

Case Report

Presentation with Personality Changes and Tinnitus to a Vascular Intervention: A Rare Case of Carotid Body Paraganglioma

Ahsan Zil-E-Ali¹, Zubair Ahmed², Amber Ehsan Faquih³, Muhammad Ishaq⁴, Muhammad Aadil¹

¹Department of Surgery, Fatima Memorial Hospital, Lahore, Pakistan, ²FMH College of Medicine, Lahore, Pakistan, ³Department of Medicine, Dow University of Health Sciences, Karachi, Pakistan, ⁴Department of Internal Medicine, Marshfield Clinic, Marshfield, WI 54449, USA

Address for correspondence: Department of Surgery, Fatima Memorial Hospital, Shadman, Lahore-54000, Pakistan.



Quick Access Code

How to cite this article:

Zil-E-Ali A, Ahmed Z, Faquih AE, Ishaq M, Aadil M. Presentation with Personality Changes and Tinnitus to a Vascular Intervention: A Rare Case of Carotid Body Paraganglioma. Journal of Medical Research and Innovation. 2018;2(2):e000109.

Doi: 10.15419/jmri.109

Publication History

Received: 04-01-2018
Accepted: 18-03-2018
Published: 19-03-2018

Editor: Dr. Varshil Mehta

Copyright: Zil-E-Ali A, Ahmed Z, Faquih A, Ishaq M, Aadil M. This is an open access article distributed under the terms of the Creative Commons Attribution License CC-BY 4.0., which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Funding: NIL

Conflict of Interest: NIL

Abstract

Carotid body paragangliomas are rare neuroendocrine neoplasms of chromaffin-negative glomus cells. This case report explains an atypical case with unusual presentation and treatment. A healthy smoker technician by profession was brought to the emergency room with coprolalia. The general physical examination did not reveal any information. His history revealed unilateral tinnitus andodynophagia, leading to a consultation by the neurologist with head imaging. Acoustic neuroma was ruled out and the caregiver was asked to elaborate the events mentioned in the history and a psychiatric examination was done. The personality changes were evaluated by the psychiatrist that showed overlapping of delirium and depression. The patient was further examined by a vascular surgeon. After careful revisiting of the history, examination and indication of tender mass in the neck by the patient's vascular surgeon, the diagnosis of carotid body paraganglioma was made which was followed by surgical resection for treatment. Carotid body paragangliomas are very vascular structures and their manipulation in a surgery setting requires expertise. This case presented with personality changes and tinnitus, a very less likely event to occur in a carotid body tumor. The present care report, thus adds on to the literature of carotid body tumors and its presenting symptoms.

Keywords: Carotid body tumor, Paraganglioma, Surgery, Vascular surgery

Introduction

Carotid body tumors, also known as chemodectoma or paraganglioma, are highly vascular glomus tumors that originate from paraganglion cells of carotid body originating at the bifurcation of the carotid artery.^[1] The present literature describes three types of carotid body paraganglioma; familial, sporadic, and hyperplastic.^[2] They attune respiratory and cardiovascular system functions when there are changes in po₂, pco₂, and blood pH.^[3] These are treated surgically and with radiotherapy when surgery is not possible.^[4] These tumors usually present with palpable neck mass, hoarseness of voice, vision changes, high blood pressure, and numbness in the tongue. The present case put forward a case report of a patient with tinnitus and neurological symptoms, including personality changes, which is a very

rare presentation indicating the diagnosis of carotid body paraganglioma.

Case Report

A 44-year-old healthy male smoker working in an industrial facility as a technician (height: 5' 7", weight: 74 kg, body mass index: 25.5) presented at midnight in the emergency department with coprolalia, jumbled conversation, and screaming episodes. The emergency physician carried a general physical examination and evaluated for focal neurological signs, which were negative. On further history from the patient's caregiver, it was known that the patient had changes in sleeping patterns and used to avoid social gatherings for past 1 month. Before this event, almost 3 months back, the patient complained of unilateral tinnitus,odynophagia, and discomfort in the neck. For these symptoms,

an otorhinolaryngologist consult was asked which led to a provisional diagnosis of acoustic neuroma. After consult with the neurologist and head imaging, the patient was clear of any condition or disease and the possibility of neuroma was ruled out.

The patient was shifted from emergency service to an indoor facility and was planned for reevaluation in the morning. After considering the past medical symptoms, the patient was again asked about the tinnitus, on which patient stated it as a “continuous annoying ringing in the right ear.” On asking about alleviating or aggravating factors, the patient stated it as, “in the morning, its better but at work its unbearable.” Regardingodynophagia, the patient told that swallowing solids are more painful than liquids and it has been undulating since the beginning. On asking the discomfort in the neck, patiently pointed a tender site. The caregivers were questioned about various personality changes, about which it was revealed that the patient had extreme personality changes which ranged from conversing with a rude language, social withdrawal, spells of forgetfulness, and multiple episodes of wakefulness in the middle of the night. The patient was also found roaming around the house at odd times. After history, the patient’s systemic examination was carried which did not show any focal abnormality, although the patient was found to be having a high blood pressure 144/98 mmHg with a heart rate of 87/min. Thus, a psychiatric consult was planned.

Initially, the diagnosis of delirium overlapping with depression was made by the psychiatric evaluation and the case was forwarded to the academic board for an expert opinion. Although the patient was conscious but was not well oriented regarding the surroundings. Psychiatrist’s opinion also suggested that the personality changes may not be associated with any major lesion in the brain, but a possibility of low perfusion of certain focal areas can be associated with such symptoms. It was difficult for a particular psychiatric condition so further evaluation was requested. On careful reevaluation of the symptoms, a vascular surgeon consult was requested, which led to magnetic resonance angiography (MRA) and extensive head and neck imaging of the patient. Figure 1 shows sagittal section of computerized tomography (CT) scan revealing a tumor-like mass on the bifurcation of the carotid artery on the right side with compression of the vascular lumen (15.5 mm × 31.5 mm × 34.8 mm).

Figure 2 shows the MRA that helped us explain the vascular nature of the tumor and extent of the growing structure. These images helped in the final diagnosis of carotid body paraganglioma that was relatable with high blood pressure and the probable site of the presence of glomus cells, the primary site of this tumor.

Considering patient’s incapacitated state, the consent was taken from the patient’s wife for a surgical intervention.



Figure 1: Sagittal section of computerized tomography scan showing the mass lesion in the carotid body

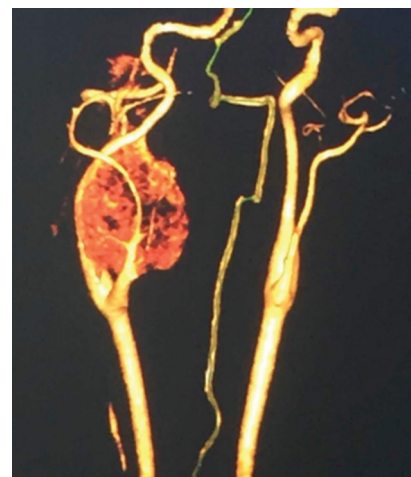


Figure 2: Image of magnetic resonance angiography showing a 3D image of vascular mass at the bifurcation

A vascular surgeon intervened and the patient was held in a supine extended neck position. The surgeon retracted sternocleidomastoid laterally and omohyoid medially. The bifurcation of the common carotid was visualized with a hard mass at the site. Vascular looping was done for the vessels and the tumor was located and excised. Figure 3 shows the technique of approaching this tumor mass and removing it by careful manipulation around this mass. The extracted sample was sent to the pathology for histological analysis that later confirmed it as a paraganglioma or more popularly known as carotid body tumor. The patient was kept under observation, and the caregivers were advised to have a follow-up in a month.

Discussion

These tumors are an asymptomatic palpable neck mass present within the anterior triangle of the neck. On examination, these masses are typically vertically fixed at the bifurcation of the common carotid (Fontaine sign). It is well established that the

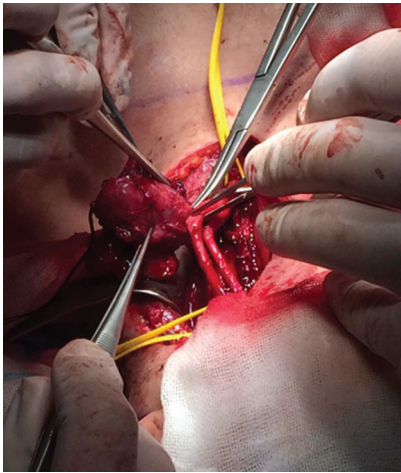


Figure 3: Intraoperative image of demarcating the lesion and using vascular looping technique to control the blood flow in the operating focus

gold standard curative treatment is surgery for carotid body tumors. The first of this kind of intervention was performed by Albert in 1889 with a more focused subadventitial dissection reported by Gordon-Taylor in 1940.^[5,6] Carotid body tumors are slow-growing hypervascular tumors that arise from paraganglionic cells and have chemoreceptor organ in it. The most common site of these tumors is at carotid bifurcation, foramen jugular, middle ear cavity, and in cervical portion. It is recommended that a complete resection should be performed because there is an incidence rate for malignant degeneration to as high as 10%, and in case of bilateral or multiple masses, a staged approach is recommended instead of simultaneously excising it to reduce the neurologic damage and cerebrovascular complications.^[4,7] In our case, the site was very similar to the published data and with the help of the imaging, diagnosis was possible. Carotid bifurcation is the site for paraganglioma and a very characteristic imaging of the tumor is seen in it. However, the presentation was very unique for this case, first, that tinnitus is not reported in the literature for carotid body paraganglioma, although it is a basic symptom for these tumors presenting at jugular vein or in the middle ear cavity due to obvious anatomical location. Presentation with tinnitus for this site is less likely and there is no report in literature similar to it. Second, an association with personality changes is not yet known. There is an ample neurological evidence that suggests the presence of foci in the brain that manage our behaviors, conscious, and unconscious perceptions, but its connection with a carotid body paraganglioma is unique and unheard of. The patient had spells of forgetfulness that may sign toward inadequate blood perfusion to the memory centers but with abusive conversation and social withdrawal which are new to us. This case was unique to be presented with personality changes. In general, the low

perfusion of certain brain areas may be associated with this process, but there is no strong evidence that can actually suggest this process. Although it was evident that no particular organic cause can be associated with tinnitus or personality change, this is important to remember that these tumors often surprise us by presenting with such new symptoms.

Overall, the presentation of these tumors and treatment plans are extensively seen in the literature, but our case is unique of its kind, showing the presentations that are very less likely to happen. It is the first reported case of a paraganglioma of carotid body, presenting with such symptoms.

Conclusion

Carotid body paragangliomas are very vascular structures and their manipulation in a surgery setting requires expertise. Furthermore, the diagnosis requires imaging, preferably a CT scan and angiogram to see the extent of the tumor and plan the surgery accordingly. Its presentation usually ranges from a palpable neck mass to other visual symptoms and blood pressure changes. This case presented with tinnitus, which is very less likely to happen in a carotid body tumor and personality changes, which are also not explainable. The report thus adds on to the literature of carotid body tumors and its presenting symptoms.

References

1. Gaillard F. Carotid Body Tumor. Radiology Reference. Available from: <https://www.radiopaedia.org/articles/carotid-body-tumour>. [Last accessed on 2018 Jan 04].
2. Medscape. Carotid Body Tumors: Background, History of the Procedure, Epidemiology. Available from: <https://www.emedicine.medscape.com/article/1575155-overview>. [Last accessed on 2018 Jan 04].
3. Ridge BA, Brewster DC, Darling RC, Cambria RP, LaMuraglia GM, Abbott WM, et al. Familial carotid body tumors: Incidence and implications. *Ann Vasc Surg* 1993;7:190-4.
4. Davidovic LB, Djukic VB, Vasic DM, Sindjelic RP, Duvnjak SN. Diagnosis and treatment of carotid body paraganglioma: 21 years of experience at a clinical center of Serbia. *World J Surg Oncol* 2005;3:10.
5. Sajid MS, Hamilton G, Baker DM, Joint Vascular Research Group. A multicenter review of carotid body tumour management. *Eur J Vasc Endovasc Surg* 2007;34:127-30.
6. Gordon-Taylor G. On carotid tumours. *Br J Surg* 1940;28:163-72.
7. Shamblin WR, ReMine WH, Sheps SG, Harrison EG Jr. Carotid body tumor (chemodectoma). Clinicopathologic analysis of ninety cases. *Am J Surg* 1971;122:732-9.