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Glomangiopericytoma simulating an intracavernous meningioma

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Abstract

Background: Glomangiopericytoma is an uncommonly encountered tumor of the nose and paranasal sinuses, accounting for <0.5% of all sinonasal tumors. Extension of these lesions to the anterior or middle cranial fossa is rare. When this occurs, diagnosing glomangiopericytoma is extremely challenging, as it is often confused with other anterior skull base tumors.

Case Description: We report a case of a giant glomangiopericytoma localizing into the cavernous sinus in a 48-year-old female who presented with mild left-sided ptosis for 48 months. The lesion simulated an intracavernous meningioma on preoperative imaging. An expanded endoscopic endonasal approach was used to debulk the portion of the lesion in the medial compartment of the cavernous sinus. Postoperatively, the patient's ptosis resolved completely, and no new deficits were sustained.

Conclusion: This is the only case of glomangiopericytoma localizing solely to the cavernous sinus reported to date.

Key Words: Cavernous sinus, endoscopic approach, glomangiopericytoma, meningioma, sinonasal-type hemangiopericytoma

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INTRODUCTION

Glomangiopericytoma is an extremely rare tumor of the nose and paranasal sinuses, accounting for <0.5% of all sinonasal tumors.[15] These tumors were previously considered under the entity of hemangiopericytomas. However, due to their distinct clinical and histological features, as well as their lower malignant and metastatic potential, they were classified independently as sinonasal-type hemangiopericytoma (glomangiopericytoma).[10,15,34] Glomangiopericytomas arise from the pericytes of Zimmerman, modified smooth muscle cells surrounding the small blood vessels of the sinonasal region.[12,23] The majority of these tumors tend to remain localized within the sinonasal regions. However, some of these tumors may grow and extend into the anterior skull base region, making a preoperative diagnosis of these tumors extremely challenging. Herein, we report the first case of a giant glomangiopericytoma occupying the cavernous sinus in a 48-year-old female presenting with 4 years history of mild left sided ptosis. Moreover, a literature review on glomangiopericytomas is presented.

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CASE REPORT

History and examination

A 48-year-old female presented with a very mild left sided ptosis for 48 months. She denied any symptoms suggestive of increased intracranial pressure or endocrinological abnormalities. Her past medical and family histories were noncontributory.

Physical examination revealed a very mild left sided ptosis without any ophthalmoplegia. The remainder of her examination was unremarkable.

Imaging

Imaging which included a computed tomography (CT) scan and a magnetic resonance imaging (MRI) revealed a large sellar/parasellar/intracavernous lesion that avidly enhanced. The lesion displaced the pituitary gland laterally to the right side as well as the left parasellar internal carotid artery inferiorly [Figure 1]. The differential diagnosis included: Meningioma, hemangiopericytoma, and cavernous hemangioma.

Surgical management

To obtain tissue diagnosis and decompress the medial compartment of the cavernous sinus, an endoscopic endonasal transellar/trancavernous approach was undertaken. Intraoperatively, the lesion proved to be extremely hemorrhagic. Eight units of blood were transfused intraoperatively. Medial decompression of the cavernous sinus was successfully accomplished.

Histopathological findings

The lesional tissue showed connective vascular arrangements in a kind of honeycomb structure, with a relatively thin walls consisting of endothelial cells and fibroblast-like cells in the stroma. Mitotic figures and necrosis were not seen. In the vascular spaces neutrophils were found, but hardly erythrocytes. The vascular walls did not contain elastin, or collagen bundles. On immunohistochemical studies, the specimen was negative for CD34 and positive for smooth muscle markers (smooth muscle actin [SMA], muscle specific actin [MSA], and desmin) and vimentin in the stroma only. CD34, ETS-related gene (ERG), and factor VIII were strongly positive in the endothelium. Based on these findings a diagnosis of glomangiopericytoma was established [Figure 2].

Postoperative course

Postoperatively, the patient's ptosis resolved completely. She did not develop any new neurological symptoms. Postoperative imaging confirmed successful medical decompression of the cavernous sinus [Figure 3]. The patient opted to not have radiation therapy at the present time. Follow-up MRI at 5 months postoperation revealed stable tumor with no interval growth/progression [Figure 4]. On her last follow-up visit 15 months postoperation, the patient was doing well with only mild intermittent dizziness and no neurological deficits.

DISCUSSION

Glomangiopericytomas are uncommonly encountered neoplasms of the sinonasal region with a characteristic perivascular myoid phenotype.[4] These tumors constitute <0.5% of all sinonasal neoplasms.[15] These tumors can occur at any age with a peak incidence in the seventh decade of life and slight female predominance.[9,30,32]

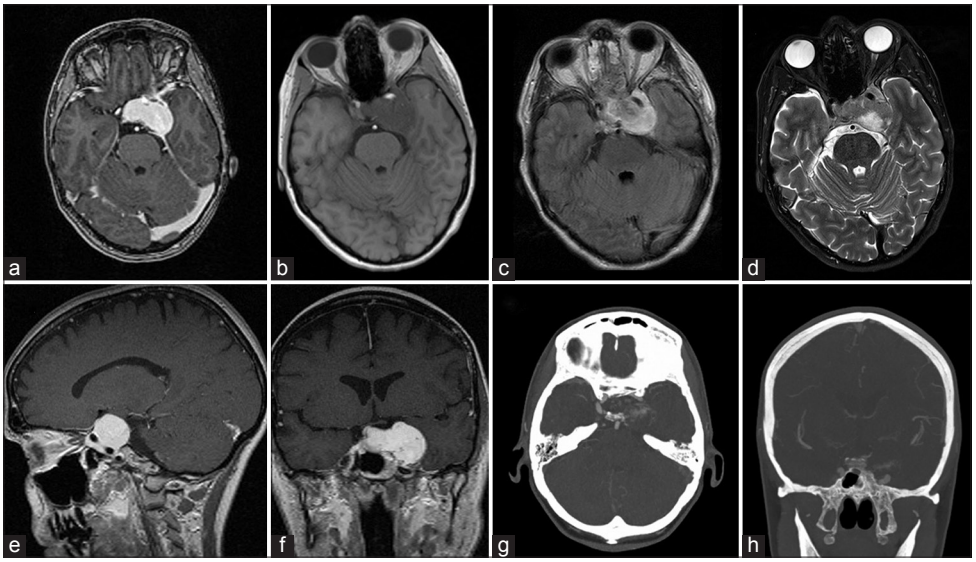

Figure 1: Preoperative imaging of the glomangiopericytoma. Axial T1 magnetic resonance imaging with gadolinium (a), T1 without gadolinium (b), flair (c), T2 (d), sagittal (e), and coronal (f) T1 with gadolinium depicting the tumor in the left cavernous sinus simulating an intracavernous meningioma. Axial (g) and coronal (h) computed tomography angiography demonstrating low vascular flow to the tumor in the left cavernous sinus

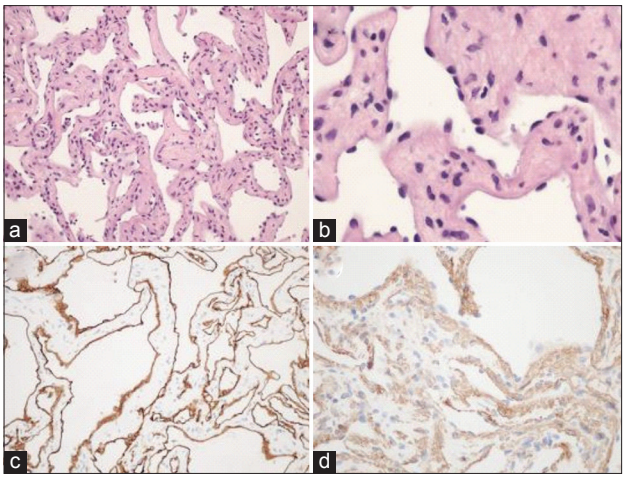

Figure 2: Microscopic and immunohistochemical examination of the resected specimen (a and b). Low and high power microscopic examination with H and E stain demonstrating connective vascular arrangements in a kind of honeycomb structure, with a relatively thin walls consisting of endothelial cells and fibroblast-like cells in the stroma. Immunohistochemical examination of the specimen showed diffuse and strong positivity to CD34 in the endothelium alone (c) and muscle-specific actin in the stroma (d)

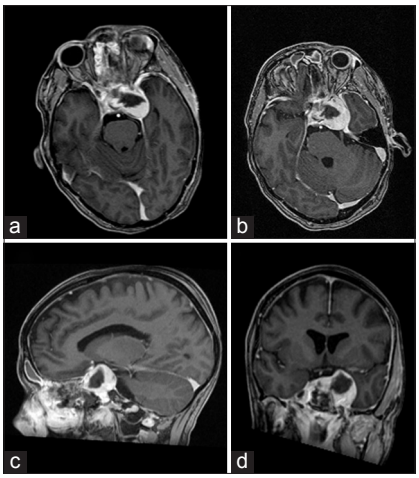

Figure 3: Immediate postoperative imaging after endoscopic tumor debulking. Axial (a and b), sagittal (c), and coronal (d) T1 magnetic resonance imaging with gadolinium depicting residual tumor in the left cavernous sinus with expected postoperative changes

The majority of these tumors are localized to the nasal cavity, with only a few cases reporting primary paranasal sinus involvement.[30] Therefore, glomangiopericytoma tends to produce symptoms localized to these locations. Nasal obstruction, epistaxis, unilateral nasal mass or polyp, sinusitis, headache, and difficulty breathing are the most common symptoms encountered in patients with glomangiopericytomas.[3,6,30] The patient in our case was in her fifth decade with an unusual presentation of mild left-sided ptosis for 4 years. Only two cases of glomangiopericytoma involving the skull base have been reported in the literature [Table 1]. The first case involved the anterior skull base secondarily to glomangiopericytoma arising from the nasal and paranasal sinuses, while in the second case the glomangiopericytoma arose directly from the skull base region.[21,28] To the best of our knowledge, our patient is the first case of glomangiopericytoma localizing solely to the left cavernous sinus.

The development of glomangiopericytomas within the sinonasal region remains elusive. Although many factors including trauma, hypertension, pregnancy, and long-term corticosteroids use have been proposed as the etiological agents for the development of these tumors, no common consensus have been reached regarding the association of these etiological factors and the development of glomangiopericytoma.[2,15,35]

Diagnosing glomangiopericytoma is challenging, as it is frequently confused with other sinonasal lesions. Thus, endoscopic evaluation and radiological imaging are essential to delineate the tumor location, extension, and guide surgical intervention. On MRI, these tumors appear isointense on T1-attentuated imaging and isointense-to-hypointense on T2-attenuated images.[22] After the administration of an intravenous contrast or gadolinium contrast, these tumors appear as enhanced soft tissue masses on CT scan and MRI.[20,22] The utility of angiography in glomangiopericytomas is debatable. Some authors advocate for its use for its value in guiding the preoperative surgical planning and embolization in order to decrease the risk of intraoperative tumor bleeding.[6,25,26] Our patient presented with a giant lesion primarily involving the left cavernous sinus. On imaging, the lesion was simulating a meningioma and our differential diagnosis at that time included meningioma, hemangiopericytoma, and cavernous hemangioma.

On gross examination, these tumors may appear as solid, firm, red or fleshy, soft, and hemorrhagic polypoid edematous masses of variable sizes (mean 3.1 cm; range 1–8 cm).[30] Microscopically, glomangiopericytomas demonstrate a diffuse, subepithelial proliferation of uniform, bland, and closely packed spindle cells growing in different fashions (i.e., fascicular, whorled, or storiform).[8,10,30] Variable sized vascular channels, forming a distinct network, with perivascular hyalinization around the small vessels can often be seen microscopically.[10,30] The nuclei are uniform and equally spaced. The cell cytoplasm is moderately eosinophilic, and the cell membranes are indistinct giving the tumor a syncytial appearance.[8,30] Mitotic rate and nuclear pleomorphism are minimal or absent, with the majority of these tumors lacking necrosis.[8,30,33] However, areas of squamous metaplasia or ulceration can rarely be seen with these tumors.[9,10,30] In addition, giant cells, clear cells, or myxoid degeneration can be depicted in a small number of glomangiopericytomas.[30]

The tumor cells are strongly positive for actin and vimentin on immunohistochemical staining.[10,21,30]

Occasionally, tumor cells are positive for SMA, laminin, factor XIIIa, MSA, and D2-40.[8,11,13,30] In addition, tumor cells are usually negative for S100, desmin, cytokeratin, Bcl-2, factor VIII, CD34, CD99, and CD117.[3,10]

It is therefore important to include glomangiopericytoma in the differential diagnosis of spindle cell tumors of the sinonasal region, which also encompass solitary fibrous tumors, glomus tumors, lobular capillary hemangiomas, angiofibromas, and leiomyomas.[8,10] The key characteristics that help in differentiating between solitary fibrous tumors and glomangiopericytoma include the presence of coarse collagen bundles and overlapping cells arranged haphazardly, as well as the positive immunohistochemical staining for CD34.[10] The tumor cells in glomangiopericytomas can be differentiated from neoplastic cells of glomus tumor by their syncytial appearance with the lack of cell borders and eosinophilic cytoplasm.[8,33] The presence of inflammatory cells, red blood cell extravasation, absence or mild nuclear atypia and mitotic figures, and the lack of necrosis can also help in differentiating glomangiopericytomas from glomus tumors.[33] Lobular capillary hemangioma usually demonstrates surface ulceration and do not grow in a fascicular fashion.[31] Moreover, the tumor cells usually exhibit vascular endothelial differentiation with CD31 and CD34 positivity and perivascular actin positivity on immunohistochemical examination.[10] In angiofibroma, the cells are stellate in shape with abundant stromal collagen and prominent vascular stroma.[10] The tumor cells in angiofibroma stain positively for β-catenin and androgen receptor.[1,18] Leiomyoma demonstrates a prominent fascicular growth pattern with the tumor cells having fibrillary eosinophilic cytoplasm and cigar-shaped nuclei with coarse chromatin. The tumor cells of leiomyoma stain positively for desmin.[6,10]

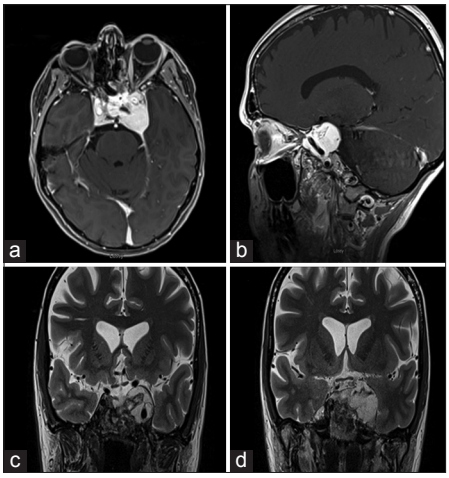

Figure 4: Postoperative imaging 5 months after tumor debulking. Axial (a) and sagittal (b) T1 magnetic resonance imaging with gadolinium and coronal T2 (c and d) showing stable tumor with no interval growth/progression

Various treatment modalities have been reported for the management of glomangiopericytomas. Some authors advocate for conservative management in the form of tumor debulking and frequent endoscopic surveillance, especially when the tumor extends into the anterior skull base.[29] Others, however, believe that the optimal management for these tumors involves radical resection of the tumor along with its extensions.[6,25,26] They believe that it is important to achieve clear margins in order to reduce the risk of tumor recurrence since these tumors are generally resistant to radiation therapy.[6,25,26] Craniofacial resection, midfacial degloving, and lateral rhinotomy have been described in the literature for the management of these tumors. However, these approaches have been associated with high incidences of morbidity and complication rates.[17,25,29]

With the recent advancements in the endoscopic techniques, some authors consider endoscopic surgery as the modality of choice for tumors involving the sinonasal and anterior skull base regions.[21] The improved visualization of the tumor with endoscopic approaches allows for complete and safe resection of the tumor. In addition, endoscopic surgery is associated with better esthetic outcomes and fewer complication rates and hospital stay.[27] Cerebrospinal fluid leakage is the most commonly encountered complication of endoscopic surgery, occurring in 5% of the cases.[27] Such complication can be managed adequately by reoperation to reposition

Table 1: Clinical information of patients with skull base glomangiopericytoma

Author	Age	Gender	Signs and symptoms	Duration	Tumor location	Treatment	Follow-up
Oosthuizen *et al*.[21]	32	Female	Nasal obstruction, anosmia, severe frontal headaches, and right-sided proptosis	6 months	Right nasal cavity with involvement of the sphenoid sinus and anterior skull base	Complete endoscopic excision with modified Lothrop's procedure	18 months
Sun *et al*.[28]	55	Female	Right parotid area tumor, intermittency pain, and foreign body sensation in right pharyngeal portion	2 months	Right skull base region	Submandibular incision with angle of mandible cut-off	
Present case	48	Female	Mild left sided ptosis	48 months	Left cavernous sinus	Medial decompression via an endoscopic endonasal transellar/ trancavernous approach	15 months

the vascularized nasoseptal flap and with the addition of a lumbar spinal drain. In our case, the patient underwent decompression of the medial compartment of the cavernous sinus via an endoscopic endonasal transellar/trancavernous approach. The patient did not experience any intra-or post-operative complications.

As opposed to hemangiopericytomas, glomangiopericytoma generally behave in an indolent manner, with extremely low malignant and metastatic potentials.[6] The prognosis of patients with glomangiopericytoma following complete surgical resection of the tumor is excellent and favorable.[6,29] The estimated 5-year survival is >90% after complete tumor resection[3,30] The completeness of the surgical resection is considered the most important factor to influence the recurrence of the tumor.[6,10,21] In the case of our patient, the extension of the tumor to the lateral compartment of the cavernous sinus and the absence of significant preoperative ophthalmoplegia (other than very mild ptosis) precluded radical resection of the tumor.

The estimated recurrence rate for glomangiopericytoma ranges from 7% to 40%; recurrence can be managed by additional surgery and/or adjuvant radiotherapy.[9,10,11,16,19,30,33,34] In addition to incomplete resection, other factors that have been reported to influence the recurrence rate include tumor size more than 5 cm, osseous invasion, severe nuclear pleomorphism, and high mitotic rate.[6,10,21] Due to the possibility of tumor recurrence, close patient surveillance with clinical and radiographic follow-up is mandatory for those patients.[3]

The role of radiation therapy in the management of residual tumors or primary lesions not amenable to surgical resection is still controversial. Glomangiopericytomas are generally radiation resistant tumors.[6,12,25,26] However, various studies have shown radiotherapy to be effective in unresectable and/or residual lesions, while others have failed to demonstrate any significant benefit.[7,22,30,33] Therefore, radical resection of glomangiopericytoma with clear margins remains the mainstay of treatment, as radiation therapy has not yet proven to improve survival.[5,14,24] The patient in this report has decided not to have radiation or further surgery at the present time.

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Conflicts of interest
There are no conflicts of interest.

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