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Bilateral visual disturbances caused by a glomus vagale: illustrative case

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BACKGROUND A glomus vagale tumor is an infrequent paraganglioma primarily characterized by auditory symptoms, cranial nerve involvement, or autonomic symptoms. However, visual involvement is not commonly observed, and to date, no cases have been reported in the literature.

OBSERVATIONS The case involves a 62-year-old female patient with a history of right carotid body tumor resection. She presented to the emergency department with a sudden decrease in visual acuity and bitemporal hemianopsia, accompanied by a left parietal headache. Initial brain magnetic resonance imaging (MRI) revealed a pituitary macroadenoma, which was completely resected. However, postoperatively, the patient developed left amaurosis. Subsequent brain MRI showed the presence of hemostatic material mixed with blood in the sellar region, causing displacement of the optic chiasm. A repeat intervention was performed, identifying bleeding from both cavernous sinuses. Head and neck angiography demonstrated a right glomus vagale tumor with abundant blood drainage into the right cavernous sinus. Embolization of the glomus vagale tumor was performed, resulting in no further bleeding and improvement of symptoms.

LESSONS The aim of this case report is to describe a rare occurrence of bilateral visual disturbances caused by bleeding in both cavernous sinuses due to venous hypertension caused by a right glomus vagale tumor.

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KEYWORDS glomus vagale; visual disturbances; venous hypertension; bleeding; sella turcica

Glomus vagale tumors are rare neoplasms that fall under the category of paraganglionic tumors. They account for less than 5% of glomus tumors occurring in the head and neck region.[1] Glomus tumors are classified based on their anatomical location, which includes carotid body tumors, glomus jugulare tumors, glomus vagale tumors, and glomus tympanicum tumors.[2] Furthermore, head and neck paragangliomas arise in a specific order of decreasing frequency. They primarily originate from the carotid body, followed by the jugular bulb, the vagus nerve, the tympanic branch of the glossopharyngeal nerve or the auricular branch of the vagus nerve, and the cervical sympathetic chain.[3]

Most paragangliomas are classified as slow-growing tumors and have a higher prevalence in females during their sixth and seventh decades of life. Glomus vagale tumors, because of their mass effect, can affect neighboring structures near the jugular foramen. This can result in a variety of symptoms, including facial paralysis, hearing loss, dysphagia, dysphonia, and other related manifestations.[4]

Given the inaccessible location for examination, the insidious growth of these lesions, and their low prevalence, patients usually present with advanced disease at the time of diagnosis; thus, paragangliomas can become an urgent and potentially life-threatening condition.[5]

Surgical treatment of paragangliomas is the only therapeutic option that can provide complete and immediate removal of the tumor but given their hypervascularization and proximity to critical neural structures, surgery may entail significant morbidity.[1] Thus, early recognition of paragangliomas that require treatment increases the chances of complete tumor removal and decreases the morbidity associated with surgery or radiosurgery.[3]

Given the challenges in diagnosing paragangliomas and the significant surgical morbidity associated with their treatment, it is noteworthy that no cases have been reported thus far regarding glomus vagale with drainage to the cavernous sinus resulting in bleeding in the sella turcica and associated visual symptoms. Here, we present

ABBREVIATIONS CT = computed tomography; MRI = magnetic resonance imaging.

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the case of a 62-year-old female patient who had a previous history of right carotid body tumor resection. The patient presented to the emergency department with visual disturbances that were later found to be associated with a pituitary macroadenoma. Despite undergoing surgical correction of the lesion, the patient experienced a worsening of visual symptoms. Subsequent extension studies were performed, confirming the presence of a right glomus vagale tumor draining into the ipsilateral cavernous sinus, which ultimately led to venous hypertension in both cavernous sinuses and bleeding in the sella turcica.

Illustrative Case

A 62-year-old female patient with a history of resected right carotid body tumor consulted the emergency department for a clinical symptom of 3 days of bilateral blurred vision associated with intense gradual headache in the left parietal region radiating to the biparietal and occipital region associated with photophobia. On neurological examination, she presented with bitemporal hemianopsia with an alteration of visual acuity.

Sella turcica magnetic resonance imaging (MRI) was conducted (Fig. 1), revealing a pituitary macroadenoma with indications of optic chiasm compression. The patient underwent transsphenoidal resection of the pituitary macroadenoma. During the surgery, minimal arterial bleeding was observed on the right side, which was effectively managed without complications. Following the procedure, the patient was transferred to the neurological intensive care unit for postoperative monitoring, and a satisfactory improvement in the initial symptoms was observed. At 48 hours postoperatively, the patient presented with a progressive decline in visual acuity in the left eye, eventually leading to amaurosis. Subsequent sella turcica MRI (Fig. 2) demonstrated intrasellar bleeding extending into the suprasellar region and causing superior displacement of the optic chiasm. No evidence of residual tumor that could account for the bleeding was found. As a result, the patient was promptly transferred to the operating room for further intervention. During surgical exploration of the sella region, hemostatic material was removed, leading to the sudden onset of profuse arterial bleeding originating from both cavernous sinuses. The affected area was promptly resealed, and the patient underwent cerebral pan-angiography, which revealed a highly vascularized mass in the high cervical region consistent with a glomus vagale tumor. Importantly, retrograde venous flow from the tumor to the cavernous sinus was observed (Figs. 3 and 4, Video 1). Embolization of the glomus vagale tumor was performed, achieving 90% occlusion of the lesion (Fig. 5, Video 2). The patient was then transferred to the operating room for removal of the hemostatic material, where slight arterial bleeding was observed compared to the previous surgery; therefore, hemostasis was performed. The postoperative period was without complications, revealed 90% improvement in visual acuity, and was without alterations of the campimetry.

VIDEO 1. Clip showing head and neck angiography. No arterial lesions of the internal carotid, external carotid, or branches are evident. There are signs of venous hypertension of the venous bundle around the odontoid process and intercavernous sinuses by prominent drainage of the right glomus vagale. Click here to view.

Management was completed with radiotherapy to avoid sellar and intracranial bleeding. The patient was discharged from the hospital 12 days after the postoperative period, to follow ambulatory control at 1 month, at 6 months, and then yearly with MRI. In the last postoperative control performed 6 months after discharge, complete resolution of visual symptoms and normal campimetry was evidenced.

VIDEO 2. Clip showing postembolization head and neck angiography. Dimethylsulfoxide and Onyx were administered until occlusion of the glomus vagale was achieved. Occlusion of approximately 90% of the glomus vagale with the disappearance of venous hypertension to the cavernous sinus and intercavernous veins. Click here to view.

Patient Informed Consent

The necessary patient informed consent was obtained in this study.

Discussion

Observations

Head and neck paragangliomas are slowly growing benign tumors, originating from specialized neural crest cells, and their doubling time is estimated to be around 13.8 years, with an annual growth rate of 0.79 mm/yr.[6] They are locally aggressive given their tendency to infiltrate various connective tissue planes and erode bone, and approximately 80% of paragangliomas are sporadic and 20% exhibit a hereditary factor.[7]

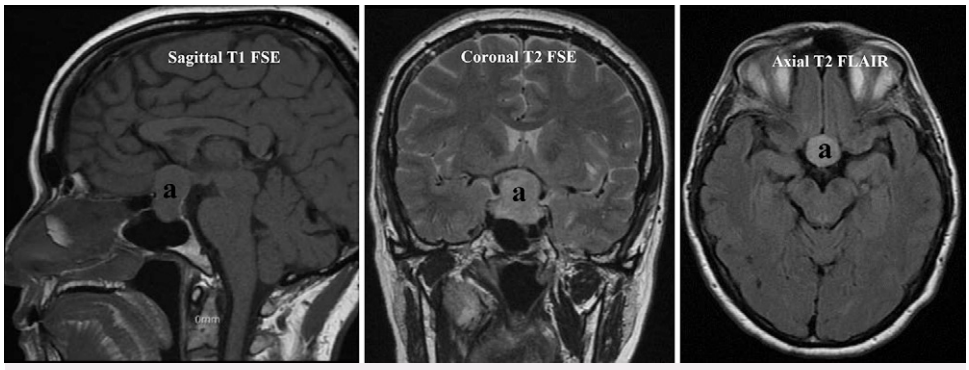

FIG. 1. Brain MRI scans showing a pituitary macroadenoma (*a*) with small foci of bleeding and signs of compression of the optic chiasm. FLAIR = Fluid-attenuated inversion-recovery; FSE = fast spin echo.

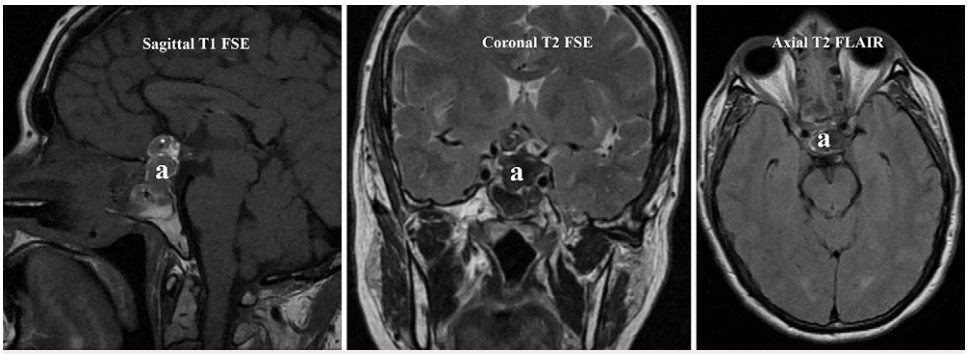

FIG. 2. Brain MRI scans showing blood and hemostatic material (*a*) in the sella turcica with suprasellar extension and superior displacement of the optic chiasm.

The glomus vagale tumor is frequently associated with middle ear involvement and tends to spread through the duct of the hypothympanic air cells, around the vagale bulb, inferior petrosal sinus, and carotid artery to the jugular foramen and posterior fossa; only 1%–3% present active secretion of catecholamines.[8,9] Glomus vagale tumors draining into the cavernous sinus and generating arterial-type bleeding are exotic; to our knowledge, this is the first reported case.

The glomus vagale tumor can remain silent for a long period of time. Symptoms can be otologic (unilateral pulsatile tinnitus, hypoacusis, and vertigo) and neurological (dysphagia, dysphonia, lingual paralysis, and shoulder weakness) and occasionally can cause tachycardia, headache, arterial hypertension, and diarrhea, among others.[9] Thus, the visual symptomatology manifested by the patient is an atypical presentation of a glomus vagale tumor that caused great deterioration in the patient's physical condition.

The principal evaluation of the glomus tumor is thin-slice computed tomography (CT) where an enhanced mass and bone windows may reveal erosion of the skull base. In this sense CT allows us to identify the location of the glomus vagale tumor. On brain MRI the glomus vagale tumor can be seen as a strongly enhanced soft tissue mass. In addition, a typical salt and pepper pattern can be demonstrated. MRI shows tumor hypervascularity, extension along the neural foramina, and multicentricity, making it the first study to be performed to evaluate a glomus vagale tumor.[10,11] Finally, angiography allows one to differentiate paragangliomas from other pathologies and allows one to identify the vessels feeding the lesion that can later be embolized. Among the arteries that generally feed paragangliomas are the ascending pharyngeal artery, the occipital artery, the external carotid artery, the internal carotid artery, the vertebrobasilar system, and the posteroinferior cerebellar artery.[6]

In the present case, the glomus vagale tumor demonstrated preferential venous drainage to the paravertebral veins and the pterygopalatine plexus, ultimately draining into the right cavernous sinus. The possibility of intratumoral arteriovenous fistulas may have contributed to the arterialized bleeding observed in the right cavernous sinus, resulting in elevated pressures. This can be analogously compared to a carotid-cavernous fistula that has ruptured into the sella turcica. Additionally, the occurrence of similar bleeding from the contralateral side (left cavernous sinus) can be explained by the connection between the cavernous sinuses through the coronary sinuses.

Regarding treatment, observation is an excellent alternative considering that approximately 65% of glomus vagale tumors remain stable and can sometimes decrease in size. Thus, if follow-up of the lesion is chosen, it should be performed through close monitoring with serial brain MRI with and without intravenous contrast to evaluate the behavior of the lesion[6,12]

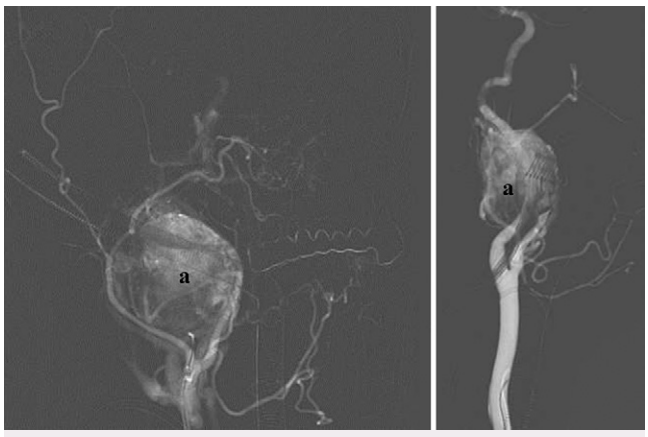

FIG. 3. Head and neck angiography showing evidence of right glomus vagale (*a*).

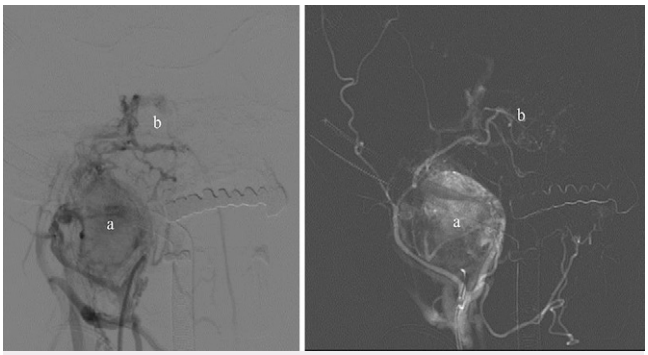

FIG. 4. Head and neck angiography demonstrating clear evidence of the right glomus vagale draining into the cavernous sinus. The images clearly depicted the presence of the glomus vagale (*a*) and its connection to the cavernous sinus (*b*).

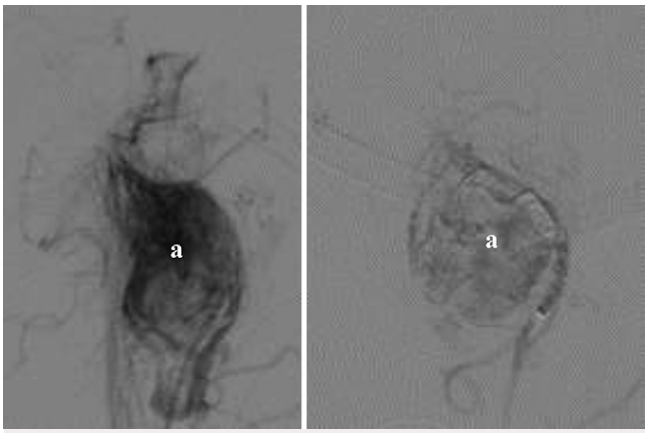

FIG. 5. Comparison of head and neck angiography before embolization (**left**) and after embolization (**right**). Significant occlusion of the glomus vagale is evident. *a:* glomus vagale.

As for a glomus vagale tumor that is symptomatic, leads to significant deterioration in the quality of life, progresses in its growth on images or presents bleeding related to the lesion that can considerably increase mortality, resection of the lesion, radiosurgery, or embolization are the options of choice.[13,14] In cases where the glomus vagale tumor represents vital urgency, as in the present case, surgery is recommended to drain and stop the bleeding; however, surgery is associated with a higher rate of complications and a higher risk of damage to adjacent cranial nerves. Despite these limitations, surgery represents the best strategy for complete removal of the paraganglioma in cases where the lesion is easily accessible surgically.[15]

On the other hand, radiosurgery is useful in the context of bilateral glomus vagale tumors or in tumors in which complete resection of the lesion was not achieved or that are difficult to access because of their location. Studies such as that by Suarez et al.[1] demonstrated that radiosurgery achieves greater tumor control, a lower rate of complications, and a lower number of cranial nerve palsies after the procedure. Thus, radiosurgery represents an excellent option for the treatment of paragangliomas.

Finally, embolization of the lesion is very difficult and is prone to revascularization and does not generate substantial benefit in terms of the relief of clinical symptoms.[16] Thus, endovascular embolization as the only treatment modality is considered palliative. In cases where the aim is to reduce the duration of surgery and intraoperative bleeding, preoperative embolization may be the best option, or in cases where there is a high risk of morbidity and mortality due to the location of the lesion.[6]

In terms of prognosis, the risk of cranial nerve injury is approximately 5%–39%, the stroke rate is 0%–3.5%, and up to 15% of patients cannot return to their preoperative daily activities. In cases in which complications occur, the rate of disability and mortality can be much higher.[6,17]

To the best of our knowledge, this is the first documented case in the literature of bilateral visual disturbances resulting from sella turcica bleeding in both cavernous sinuses due to venous hypertension in the vascular bed caused by a glomus vagale tumor. This exceptional case offers valuable insights into the exploration of potential clinical manifestations and the underlying pathophysiological mechanisms contributing to such complications in glomus vagale tumors.

Lessons

The glomus vagale tumor is a rare lesion typically characterized by stable symptomatology primarily involving cranial nerves, hearing, and autonomic functions. However, in certain complicated cases, it can present with a diverse range of symptoms that are not easily attributed to the lesion. The current case serves as an example of a complex glomus vagale tumor with drainage into the cavernous sinus, leading to bleeding in the sella turcica and resulting in bilateral visual disturbances. This case highlights the atypical manifestations and potential complications associated with glomus vagale tumors.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: all authors. Acquisition of data: Guzman. Analysis and interpretation of data: Garcia Rairan, Jimenez Hakim. Drafting the article: Garcia Rairan, Guzman. Critically revising the article: Garcia Rairan, Jimenez Hakim, Guzman. Reviewed submitted version of manuscript: Garcia Rairan, Jimenez Hakim. Approved the final version of the manuscript on behalf of all authors: Garcia Rairan. Study supervision: Garcia Rairan.

Supplemental Information

Videos

Video 1. https://vimeo.com/829059858.

Video 2. https://vimeo.com/829062517.

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