A case of metastatic adenocarcinoma from an unknown primary involving the aortic bifurcation

Lenny Suryani Safri, MD, MS,[a] Mohamad Syafeeq Faeez Md Noh, MD,[b] Ahmad Rafizi Hariz Ramli, MD, MS,[c] Suria Hayati Md Pauzi, MD, Dr Path,[d] Mohamad Azim Md Idris, MBChB, MS,[a] *and* Hanafiah Harunarashid, MBChB, FRCS,[a] *Cheras, Serdang, and Petaling Jaya, Malaysia*

ABSTRACT

Aortic malignant neoplasms are rare; these may be primary or secondary (metastatic). Increasing use of cross-sectional imaging has allowed better detection and diagnosis of these conditions. We encountered a young woman presenting with acute abdomen who was found on cross-sectional imaging to have a malignant tumor involving the aortic bifurcation. An en bloc excision of the tumor was performed, with distal aorta reconstruction using an aortoiliac Dacron graft; this was complicated with infection and graft occlusion, necessitating total removal and extra-anatomic bypass. A pathologic diagnosis of metastatic adenocarcinoma involving the aortic bifurcation, with an unknown primary, was made. (J Vasc Surg Cases and Innovative Techniques 2018;4:160-2.)

The aorta is a rare site for tumor deposition. However, this does not preclude the occurrence of primary and secondary aortic malignant neoplasms. Brodowski,[1] in 1873, reported the first case of an aortic malignant neoplasm, a fibrosarcoma of the thoracic aorta. Since then, there have been isolated case reports and case series of aortic malignant neoplasms in the literature. Metastatic tumors involving the aorta are relatively more common than primary tumors. In the thoracic region, these usually originate from the lungs, esophagus, and thymus; in the abdomen, germ cell tumors and tumors of retroperitoneal origin are commonly implicated.[2] Adenocarcinomas are tumors originating from glandular tissue, usually affecting the lungs, gastrointestinal tract, breasts, and thyroid gland. Metastasis to the aorta is very rare. We report a young woman encountered in our center, presenting with acute abdomen, eventually diagnosed with metastatic adenocarcinoma involving the aortic bifurcation, with an unknown primary.

Informed consent has been obtained before publication of this article.

CASE REPORT

A 27-year-old woman with no underlying medical illness presented to our center with a 2-week history of progressively worsening abdominal pain associated with constipation and constitutional symptoms. There was no history of trauma or surgery. She was afebrile and normotensive, and her heart rate was 90 beats/min. Palpation of the abdomen revealed no mass, with maximal tenderness elicited at the lower abdomen. An abdominal ultrasound examination showed presence of vague para-aortic hypoechoic masses, measuring 3.0 × 2.7 cm in the largest dimension. Contrast-enhanced computed tomography (CT) of the abdomen and pelvis showed a heterogeneously enhancing para-aortic mass, measuring approximately 3.3 × 4.5 × 6.5 cm with terminal aortic wall and aortic bifurcation encasement (Fig). Left ureteric compression with resultant proximal hydroureter and hydronephrosis was present. The rest of the abdominal organs were grossly normal. Biochemical parameters showed a raised white blood cell count at 12 × 10^9/L (normal, 4-11 × 10^9/L). Serum carbohydrate antigen 19-9 level was also found to be raised, with a value of 92 U/mL (normal, <37 U/mL). Ultrasound-guided biopsy showed a moderately differentiated adenocarcinoma, with possible pancreatic origin. Immunohistochemical staining was positive for cytokeratin 7, cytokeratin 20, carbohydrate antigen 19-9, and carcinoembryonic antigen. In view of the possible pancreatic origin, a pancreatic protocol CT as well as a positron emission tomography scan was arranged, both with normal findings in relation to the pancreas. Abdominal CT did not demonstrate other hepatobiliary lesions. Findings on gynecologic examination and upper endoscopy and colonoscopy were normal. On exploration, a retroperitoneal mass, encasing the aortic bifurcation with infiltration into the left common iliac vein and left ureter, was demonstrated intraoperatively. Inspection of the rest of the organs was unremarkable. An en bloc excision of the tumor was performed, with distal aorta reconstruction using an aortoiliac Dacron graft. The affected left common iliac vein was resected and ligated. Excision of the involved left ureter was achieved, with primary anastomosis over a double J stent. Pathologic examination of

From the Department of Surgery[a] and Department of Pathology.[d] Faculty of Medicine, Universiti Kebangsaan Malaysia Medical Center, Cheras; the Department of Imaging, Faculty of Medicine and Health Sciences, Universiti Putra Malaysia, Serdang[b]; and the Department of Surgery, Faculty of Medicine, Universiti Malaya Medical Center, Petaling Jaya.[c]

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Correspondence: Lenny Suryani Safri, MD, MS, Clinical Lecturer and Vascular Surgeon, Department of Surgery, Faculty of Medicine, Universiti Kebangsaan Malaysia Medical Center, Jalan Ya'acob Latif, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia (e-mail: lennysuryanisafri@yahoo.com).

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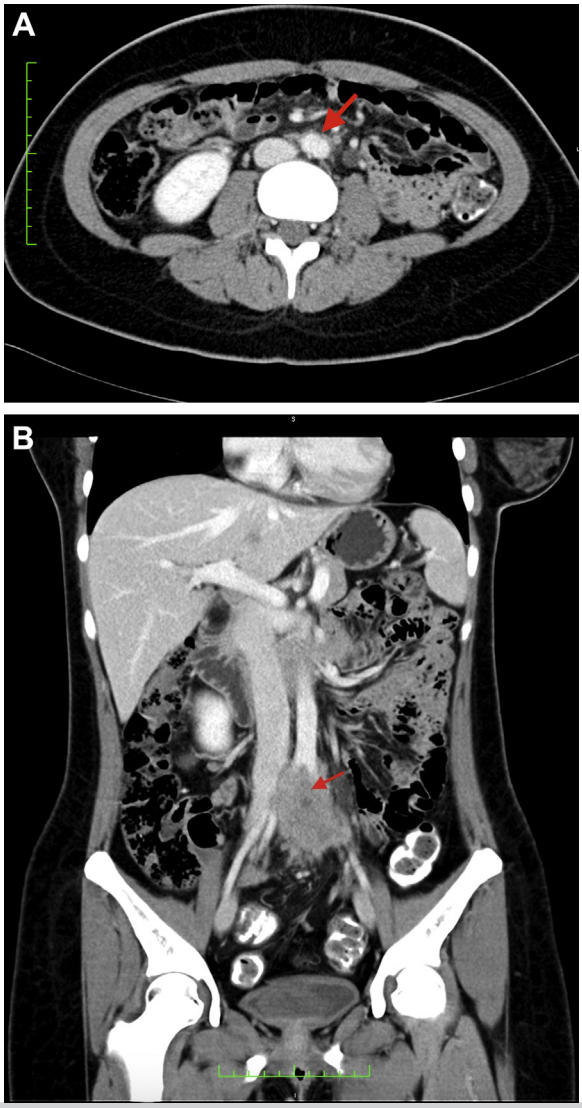

Fig. Contrast-enhanced computed tomography (CT) images, in axial **(A)** and coronal **(B)** section, showing the lesion abutting the terminal abdominal aorta and encasing the aortic bifurcation (*arrow*).

the resected specimen confirmed encasement but no invasion into the aortic bifurcation and ureter; both ureteric and iliac vessels were free of malignant disease. There was abutment (and no invasion) of the terminal abdominal aorta. The tumor cells were <1 mm from the anterior and posterior margins, 2 mm from the right and left lateral margins, 3 mm from the superior margin, and 5 mm from the inferior margin. There was early graft infection, with graft occlusion and perigraft abscess formation on day 13 after surgery, which necessitated total removal of the infected graft and drainage of the abscess. Cultures grew extended-spectrum β-lactamase *Escherichia coli*; however, the source was unknown. Appropriate antibiotics were instituted. An axillobifemoral anastomosis was performed. She was then admitted to the intensive care unit for close monitoring and management and was discharged home 7 days later. She was referred to oncology for adjuvant chemoradiation, with 6 cycles of cisplatin and paclitaxel as well as radiotherapy to the affected region. CT during a period of 2 years at 4- to 6-month intervals showed no evidence of recurrent malignant disease. The patient then died of a road traffic accident.

DISCUSSION

Aortic malignant neoplasms are rare clinical entities, with a multitude of clinical manifestations and imaging findings but no widely accepted consensus for operative intervention. Isolated case reports and case series have attempted to describe their various clinical presentations, imaging findings, and treatment strategies. To this day, evidence-based information for the most appropriate algorithm in management of these cases is still lacking. A case-by-case approach is most appropriate, at present.

Malignant neoplasms affecting the aorta may be either primary or secondary (metastatic). Of these two, secondary malignant neoplasms are more common.[2] Primary aortic malignant neoplasms more commonly affect men, with a male to female ratio of 2:1 to 5:1. The mean age is 60 years, and both thoracic and abdominal aortas are affected equally.[3,4] Because of the old age at presentation, imaging findings, when present, are commonly attributed to thrombosis and atherosclerotic disease. This contributes to the difficulty in early diagnosis.

Secondary (metastatic) aortic malignant neoplasms are more commonly seen and are more readily diagnosed, possibly owing to the knowledge of a pre-existing primary tumor. There appears to be no age predilection. In the thoracic region, invasion from the lungs, esophagus, and thymoma is the usual scenario.[2] Retroperitoneal sarcomas and germ cell tumors are the most common malignant neoplasms invading the abdominal aorta, potentially with resultant aneurysm or pseudoaneurysm formation and rupture.[5-7]

A wide spectrum of clinical manifestations may accompany aortic malignant neoplasms, regardless of whether the tumor is primary or metastatic. These include embolic events to the brain, lower extremities, and abdominal viscera manifesting as hypertension, intestinal infarcts, peripheral vascular disease, or acute abdomen.[2] Uncommonly, spontaneous aortic rupture with massive hemorrhage may occur; this has been reported in the literature.[8,9]

The scarcity in the literature of metastatic carcinoma involving the aorta (Table) hampered us in terms of locating the primary tumor. The biopsy result and immunohistochemical staining suggesting a pancreatic origin led us to pursue structural and functional imaging to identify the primary tumor. The findings were normal. We postulated, after taking into account the possible pancreatic origin, that the tumor excised from the aortic bifurcation may have originated from either the aorta itself or, more plausibly, the gastrointestinal tract in the

Table. Cases of malignant disease with secondary aortic involvement or metastasis in the English literature in the past 15 years

Year	Primary tumor	Site of aortic involvement or metastasis	Treatment
Jerius et al, 2003	Testicular germ cell tumor	Infrarenal abdominal aorta	Chemotherapy + floppy aortic graft reconstruction
Zerati et al, 2007	Embryonic carcinoma	Infrarenal abdominal aorta	Chemotherapy + aortobi-iliac grafting
Busuito et al, 2012	Endometrial stromal sarcoma	Infrarenal abdominal aorta	En bloc tumor resection and in situ aortic replacement + hormonal therapy
Tanaka et al, 2017	Retroperitoneal dedifferentiated liposarcoma	Abdominal aorta	En bloc resection, with extra-anatomic femoral-femoral crossover after left axillofemoral bypass

form of ectopic pancreatic tissue. When present, ectopia of the pancreatic tissue is mostly located within the gastrointestinal tract (70%-90% cases).[10]

To this day, there is no consensus on the choice of oncologic intervention by chemoradiation. In this case, our patient received 28 fractions of radiotherapy, with combination chemotherapy with paclitaxel and cisplatin. After chemoradiation, CT during a period of 2 years at 4- to 6-month intervals showed stable disease, with no evidence of recurrence. Bohner et al[11] and Majeski et al[12] demonstrated in their experience that when limited metastasis or unresectable local recurrence is present, control with chemoradiation for a long period is possible.

CONCLUSIONS

Aortic malignant neoplasms are rare clinical entities with various clinical manifestations. Accurate diagnosis, followed by appropriate management, requires a multidisciplinary approach. Operative intervention, at present, is undertaken on a case-by-case basis. Our experience in managing this rare case of metastatic adenocarcinoma involving the aortic bifurcation is testament to the challenges involved in managing these rare surgical conditions.

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