An ectopic paraganglioma supplied by the subclavian artery in the right supraclavicular fossa

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ABSTRACT

Paragangliomas are rare and highly vascularized neuroendocrine tumors originating from neural crest-derived paraganglionic tissue surrounding the autonomic nerve. In this case, we report a nonfunctional paraganglioma located in the right supraclavicular fossa. Both computed tomography and paraffin-embedded sections diagnosed paraganglioma. The paraganglioma was presumed to be derived from the paraganglia of the right cervical parasympathetic nerve and primarily supplied by the right subclavian artery. Because of the abundant blood supply, we performed arterial embolization before the operation. This ectopic paraganglioma is considered a rare report of a paraganglioma supplied by the subclavian artery. (J Vasc Surg Cases and Innovative Techniques 2019;5:91-4.)

Keywords: Ectopic paraganglioma; Subclavian artery; Embolization

Paragangliomas can grow anywhere in the body from the skull base to the bladder along paraganglia because they originate from neural crest cells, which are present throughout the entire body during development and growth. The incidence of paragangliomas in the head and neck is 0.03%, whereas carotid body tumors (CBTs), glomus jugulare tumors (GJTs), and vagal paraganglioma account for 98% of all paragangliomas.[1] Paragangliomas are often supplied by the carotid artery, internal carotid artery, and external carotid artery, yet tumors supplied by the subclavian artery are rarely reported. We obtained the consent of the patient for publication of this report.

CASE REPORT

A 46-year-old man presented with an 8-month history of a painless mass located at the supraclavicular fossa in the right side of the neck. A 10-cm L-type incision in the right side of the neck was present because the patient had undergone an unsuccessful operation with diagnosis of a right thyroid nodular goiter in a community hospital 4 months earlier. The operation had to be terminated because of aggressive intraoperative bleeding on dissection of the mass, which was mistaken for a thyroid goiter. The man did not complain of any headaches, palpitations, or hypertension.

After admission to this hospital, an enhanced computed tomography (CT) angiography scan was performed. Plain CT revealed a 46- × 65- × 49-mm well-defined mass, with the top portion reaching to the bifurcation of the carotid artery and the bottom portion located on the top of the pleura (Fig 1, *A*). The mass pushed the right lobe of the thyroid, sternocleidomastoid, and carotid artery anteriorly. Enhanced CT revealed plaque enhancement related to vascularization in the mass, which was not visible on the plain CT scan (Fig 1, *B-D*). Furthermore, three-dimensional CT angiography clearly demonstrated that the mass was extremely vascularized, and the blood supply mainly originated from the right subclavian artery (Fig 2). The imaging study suggested a diagnosis of paraganglioma. Thyroid peroxidase antibody and total thyroxine levels in the venous blood were normal with values of 100.91 IU/mL and 167.90 nmol/L, respectively. Epinephrine and norepinephrine in the venous blood were within the normal ranges. Vanillylmandelic acid was not examined. Taking these findings together, we presumed it was a nonfunctional paraganglioma.

Considering the high degree of vascularization, we performed vessel embolization before the operation. Under local anesthesia, the right femoral artery was punctured to perform arteriography. Two main vessels branching off the right subclavian artery were found to supply the tumor. One vessel supplied the bottom of the tumor, and the other relatively robust vessel encircled the tumor and fed most of the tumor (Supplementary Video, *A*). Two branches were embolized using a Guglielmi detachable coil (Cook Medical, Bloomington, Ind). After the procedure of embolization was finished, the blood flow to the tumor was greatly reduced to 70% of the original blood supply (Supplementary Video, *B*). Twenty-four hours after embolization, open surgery was performed under general anesthesia. The incision was the previous L for cosmetic reasons. The incision was deepened by cutting the superficial fascia and the platysma. The right sternocleidomastoid was retracted laterally. The tumor was exposed, and the surrounding adipose tissue

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This investigation was financed by the National Natural Science Fund (No. 81070257) and the Science and Technology Planning Project of Guangdong Province (No. 2014A020212481).

Author conflict of interest: none.

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2468-4287

https://doi.org/10.1016/j.jvscit.2018.09.009

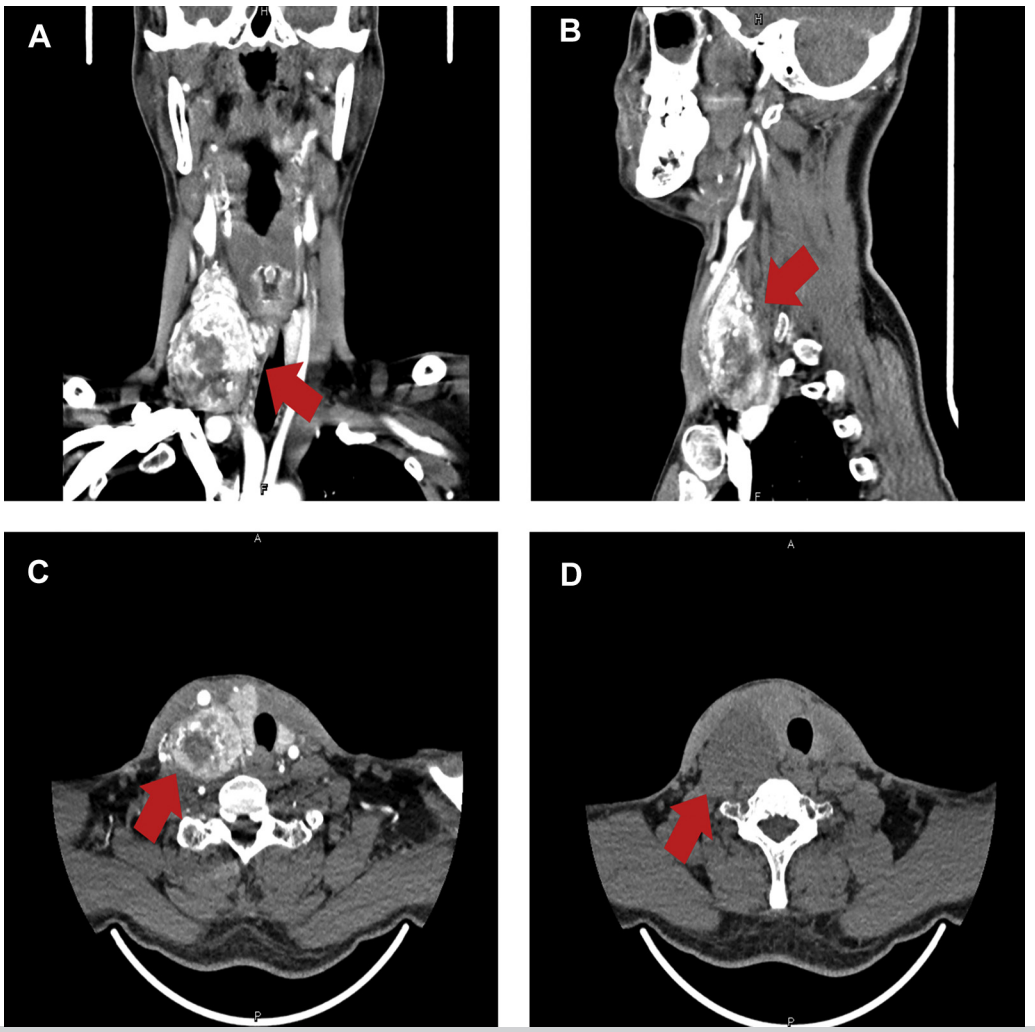

Fig 1. Plain and enhanced computed tomography (CT) of the neck: coronal **(A)**, sagittal **(B)**, and axial **(C, D)** views. The *arrows* indicate the neck mass.

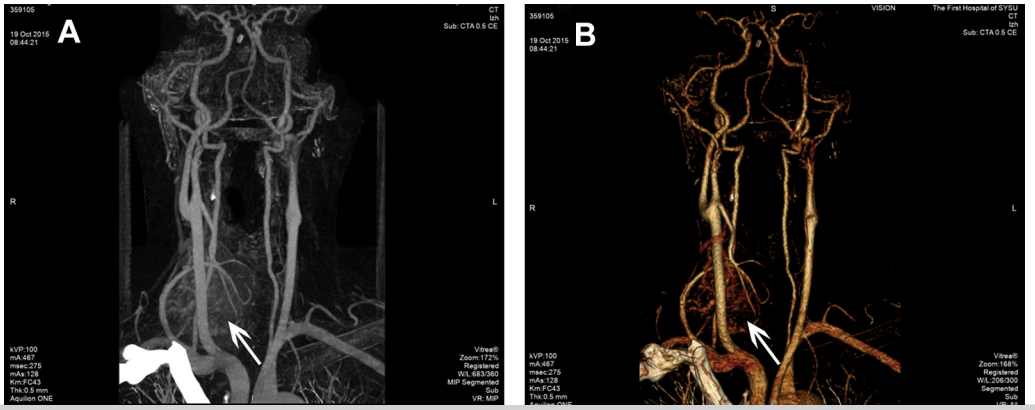

Fig 2. Three-dimensional computed tomography (CT) angiography of the tumor in the volume rendered **(A)** and maximum intensity projection **(B)** models. The *arrows* indicate the blood supply of the neck mass.

and tumor feeding vessels were dissected and ligated. The mass was not relevant to the vagus nerve. The lower border of the mass was dissected meticulously to protect the right subclavian vein. Control of intraoperative bleeding was easily obtained. The tumor was successfully removed. Intraoperative images before (Supplementary Fig 1, *A*) and after resection (Supplementary Fig 1, *B*) are provided. The patient recovered uneventfully and was discharged from the hospital.

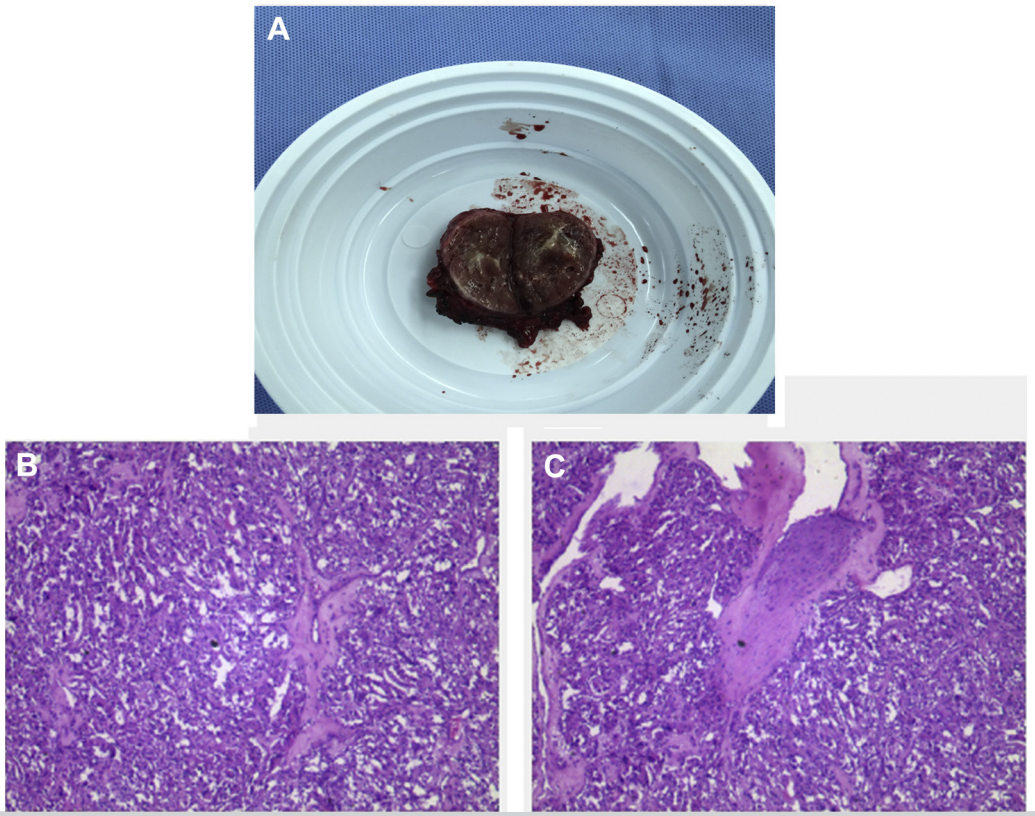

Fig 3. Histologic features of the paraganglioma. **A,** The size of the paraganglioma is approximately 46 × 65 × 49 mm. **B,** The tumor cells are organically arranged. Vitreous degeneration in the extracellular matrix is also evident. An enlarged nucleus of anachromasis, capillary networks, and ganglia next to tumor cells support the diagnosis of paraganglioma (hematoxylin and eosin, magnification ×40) **C,** Tumor cells stained by hematoxylin and eosin (magnification ×100).

Postoperatively, both frozen section examination and paraffin-embedded sections (Fig 3) demonstrated a well-encapsulated tumor arranged in a typical "Zellballen" pattern. No significant cytologic atypia was identified. Intercellular substance analysis revealed hyaline degeneration. The histopathologic examination supported the diagnosis of a paraganglioma. Pathologists ordered immunohistochemical staining for chromogranin A, synaptophysin, CD56, and neuron-specific enolase, but the patient declined.

At the last outpatient follow-up 2 years after operation, recurrence of the tumor in the neck was not detected by physical examination, follow-up enhanced CT (Supplementary Fig 2), or CT angiography (Fig 4).

DISCUSSION

Paragangliomas arise from the sympathetic and parasympathetic nervous systems. These tumors are mainly manifested as CBTs, GJTs, and vagal paragangliomas and represent rare neuroendocrine tumors.[2-4] Surgical removal is the most favorable treatment strategy for a neck mass thought to be a paraganglioma.[5] Paragangliomas in the head and neck can be unilateral or bilateral and typically grow slowly. Therefore, few patients complain of significant discomfort and merely present with a mass in the neck. As the tumor grows, however, it will invade the surrounding tissue and cause tumor mass effects, such as dizziness, palpitation, and dyspnea. A small proportion of paragangliomas secrete vasoactive substances, such as catecholamines, which can lead to typical hypertension and metabolic disorders and contribute to a diagnosis of functional paraganglioma.[6,7] Ultrasound, CT, magnetic resonance imaging (MRI), and digital subtraction angiography are helpful for diagnosis of paragangliomas; however, pathologic examination is the "gold standard." A typical "salt and pepper" appearance on MRI facilitates a diagnosis of vagal paraganglioma.

The tumor in this case was located beneath the proximal segment of the right carotid artery in the neck, at the supraclavicular fossa, and it was not relevant to the vagus nerve. According to the Glenner and Grimley classification, we presume that the paraganglioma originated from a paraganglion involved in the right cervical parasympathetic nervous system.[8] The rarity of this case is that the blood supply arteries to the tumor are unusual. To our knowledge, paragangliomas located in the right side of the neck are rarely reported to be fed by the vessels originating from the subclavian artery.

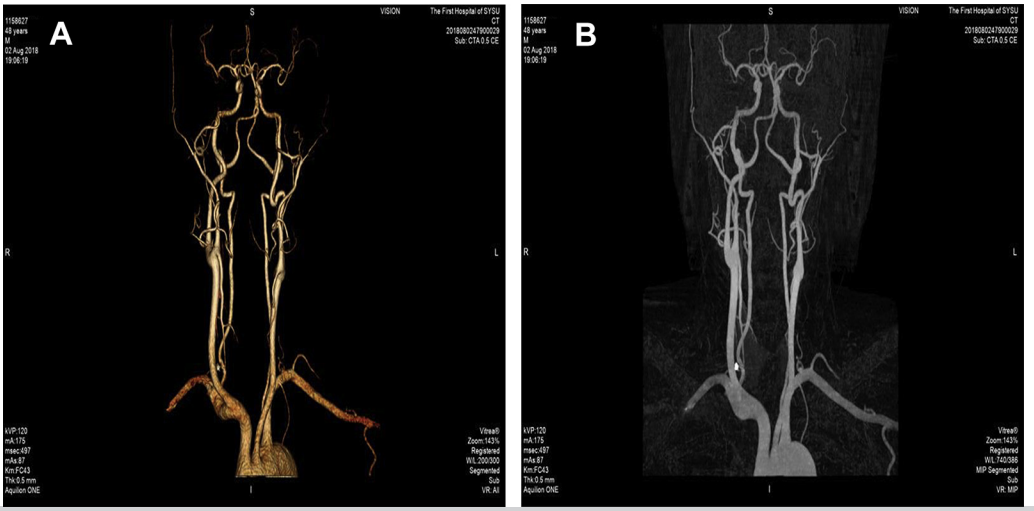

Fig 4. Postoperative three-dimensional computed tomography (CT) images of the neck. **A,** Volume rendered model. **B,** Maximum intensity projection model.

This paraganglioma was not located in a typically reported location in the neck. It grew and pushed the right thyroid lobe anteriorly. The first unsuccessful operation in another hospital was undertaken because of the misdiagnosis of thyroid tumor. The blood supply of the tumor was abundant, and the operator could not control the bleeding intraoperatively. However, current state-of-the-art helpful examinations, such as CT and MRI, can assist in the differential diagnosis by showing clear outlines of the tumor location, adjacent tissue, and blood supply. It is necessary to perform embolization before the operation to control bleeding for successful removal of the tumor by open surgery. Most of the neck paraganglioma was not functional. However, preoperative catecholamine levels should be detected to exclude a functional paraganglioma.

Resection of a paraganglioma does not suggest that treatment can be terminated. Paraganglioma metastasis and recurrence are possible because a pathologic diagnosis cannot determine whether the lesion is malignant or benign. Therefore, patients must undergo regular re-examination to exclude paraganglioma recurrence.

CONCLUSIONS

CBT, GJT, and paraganglioma are the common paragangliomas in the neck. They are usually located in the bifurcation of the carotid artery or above. A neck mass at the supraclavicular fossa is rarely diagnosed as a paraganglioma. To the best of our knowledge, this is the first description in the English literature of a paraganglioma located at the supraclavicular fossa supplied by the branches of the right subclavian artery. Although it is typically reported to exhibit a close association with the carotid artery, a paraganglioma with blood supplied from the subclavian artery should be part of the differential diagnosis.

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Submitted Feb 11, 2018; accepted Sep 28, 2018.