as a distinct entity by the World Health Organization (WHO). **Case Presentation**: A male patient in his early 60s presented with new-onset right arm and leg weakness/numbness, who was incidentally found to have a left ethmoid sinus mass with extension in the olfactory fossa. On CT and MRI, the mass enhanced with well-defined borders and eroded the bone, but without dural enhancement. The mass was surgically excised, and pathology confirmed the diagnosis of glomangiopericytoma by microscopic appearance and staining. **Discussion**: Glomangiopericytoma has less than 0.5% incidence of all neoplasms of the sinonasal cavity, making it rare. Most diagnosed patients are in their 6th or 7th decade of age, with a slight female predominance. Treatment is complete surgical excision, with excellent prognosis, although there is up to 17% local recurrence. Despite the non-specific appearance on CT and MRI, imaging can help provide differential diagnosis, tumor extent, size, and reassuring non-aggressive characteristics of the tumor prior to surgery. GPC tumors are relatively resistant to radiation and chemotherapy. **Conclusion**: It is important to recognize glomangiopericytoma in the differential of masses of the nasal cavities or paranasal sinuses, as they rarely warrant aggressive treatment beyond local excision. Each reported case of glomangiopericytoma helps to build guidance for imaging and treatment since GPC is rare and not well-represented in the medical literature.

INTRODUCTION

Sinonasal glomangiopericytoma is a rare mesenchymal neoplasm arising in the nasal cavity or paranasal sinuses [1,2]. It is considered a borderline low malignant potential soft tissue tumor [1-5]. It is also called sinonasal-type hemangiopericytoma, which arises from the pericyte cells surrounding the capillaries [1]. The first reported case was in 1942 by Stout and Murray, which was originally classified as hemangiopericytoma [2,3,5-

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Abbreviations: GPC, glomangiopericytoma; CT, computed tomography; CTA, computed tomography angiogram; MRI, magnetic resonance imaging; ENT, ear, nose, and throat.

Keywords: sinonasal, glomangiopericytoma, neoplasm, skull base, rare

Author Contributions: Each author has contributed to this manuscript including conception, design, review, and editing. The authors have no outside source of funding as a conflict of interest.

7]. The lesion was thought to fall in the spectrum between glomus tumors and capillary hemangioma; hence the term hemangiopericytoma [2]. Since that initial description, the definition of the disease has been associated with controversy. In 1976, it was described by Compagno as "haemangiopericytoma like" due to low incidence of metastasis and mortality [5]. Glomangiopericytoma (GPC) differs from a conventional soft tissue hemangiopericytoma in location, biologic behavior, and histologic features [2]. GPC is an indolent tumor and a variant of hemangiopericytoma, the latter being more aggressive and invasive [8]. The World Health Organization (WHO) classified glomangiopericytoma as a distinct entity in 2005 [4,6-9]. The detection of GPC can be facilitated by the use of CT or MRI. However, the definitive diagnosis is achieved by tissue sampling after complete resection or biopsy [6]. It has an excellent prognosis with surgical excision. The tumor has tendency for local recurrence.

CASE PRESENTATION

A male patient in his early 60s with history of atrial fibrillation was off anticoagulation for planned outpatient laparoscopic cholecystectomy. The patient has no history of cancer or sinonasal disease. He drinks alcohol occasionally and is a former smoker, who quit smoking over 30 years ago. While the patient was in the pre-op area for his elective cholecystectomy surgery, he had sudden onset right arm and leg weakness/numbness. Subsequently, stroke code was activated. A computed tomography angiogram (CTA) head and neck was done, which was negative for stroke or acute arterial disease. His neurologic symptoms improved spontaneously and went back to baseline. The likely diagnosis for his neurologic symptoms was transient ischemic attack in the setting of Eliquis being held for surgery. Incidentally, a mass was found in the CTA involving the left ethmoid sinus and olfactory fossa. The patient had no epistaxis, no facial numbness, no disturbance in smell or taste. CT without contrast demonstrated a mass isodense to soft tissue, without any calcification. The 2.5 cm left ethmoid mass demonstrated arterial enhancement, eroding the ethmoid cribriform plate and left lateral lamella (Figure 1A-B). However, there was no adjacent periosteal reaction or hyperostosis, suggesting a nonaggressive neoplasm. The mass protruded through the bone into the anterior cranial fossa. On MRI, the mass was isointense to brain parenchyma on T1 and T2 sequences (Figure 1C), with no restricted diffusion, and homogenous contrast enhancement (Figure 1D). There was no dural enhancement or thickening. There was only minimal mass effect, and no adjacent cerebral edema.

Endoscopic biopsy of the left ethmoid mass was performed. The pathology report showed low-grade spindle cell neoplasm, favoring glomangiopericytoma. It showed submucosal bland spindle cells with thick-wall vessels and adipocytes (Figure 2). CD34, ERG, and SMA highlighted the vessels, but not the spindle cells. Beta catenin was diffusely positive. S100, EMA, STAT6, and HMB-45 were negative. Ki-67 demonstrated a proliferative index of approximately 1%.

The decision was made to endoscopically excise the tumor with a collaborative operation by ENT and neurosurgery. While there was osseous erosion, the dura remained without tumor invasion. The pathology report confirmed the final diagnosis of sinonasal glomangiopericytoma with negative resection margins. No chemotherapy or radiation therapy was planned because of the improved 10-year survival without these adjuvant treatments. The patient was doing well post-operatively at 21 months following the complete resection of the tumor without clinical evidence for recurrence.

DISCUSSION

Glomangiopericytoma is a rare sinonasal perivascular tumor with low malignant potential [7]. Its incidence is less than 0.5% of all neoplasms of the sinonasal cavity, which is rare [1,4-6,8-11]. Most diagnosed patients are in their 6th or 7th decade of age [7], which applies to our patient. There is a slight female predominance [2]. GPC is generally localized to the nasal cavity, with paranasal involvement and skull base extension uncommon [10]. If paranasal sinus involvement is present, the ethmoid and sphenoid sinus are most commonly affected. Our patient had a left ethmoid sinus tumor. The most commonly reported clinical presentation was epistaxis (78%), followed by nasal obstruction (52%), and then headache (17%) [1]. In our case, the patient was without sinonasal symptoms. Its etiology is still not clear. However, there are suggestions that high vascularity caused by trauma, pregnancy, hypertension, or usage of corticosteroids might be involved in its pathogenesis [7].

GPC has characteristics similar to sinonasal polyps and could be confused for it [6]. The glomangiopericytomas are submucosal in nature, which allows differentiation from the more common mucosal tumors such as squamous cell carcinomas or inverted papillomas [4]. Differential diagnosis for a submucosal tumor includes solitary fibrous tumors (hemangiopericytoma), lobular capillary hemangiomas, nasopharyngeal angiofibromas, desmoid type fibromatosis, and glomus tumors [1,4,7]. Differentiating these entities in the head and neck can be difficult because of some overlapping imaging findings, making tissue sampling (either biopsy or resection) the criterion standard for diagnosis.

Preoperative workup may include endoscopy, CT, and MRI to assess extent, size, and characteristics of the

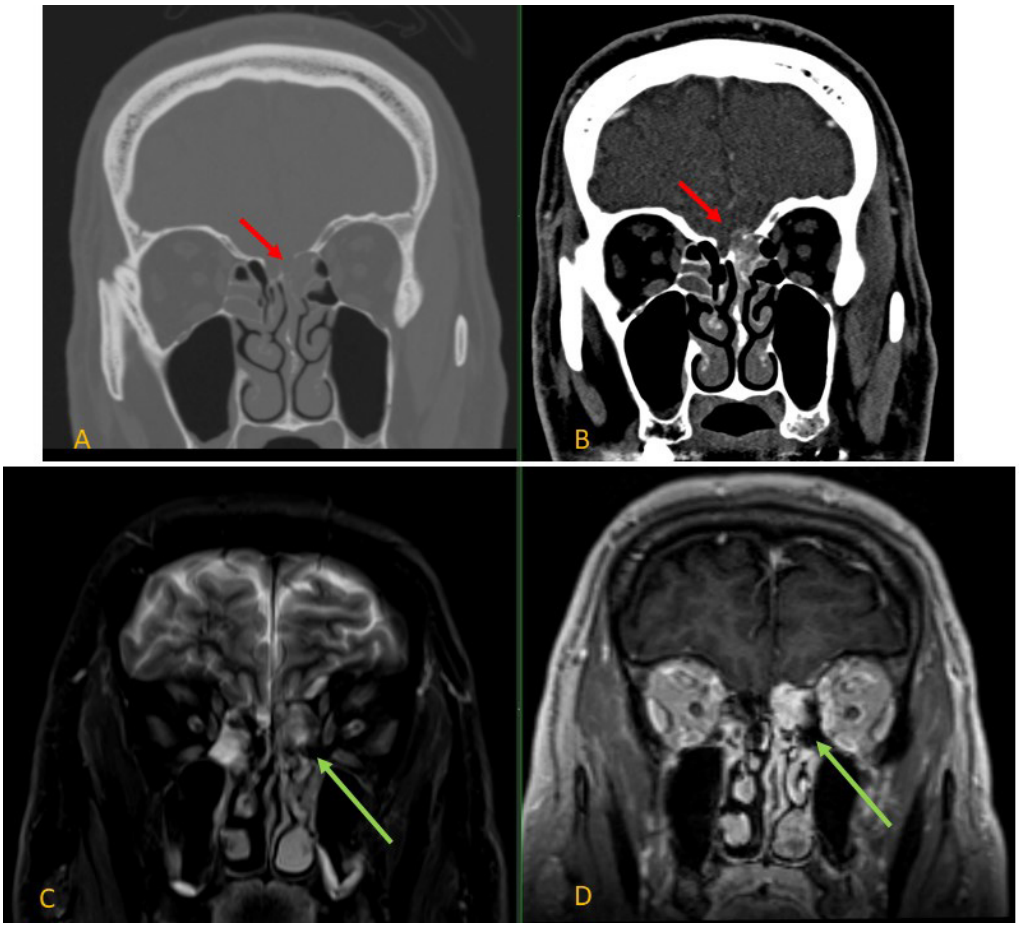

Figure 1. Coronal CT in bone window (**A**) demonstrates the left ethmoid mass eroding the cribriform plate and left lateral lamella. Coronal CTA in soft tissue window (**B**) shows the mass displaying arterial enhancement. Coronal T2 MRI (**C**) demonstrates isointensity of the left ethmoid mass as compared to the brain parenchyma. Coronal T1 post contrast MRI (**D**) displays homogenous enhancement of the mass.

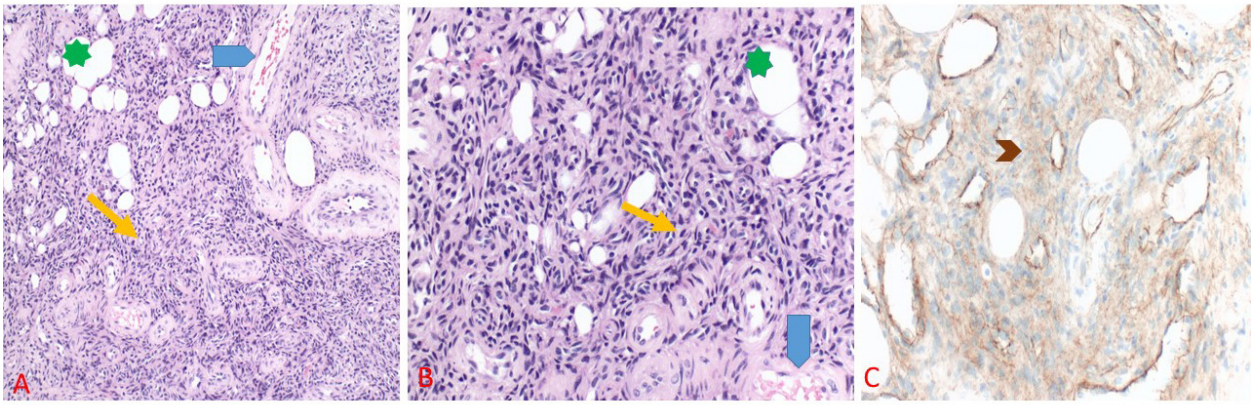

Figure 2. Low power (**A**) and high power (**B**) magnification demonstrate submucosal bland spindle cells with admixed thick-wall vessels and adipocytes, compatible with glomangiopericytoma. (**C**) Beta catenin shows cytoplasmic staining. While not the typical cytoplasmic and nuclear staining seen in glomangiopericytoma, the morphology is very classic. Orange arrow: submucosal bland spindle cells; blue arrowhead: thick-wall vessels; green star: adipocytes; and brown chevron: cytoplasmic beta staining.

tumor for appropriate preoperative planning [10]. On contrast-enhanced CT and MRI, GPC shows avid and homogeneous enhancement [4]. On imaging, the glomangiopericytomas appear as well-defined, round or lobulated soft tissue masses. These tumors frequently show erosive bone remodeling, especially in the sinonasal cavity. Our study also revealed a well-defined soft tissue tumor in the left ethmoid sinus with homogenous enhancement on CT and MRI, and with bony erosion. On MRI, there has been variable T2 appearance including T2 hyperintensity and intermediate signal [4]. Our case, the tumor showed T2 isointensity to adjacent brain parenchyma.

After acquiring tumor tissue, diagnosis is mainly based on the histological characteristics showing epithelioid cells with myoid cell differentiation and immunohistochemistry [8]. GPC is diagnosed by characteristic histology showing uniform proliferation of oval to short spindle shaped cells under the epithelium [1]. On immunohistochemistry, it is positive for cytoplasmic SMA and Vimentin, and nuclear beta catenin in 80-100%. GPC tumor cells are negative for CD34, AE1/AE3, Bcl-2, CD99, CD117, Factor VIIIR Ag, S-100 protein, and STAT6. High Ki-67 index (>10%) is a prognostic factor for aggressive behavior. Nuclear staining for beta catenin is diagnostic marker of GPC [1,3]. In our case, the tumor also had submucosal bland spindle cells, and diffusely positive for beta catenin. It also demonstrated negative staining for CD34, S100, and STAT6. However, it was negative for SMA, unlike the literature. Our case demonstrated Ki-67 proliferative index of approximately 1%, indicating more indolent behavior.

The treatment of choice is complete surgical resection [6,7]. Prognosis is favorable with a 5-year survival rate of 88% [1]. The reported recurrence is 17% [3,6]. Recurrence is thought to be due to incomplete surgical excision [7], which can be managed by additional surgery [1]. The tumor is relatively resistant to chemotherapy and radiation therapy, so these adjuvant therapies are initially contraindicated [5]. Our patient is known to be doing well at their last follow-up 21 months following the complete resection of his tumor. This strengthens the current clinical practice of improved survival with surgery alone and prevents worse outcomes seen with initial treatments by chemotherapy or radiation therapy. Most recurrences arise within 5 years of excision [10]. Local recurrence has been shown to occur up to 12 years after initial excision, with most local recurrences happening within a year of removal are likely due to incomplete excision. Rates of metastasis are 5-10% and are usually preceded by multiple recurrences. In the rare case of metastatic disease, a combination of radiotherapy and chemotherapy can be used as adjuvant or palliative therapy.

For tumor resection, an endoscopic approach has some advantages over more traditional open approaches, including clear visualization of each wall of the nasal cavity and minimum invasion of intact tissues reducing postoperative facial deformity or abnormal subsequent deformity of the nose and paranasal sinuses [7]. Even though intranasal endoscopic treatment can be effective in most cases, highly vascularized tumors or large tumors can make it difficult. Preoperative embolization can be considered to reduce the volume of bleeding during surgery [7], especially for large or highly vascular tumors [10]. Post-operative management should include long-term surveillance with nasal endoscopy at regular intervals considering the local recurrence of the tumor and/or MRI and CT [10].

CONCLUSION

Sinonasal glomangiopericytoma is very rare amongst sinonasal tumors. It is important to recognize glomangiopericytoma in the differential diagnosis of masses of the nasal cavity or paranasal sinuses, as they tend to behave in an indolent fashion, rarely warranting aggressive treatment beyond local excision. The definitive diagnosis of GPC remains by tissue sampling; however, CT and MRI can facilitate the differential diagnosis, staging of the extent of disease, and pre-operative planning for paranasal sinus anatomic variants that could complicate surgical resection. Our case supports the literature evaluation of imaging findings and treatment, supporting the current evaluation and treatment of GPC, which remains underrepresented in the medical literature. Our goal is to contribute to the data pool for this rare disease to help make guidelines to facilitate future diagnosis and management. Case reports are the means by which we accumulate data for rare diseases.

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