Case report - Vascular general

A primary angiosarcoma in the aorta

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Abstract

A 60-year-old woman after aortic valve replacement had an acute abdominal pain. Computed tomography demonstrated a tumor in the aorta, which originated from the distal portion of the thoracic aorta and extended to the aortic bifurcation. At autopsy, the orifices of celiac, superior and inferior mesenteric, right and left renal arteries were consistently occluded by incursion of the tumor. We present herein a huge primary sarcoma in the abdominal aorta, which has been reported as extremely rare.

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Keywords: Tumor; Abdominal aorta; Intimal sarcoma

1. Clinical presentation

A 60-year-old woman with severe aortic stenosis and regurgitation underwent valve replacement. On the fourth postoperative day, she had an acute abdominal pain, and computed tomography demonstrated an extremely long tumor in the aorta, which was arising from the distal portion of the thoracic aorta and extended to the aortic bifurcation. Transesophageal echocardiography revealed that the tumor had considerable mobility in the blood flow. This would be the reason why the pulsation of the bilateral femoral arteries was palpable before the operation. Visceral ischemia was rapidly and extensively progressive. She died of multiple organ failure.

At autopsy, the tumor was attached to the thoracic aorta, and developed only in the aortic lumen. The tumor was arising from the intima of the aorta. The media and adventitia were intact. The orifices of celiac, superior and inferior mesenteric, right and left renal arteries were entirely occluded by incursion of the tumor (Fig. 1).

The tumor was examined immunohistologically. Staining for vimentin revealed abundant microvasculature. Staining for desmin, neurofilament, CD-34, alpha smooth muscle actin, epithelial membranous antigen and cytokeratin was consistently negative. These findings suggested that characteristics of this tumor coincided with that of intimal sarcoma of the endothelial cell type (Fig. 2).

2. Comments

The primary sarcoma in the aorta is extremely rare. Intimal sarcoma of the aorta was more common in male than in female, and the most frequent site was infrarenal portion [1]. Aortic sarcoma can be divided into two growth patterns, which are luminal and mural patterns. The tumor of luminal growth pattern is more popular than that of mural pattern, and often embolizes the distal peripheral artery or occludes its origin [1]. For precise diagnosis of intimal sarcoma, magnetic resonance imaging was reported practical [1, 2]. Usefulness of ultrasonography was also reported, because it revealed the inhomogeneous mass unlike a thrombus and suggestive of a tumor [3]. On the contrary, in contrast-enhanced computed tomography, the tumor could not be easily distinguished from the thrombus in the aorta. Since the major location of the metastasis was bone, scintigraphy was recommended when the malignant aortic tumor was suspected [1]. For patients with no metastatic lesion, artificial graft replacement or endarterectomy with confirmation of negative margin of resection may be the effective surgical procedures of choice [1, 4].

Fig. 1. Longitudinally incised descending thoracic and abdominal aorta. IMA, inferior mesenteric artery; LRA, left renal artery; SMA, superior mesenteric artery.
Primary aortic intimal sarcoma is divided as two types, mesenchymal cell type and endothelial cell type from the immunohistochemical staining using mesenchymal markers and endothelial markers. Regardless of cell type, the prognosis of patients presenting bowel ischemia or with metastasis in bone or other organs, such as spleen, liver, or skin, is extremely poor \([1–3, 5]\), because chemotherapy and radiation are not effective. The survival time in the majority of reported cases was less than two years.

In the present case, the tumor was endothelial cell type, of luminal growth pattern, and did not invade the aortic wall. There was no obvious metastasis in the peritoneal cavity at autopsy. Unfortunately, we could not identify this huge tumor in the aorta before occlusion of the visceral arteries. Although computed tomography without contrast enhancement was routinely performed as screening before valvular heart surgery, the tumor in the aorta was not obviously demonstrated. We presumed that the increase of cardiac output, disappearance of regurgitant flow in the aorta after prosthetic valve implantation and concomitant anomalous coagulability caused such a sudden abdominal ischemia.

In conclusion, aortic intimal sarcoma is a rare clinical entity. Prompt diagnosis before organ ischemia and systemic metastasis is difficult because of its various clinical manifestation and rarity, but is essential for survival.

References