

A rare location of the glomus tumor in the abdominal aorta

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ABSTRACT

Glomus tumors are rare, mostly benign, and often superficial tumors that commonly occur on the upper and lower extremities. Whereas other locations have been described in the literature, we report the first case of glomus tumor in the abdominal aorta in a 21-year-old patient. (*J Vasc Surg Cases and Innovative Techniques* 2019;5:163-6.)

Keywords: Glomus tumor; Vessel tumor; Aortic disease

Glomus tumors are rare benign tumors initially labeled “painful subcutaneous tubercle” by Wood¹ in 1812. In 1924, Masson² histologically described glomus tumors as being composed of cells called myopericytes or apparently differentiated putative perivascular myoid cells. In 1934, Popoff³ defined the structure of the glomus as anastomotic connections between arterioles and venules resulting in shunts that contribute to thermoregulation.

The most common presentation of a glomus tumor is a painful subungual nodule in a finger. It can also be found in subcutaneous tissue of the extremities⁴ as well as in other locations. Whereas the fingers are the most common location for glomus tumors, approximately 35% are extradigital. To our knowledge, this article presents the first case of glomus tumor in the aorta. The patient’s informed consent was obtained for this publication.

CASE REPORT

This case describes the clinical course of a 21-year-old woman who presented to our cardiovascular surgery department with a 6-month history of abdominal and low back pain. An expansible and pulsatile abdominal mass was detected. Abdominal ultrasound revealed a well-defined hypoechoic mass between aorta and vena cava in close contact with the blood vessel. Abdominal and pelvic magnetic resonance angiography revealed a retroperitoneal tumor measuring 52 × 41 × 48 mm involving about three-quarters of the infrarenal abdominal aortic circumference. No lymphadenopathy was detected.

Computed tomography (CT)-guided biopsy found a richly vascularized tumor composed of sheets of round cells with a round

or oval nucleus and eosinophilic cytoplasm, consistent with a glomus tumor. This solid sheet of cells was organized around vessels of different sizes. There was no cellular atypism and no significant mitotic activity.

These tumor cells stained strongly for smooth muscle actin and caldesmon. The neuroendocrine cell markers chromogranin and synaptophysin were negative. Only CD34 was detected in the blood vessels. The Ki67 proliferation index was about 10% and heterogeneous. Based on these histologic and immunohistochemical findings, the diagnosis of glomus tumor was made.

After a 2-month observation period needed for confirmation of histology findings in the expert center and to make a surgical decision, the patient again presented with acute abdominal pain. Repeated CT revealed that the tumor was enlarging (Fig 1), with a diameter up to 61 × 45 × 51 mm. An in-tumor hematoma in the aortic wall was also detected.

The patient was urgently taken to the operating room for tumor resection and abdominal aortic replacement with a 14-mm tube graft. Operative time was 4 hours 36 minutes, and aortic clamping lasted 49 minutes. Pathologic examination confirmed a nonmalignant glomus tumor. The tumor invaded the aortic wall 40 mm distal to the renal arteries. It was resected proximally and distally in the intact zone, which stopped approximately 20 mm before the aortic bifurcation. The tumor had not invaded the vena cava; thus, no reconstruction was needed. A wide débridement of all tissues around the abdominal aorta and vena cava was performed. Intraoperative blood loss was 700 mL. The postoperative course was uneventful, and the patient was discharged after 9 days.

Gross examination revealed a white, nonspecific, elastic 68- × 70- × 65-mm mass that weighted 70 g and a hematoma in the center of the tumor (Fig 2). Histologic analysis of the biopsy specimen revealed a tumor developing in contact with the aortic wall media. Microscopic analysis confirmed a tumor consisting of a proliferation of round uniform cells with well-defined cell membranes formed around vascular spaces of varying sizes. Immunohistologic and immunochemical staining showed a strongly positive signal for smooth muscle actin (Fig 3). No evidence of malignant disease was found. Nevertheless, the tumor was considered as having an “uncertain malignant potential” because its dimensions were >20 mm and its location was deep according to World Health Organization 2012 classification.

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Fig 1. Preoperative computed tomography (CT) axial image of a tumor mass including active hemorrhage component.



Fig 2. Gross examination view of nonspecific elastic white mass invading aorta wall.

The follow-up period included a clinical examination and a Doppler ultrasound examination at 1 month, 6 months, and 12 months and then once a year. The Doppler ultrasound examination demonstrated a patent graft. Neither tumor recurrence nor complications were observed during a 5-year follow-up period.

DISCUSSION

Glomus tumor is known as a mesenchymal neoplasm composed of cells similar to modified smooth muscle cells of a normal glomus body.⁴ The glomus body is a normal histologic thermoregulating subcutaneous structure³ consisting of an arteriovenous shunt supplied by nerve fibers performing a temperature-regulating function. These systems are mostly present in the dermis of the digits, the palms, and the soles of the feet. Glomus tumors are usually benign and rare. Their incidence is estimated at <2% of all soft tissue tumors. Malignant forms of glomus tumors are exceedingly rare.⁴

In general, three histologic types of glomus tumors have been described: solid glomus tumors, glomangiomas, and glomangiomyomas. A solid glomus tumor consists of a nest of glomus cells surrounding the capillaries;

this makes up approximately 75% of all glomus tumors. Glomangiomas are characterized by cavernous hemangioma-like vascular structures surrounded by small clusters of glomus cells. A specific smooth muscle bundle is usually found in glomangiomyomas.⁴

The distinction between benign and malignant glomus tumors relies on the presence of nuclear atypical and mitotic activity, atypical mitotic figures, or a frank sarcomatous change. A total of 75% of glomus tumors occur in the hand, especially in the fingertips. They are typically located in the subungual area^{5,6} of the upper and lower extremities.⁷⁻⁹

As for vascular localizations, a rare venous glomus tumor was described¹⁰ clinically similar to varicose vein malformation; it was treated surgically. To our knowledge, ours is the first published report of an arterial or abdominal aortic location of a glomus tumor. Some of the unusual extradigital¹¹ locations are the oral mucosa, intraneural, gastric, mesocolon, kidney, and other locations¹²⁻¹⁶ not known to contain glomus cells under normal conditions. A glomus tumor can arise from perivascular cells that can differentiate into glomus cells.⁷ Clinical cases of malignant glomus tumors have also been described in the literature.^{16,17}

In our case report, the initial diagnosis of a glomus tumor in an aortic location was difficult to make because of the nonspecific clinical presentation. A differential diagnosis was required between aortic aneurysm, abdominal aortic tumor, and atheroma. Clinical symptoms, such as abdominal pain and abdominal pulsatile mass, can be suggestive of an abdominal aortic aneurysm. However, in this case, only the patient's age contributed to the possibility of a tumor origin of this mass. Magnetic resonance imaging with gadolinium and a CT scan provide a clear image of the tumor before surgery, making them effective methods to guide the decision-making process.¹⁸ However, the diagnosis of glomus tumor can be confirmed only after a pathologic examination. The CT-guided biopsy led the surgeon to a potentially tumoral character of the abdominal mass.

Our case of glomus tumor should be considered as having an uncertain malignant potential because of its large dimensions and its deep location. The urgent clinical presentation of the case (ie, pain and increasing diameter of the tumor mass), intratumoral heterogeneous CT scan images, high risk of an aortic rupture, and uncertain malignant potential were decisive elements for the surgical intervention. The hematoma detected on the CT scan could have been provoked by the CT-guided biopsy.

The high risk of an aortic rupture requires attention in diagnosis and treatment of emergent aortic tumor masses.^{18,19} We believe that surgical treatment of glomus tumors is the best curative approach, especially in an emergency situation. Complete resection of the glomus tumor is necessary to avoid recurrence. However, this

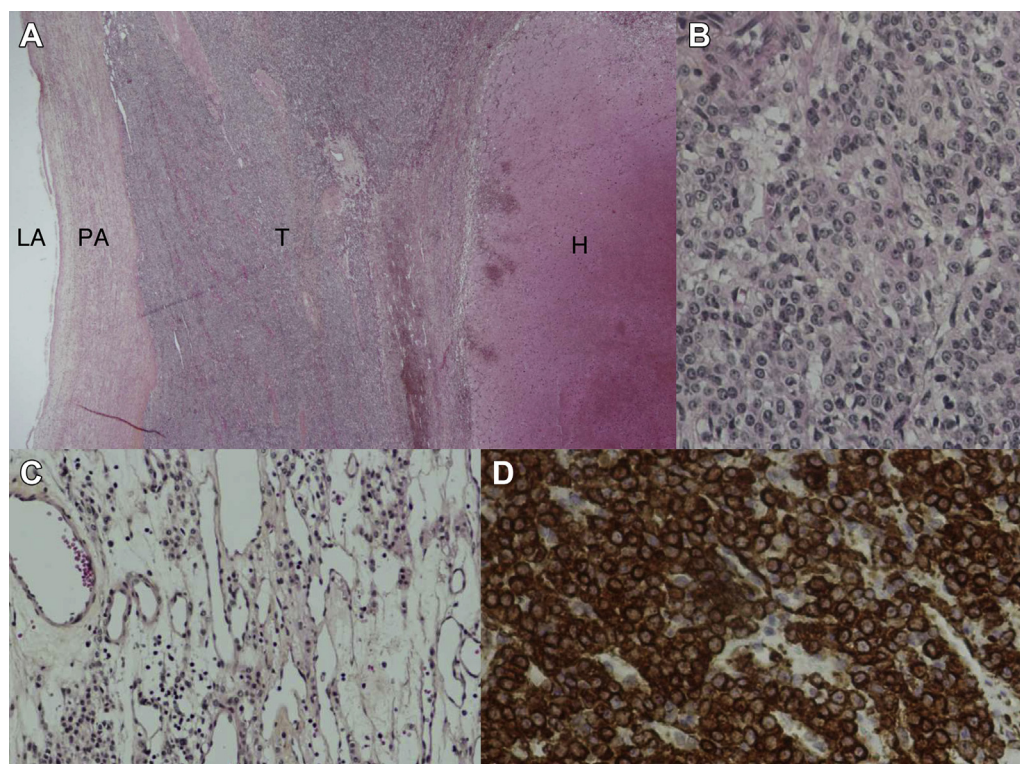


Fig 3. Section of hematoma located in the middle of the tumor. **A**, Aortic lumen (LA), aortic wall muscle (PA), tumor (T), and hemorrhage (H) on hematoxylin-phloxine-saffron (HPS) staining (magnification $\times 20$). **B**, A solid zone (HPS stain, magnification $\times 200$). **C**, A vessel-rich zone. Different-size vessels surrounded by little round cells (HPS stain, magnification $\times 100$). **D**, Actin muscle strong positivity after immunohistochemical marking (magnification $\times 200$).

approach is feasible in only 50% of all aortic tumor cases. Significant perioperative comorbidity has been reported; mean perioperative blood loss is approximately 2.4 L¹⁹ but in our case was limited to 700 mL.

CONCLUSIONS

Glomus tumors in aortic locations are rare and difficult to diagnose as they require rigorous preoperative analysis. Glomus tumors can be safely treated surgically to completely resect the tumor to avoid recurrence. A positive prognosis may be expected even in cases of uncertain malignant potential in young patients.

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