

## Case Report

# Carotid body tumor imitator: An interesting case of Castleman's disease

Hakeem J. Shakir<sup>1,2</sup>, Sara M. Diletti<sup>2</sup>, Alexandra M. Hart<sup>3</sup>, Joshua E. Meyers<sup>1,2</sup>, Travis M. Dumont<sup>1,2,4</sup>, Adnan H. Siddiqui<sup>1,2,5,6,7</sup>

Departments of <sup>1</sup>Neurosurgery and <sup>2</sup>Radiology, Jacobs School of Medicine and Biomedical Sciences, State University of New York, University at Buffalo, <sup>3</sup>Department of Neurosurgery, Kaleida Health, Gates Vascular Institute, Buffalo General Medical Center, <sup>4</sup>Department of Pathology, Kaleida Health, Buffalo General Medical Center, State University of New York, University at Buffalo, <sup>5</sup>Toshiba Stroke and Vascular Research Center, State University of New York, University at Buffalo, <sup>6</sup>Jacobs Institute, Buffalo, New York, <sup>7</sup>Department of Neurosurgery, Department of Medical Imaging, Division of Surgery, University of Arizona, Tucson, Arizona, USA

E-mail: Hakeem J. Shakir - [hshakir@ubns.com](mailto:hshakir@ubns.com); Sara M. Diletti - [saradile@buffalo.edu](mailto:saradile@buffalo.edu); Alexandra M. Hart - [amhart@buffalo.edu](mailto:amhart@buffalo.edu); Joshua E. Meyers - [jmeyers@ubns.com](mailto:jmeyers@ubns.com); Travis M. Dumont - [tdumont@surgery.arizona.edu](mailto:tdumont@surgery.arizona.edu); \*Adnan H. Siddiqui - [asiddiqui@ubns.com](mailto:asiddiqui@ubns.com)

\*Corresponding Author

Received: 23 March 15 Accepted: 18 June 15 Published: 07 December 15

**This article may be cited as:**

Shakir HJ, Diletti SM, Hart AM, Meyers JE, Dumont TM, Siddiqui AH. Carotid body tumor imitator: An interesting case of Castleman's disease. *Surg Neurol Int* 2015;6:181. <http://surgicalneurologyint.com/Carotid-body-tumor-imitator:-An-interesting-case-of-Castleman's-disease/>

Copyright: © 2015 Shakir HJ. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

## Abstract

**Background:** There are very few reports in the literature of Castleman's disease affecting the carotid artery and a single previous report of a case of Castleman's disease of the neck originally mistaken as a carotid body tumor.

**Case Description:** We describe a rare case of Castleman's disease, manifesting with classic radiographic hallmarks of a carotid body tumor. The postoperative pathologic examination identified the resected mass as Castleman's lymphadenopathy. The management of this particular case is discussed, and the findings are highlighted.

**Conclusions:** We present a unique case of a tumor initially and incorrectly diagnosed as a carotid body tumor. However, after comprehensive treatment with endovascular and surgical modalities and subsequent pathologic examination, the diagnosis of this rare entity was made.

**Key Words:** Carotid body tumor, Castleman's disease, lymphadenopathy, paraganglioma

**Access this article online****Website:**

[www.surgicalneurologyint.com](http://www.surgicalneurologyint.com)

**DOI:**

10.4103/2152-7806.171239

**Quick Response Code:**

## INTRODUCTION

Carotid body tumors, typically found at the carotid bifurcation, are rare entities among head and neck neoplasms. Most (60–70%) head and neck paragangliomas occur in the region of the carotid bifurcation.<sup>[5]</sup> Additional tumors in this region that may be indistinguishable radiographically from carotid body tumors include glomus vagale tumors arising from the vagus nerve and schwannomas arising from any sensory component of the cervical cranial nerves. Carotid body tumors are histopathologically characterized as paragangliomas. Only

1–2% of all carotid body tumors are secretory, releasing catecholamines.<sup>[8]</sup>

The patient described in this report presented with a nonsecretory tumor causing widening of the carotid bifurcation. Although observation for small, nonsecreting tumors may represent an option, typically surgical intervention is required for cases involving the carotid artery. Without intervention, progressive symptomatology due to damage of the neighboring vagus and hypoglossal nerves in addition to compromise of the integrity of the carotid artery itself should be expected.<sup>[5]</sup> In our patient, a diagnosis of carotid body tumor was made.

Castleman's lymphadenopathy was not suspected until after the pathologic examination. We aim to discuss our course of action and postoperative findings as well as the characteristics of this rare and frequently asymptomatic disease.

## CASE REPORT

### Examination

A 46-year-old African-American man presented with an enlarging, tender, left-sided neck mass. The initial evaluation consisted of magnetic resonance and computed tomographic (CT) imaging that demonstrated a 7-cm mass with T1 signal and T2 signal contrast enhancement [Figure 1]. On the CT scan, the mass was revealed to be hyperdense. A presumptive diagnosis of nonsecretory paraganglioma was made after catecholaminergic products were absent in serum and urine analyses. Given the size and progressive growth of the mass and symptomatic status of the patient, resection was planned.

After informed consent was obtained, digital subtraction angiography and balloon occlusion testing were performed. Angiography confirmed a 7-cm vascular tumor with very thin vascular channels arising directly from the walls of the common and internal carotid arteries. There was no pedicle large enough to be suitable for embolization [Figure 2]. This pattern was unusual because paragangliomas are extremely vascular with very well developed vascular channels that usually arise off the external carotid artery and have a pattern of arborization that is highly amenable to endovascular embolization. In this case there was the appearance that the vascular channels were direct offshoots of the vasa vasorum of the

affected vessels. In addition, there was irregularity and dilatation in the region of the carotid bulb suggestive of carotid wall involvement, which is possible but rare in paragangliomas. The patient tolerated the balloon occlusion test; and because the tumor was considered to be invading the wall of the carotid artery, we elected to endovascularly sacrifice the internal carotid artery (ICA) in preparation for surgical resection. The purpose of this sacrifice was to starve the tumor of its blood supply and minimize the degree of intraoperative bleeding. The endovascular sacrifice of the ICA was accomplished without incident. One-week later, the patient was taken to the operating room for resection.

### Operation

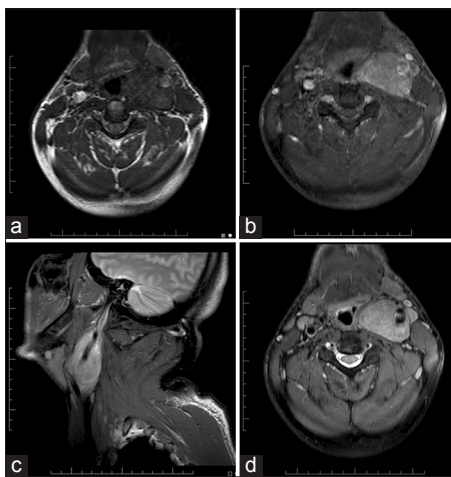
Surgical exposure of the carotid sheath revealed a large mass in the region of the carotid bifurcation. Exposure from the skull base to the omohyoid muscle allowed sequential separation of the jugular vein and the vagus, accessory, and hypoglossal nerves from the mass. The entire carotid bifurcation was dilated without separation of planes between the internal and external carotid arteries, carotid bulb, and tumor. The common, internal, and external carotid arteries were subsequently ligated away from the tumor; and the tumor and neighboring arterial segments were resected *en bloc* and sent for pathologic examination. Four distinctive lymph nodes appreciated during neck dissection were additionally submitted for examination. After resection of the mass, the sympathetic chain ganglia below the carotid sheath became apparent and appeared uninvolved.

### Postoperative course

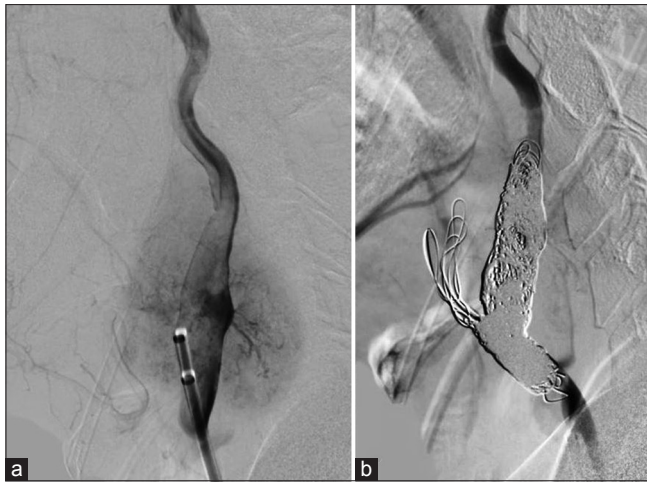
The patient woke up without deficits; however, on the second postoperative day, he developed mild hoarseness and swallowing difficulty with an enlarging neck mass. He was taken emergently for the evacuation of a neck hematoma. No active bleeding sites were noted. Thereafter, the patient made an uneventful recovery and was discharged home without evidence of cranial neuropathies.

### Pathological findings

Gross examination of the *en bloc* specimen revealed a segment of artery surrounded by a mass measuring 4.8 cm × 4.0 cm × 2.4 cm. The tumor had a tan, myxoid surface with yellow tissue, and hemorrhagic changes next to the thrombosed arterial segment. Microscopic examination revealed a large circumscribed cervical lymph node with a large elastic artery coursing through it. The lymph node showed distinctive features including follicles composed of concentric rings of mantle zone lymphocytes around germinal centers as well as prominent vascularity of the germinal centers, often showing a single penetrating arteriole [Figure 3a and b]. Immunostaining patterns from the lymph node revealed CD23, cyclin D1, BCL2, CD20, CD3, CD138, CD56,



**Figure 1:** Axial T1-weighted magnetic resonance images without (a) and with (b) gadolinium demonstrating a 7-cm homogeneously enhancing left neck mass circumferentially encasing the left internal carotid artery, extending from C2 to C4. Sagittal (c) and axial (d) T2-weighted magnetic resonance images, again showing the left neck mass encasing the left internal carotid artery

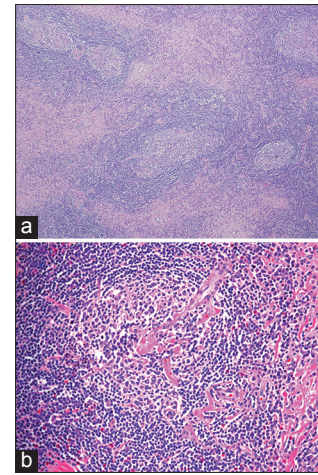


**Figure 2: Mid-to-late arterial phase of a left common carotid artery injection before (a) and after coil embolization (b) of the common, internal, and external carotid showing reduced vascularity of the mass**

kappa/lambda, CD30, and CD15, which excluded the diagnosis of lymphoma; and Congo red staining excluded amyloidosis. Additional adjacent benign lymph nodes submitted for microscopic examination showed some of the above-mentioned features of Castleman's lymphadenopathy.

## DISCUSSION

Castleman *et al.*<sup>[1]</sup> first distinguished localized mediastinal lymph node hyperplasia from thymoma in 1956. Benign lymphoproliferative masses are the differentiating factors of Castleman's disease.<sup>[2]</sup> The disease presents as two types: unicentric and multicentric. The unicentric type involves a localized mass, and surgical resection is curative for 90% of these cases. Multicentric Castleman's lymphadenopathy presents as the systemic proliferation of the disease at multiple sites. Our patient displayed the unicentric variety with cervical localization. The unicentric type can be further divided into the hyaline vascular and plasma cell types. The hyaline vascular type of unicentric Castleman's disease is more common and occurs in 90% of cases, including ours.<sup>[2,3,9]</sup> This type is characterized by large lymphoid follicles, vasculature with prominent hyalinization, and circular layers of lymphocytes. The "onion skinning" of the rings of mantle zone lymphocytes and the single penetrating central vessel producing a lollipop-like appearance are characteristic features of the hyaline vascular form of Castleman's disease.<sup>[4]</sup> The remaining 10% are the plasma cell type, characterized by the substantial growth of polyclonal plasma cells in interfollicular areas; and afflicted patients can present with anemia, pyrexia, night sweats, and hyperglobulinemia.<sup>[6,9]</sup> Of the 183 cases of Castleman's disease reported in the literature, most (70%) of these lesions were found in the mediastinum, and



**Figure 3: (a) Hematoxylin and eosin (H and E) stain at low-power magnification (x40) demonstrating lymph node hyperplasia with vascular proliferation magnification. (b) H and E stain at high-power magnification (x200) showing follicle containing a centrally located single prominent radiating vessel and concentric rings of mantle zone lymphocytes, characteristic of Castleman's lymphadenopathy**

patients were typically asymptomatic.<sup>[9]</sup> Only 14% of the lesions were localized in the neck.<sup>[7]</sup>

Making a clinical diagnosis of Castleman's lymphadenopathy is difficult as a result of the disease's rarity and asymptomatic presentation and because it can often be mistaken for lymphoma.<sup>[2,3,6,9]</sup> Prognosis for the hyaline vascular type is favorable as surgical removal is usually curative. The plasma cell type may be inoperable and may instead benefit from radiotherapy, chemotherapy, or steroids.<sup>[3,9]</sup> In our case, radiation potentially could have been used; however, the mainstay of treatment for this lesion is surgical resection. Furthermore, biopsy of the lesion, although seemingly practical compared to undertaking a more invasive resection to determine the pathology of the lesion, would be dangerous given the location and anatomical distortion of the lesion. Patients with the hyaline vascular type usually have no recurrences after complete surgical removal; however, patients with the plasma cell type may be prone to recurrence.<sup>[3]</sup>

Hanzel *et al.*<sup>[7]</sup> made a diagnosis of carotid body chemodectoma in their patient and discovered the Castleman's disease only during the histopathologic examination, as is the situation with most of these cases. In our case, re-review of the angiogram revealed features that in hindsight were atypical for carotid body tumors and hinted at a different pathology. First, the vascular blush [Figure 2a], though present, was very diffuse and consisted of very small blood vessels instead of the enlarged channels typical for paragangliomas.<sup>[5]</sup> Second, no obvious enlargement of the external carotid artery branches supplying the tumor was present. Third, the vascular channels supplying the tumor appeared to be arising directly from the vasa vasorum of all the affected vessels including the common, internal, and external carotid arteries.

Finally, the entire carotid bulb was abnormally dilated, and its wall appeared highly irregular and suggested tumor invasion. Although carotid invasion is possible particularly in head and neck cancers, such invasion typically does not result in dilation of the carotid bifurcation, which is characteristic of carotid body paragangliomas.<sup>[5]</sup> The difficulty encountered in our experience was that we were compelled to sacrifice the affected carotid artery. In spite of adequate exposure, grafting or preservation of the vessel was unobtainable given the invasion of tumor into the carotid wall. Performance of the preoperative balloon test occlusion was critical for safe and complete resection of this tumor without need for a bypass procedure, which may have been difficult and required intracranial exposure given the cranial extent of this tumor to the skull base and carotid canal.

We describe a rare case of Castleman's disease masquerading as a carotid body tumor. Surgeons who are confronted with similar head and neck pathology should pay close attention to the involvement of the carotid vessel wall, as highlighted in our case. Hints of a different pathology were apparent; however, it was only after a pathological examination that we made the diagnosis of Castleman's disease.

## ACKNOWLEDGMENTS

We would like to thank Paul H. Dressel BFA for the preparation of the illustrations and Debra J. Zimmer for editorial assistance.

Dr. Siddiqui declares the following financial relationships (all outside the submitted work): research grants–National Institutes of Health (co-investigator: NINDS 1R01NS064592-01A1 and NIBIB 5 RO1 EB002873-07),

University at Buffalo (Research Development Award); financial interests–Hotspur, Intratech Medical, StimSox, Valor Medical, Blockade Medical, Lazarus Effect; consultant–Codman and Shurtleff, Inc., Concentric Medical, Covidien Vascular Therapies, GuidePoint Global Consulting, Penumbra, Stryker Neurovascular, Pulsar Vascular; speakers' bureaus–Codman and Shurtleff, Genentech; National Steering Committees for Penumbra 3D Separator Trial, Covidien SWIFT PRIME Trial; advisory board–Codman and Shurtleff, Covidien Vascular Therapies; honoraria–Abbott Vascular and Codman and Shurtleff, Inc. for training other neurointerventionists in carotid stenting and for training physicians in endovascular stenting for aneurysms.

## REFERENCES

1. Castleman B, Iverson L, Menendez VP. Localized mediastinal lymphnode hyperplasia resembling thymoma. *Cancer* 1956;9:822-30.
2. Chaloupka JC, Castillo M, Hudgins P. Castleman disease in the neck: Atypical appearance on CT. *AJR Am J Roentgenol* 1990;154:1051-2.
3. Chen CC, Jiang RS, Chou G, Wang CP. Castleman's disease of the neck. *J Chin Med Assoc* 2007;70:556-8.
4. Cronin DM, Warnke RA. Castleman disease: An update on classification and the spectrum of associated lesions. *Adv Anat Pathol* 2009;16:236-46.
5. 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.
6. Flórez J, García-Pardo G, Auguet T, Sirvent JJ, Bel M, Richart C. Pelvic mass in 46-year old man. *Postgrad Med J* 1997;73:371-3.
7. Hanzel P, Hanzel CV, Hanzel SI, Szepe P, Zelenak K, Hajtman A. Castleman's disease mimicking carotid body tumor. *Acta Med Martiniana* 2012;12:39-44.
8. Manzoor T, Ahmed B, Najam A, Ayub Z. Carotid body paraganglioma. *J Coll Physicians Surg Pak* 2009;19:523-5.
9. Olscamp G, Weisbrod G, Sanders D, Delarue N, Mustard R. Castleman disease: Unusual manifestations of an unusual disorder. *Radiology* 1980;135:43-8.