INTRODUCTION

Primary malignancy of the aorta is a very rare disease and only approximately 150 cases have been reported in the literature, including approximately 40 cases of aortic angiosarcoma (AAS) [1]. Because of its rarity, it is very difficult for a clinician to differentiate AAS from other non-malignant diseases of the aorta, such as abdominal aortic aneurysm (AAA), atheroma, and mural thrombus [2,3]. Because the prognosis of primary AAS is extremely poor, clinical suspicion is key to early diagnosis and proper surgical treatment, especially in cases with atypical rapid growth of an aortic abdominal aneurysm with a thrombotic mass. Herein, we report a case of angiosarcoma in the abdominal aorta mimicking an infected aneurysm and present computed tomography and positron emission tomography findings.

Key Words: Aorta, Hemangiosarcoma, Angiosarcoma, Aneurysm, Positron-emission tomography

INTRODUCTION

Primary malignancy of the aorta is a very rare disease and only approximately 150 cases have been reported in the literature, including approximately 40 cases of aortic angiosarcoma (AAS) [1]. Because of its rarity, it is very difficult for a clinician to differentiate AAS from other non-malignant diseases of the aorta, such as abdominal aortic aneurysm (AAA), atheroma, and mural thrombus [2,3]. Because the prognosis of primary AAS is extremely poor, clinical suspicion is key to early diagnosis and proper surgical treatment. Herein, we report a case of AAS mimicking an infected aneurysm treated by complete excision. The diagnosis was made unexpectedly after a permanent pathologic examination.

CASE

A 62-year-old man was referred to the vascular clinic with a rapidly growing AAA. The diameter of the saccular outpouching aneurysm in the infrarenal aorta increased from 18×19 mm to 21×36 mm in 2 months. He presented with symptoms of abdominal pain without fever. Laboratory tests revealed no abnormalities and the leukocyte count (6,630 /µL) and C-reactive protein level (0.34 mg/dL) were within the normal range. With suspicion of an infected aneurysm, positron emission tomography (PET) was performed and showed hypermetabolic lesions in the aorta, bilateral common iliac artery, and right inguinal nodes, suggesting an infected aneurysm and reactive inflammation (Fig. 1). Under clinical suspicion of an infrarenal infected aneurysm, emergency operation was performed via a midline transperitoneal approach, including excision of the whole aortic aneurysm and surrounding tissue, in situ reconstruction with a rifampicin-soaked 16-8 mm Dacron bifurcated graft (Hemashield; Maquet, Rastatt, Germany), and omental wrapping. There was no gross pus, but peri-aneurysmal inflammation was present and compatible with
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an infected aneurysm. The microbiologic culture showed no bacterial growth in either the blood or aortic tissue. The patient recovered well without any complications. However, the histologic examination revealed intravascular epithelioid angiosarcoma. Although frozen sections were omitted during the operation, the resection margin was free and lymph node metastasis was negative in nine nodes. The tumor was confined to the aortic wall and thrombus, the tumor cells were mixed with a fibrin clot, and most of the lesion consisted of necrotic thrombus (Fig. 2). R0 resection was performed fortunately and no additional therapy was given. The patient was doing well with no recurrence until the follow-up computed tomography (CT) angiography after 2 years, which unfortunately revealed metastatic lesions inside the aortic graft down to the common iliac arteries and a left adrenal mass encasing renal vessels (Fig. 3). He has remained alive for 3 years postoperatively on palliative chemoradiotherapy.

DISCUSSION

Angiosarcoma is a rare malignant neoplasm that develops in the inner lining of the blood and lymph vessels. They can develop anywhere in the body, but the most commonly

Fig. 1. Initial outside computed tomography revealed a 18×19 mm saccular outpouching lesion of aorta just above the aortic bifurcation (arrow in A). The lesion increased up to 21×36 mm in size within 50 days (arrow in B). Positron emission tomography revealed hypermetabolic lesions in the aorta (arrows in C and D).

Fig. 2. Pathologic specimen of the resected aorta. Most of the lesion consisted of necrotic thrombus in the gross specimen (A). Microscopic view showed tumor cells with necrotic cells (B, C) (Hematoxyline & eosin stain, magnification, ×100 in B, ×200 in C).
involved sites are the skin, breast, liver, and spleen [4]. The possible causes include past radiation therapy, persistent lymphedema, or carcinogenic chemicals. The tumor is characterized by rapidly proliferating, extensively infiltrating anaplastic cells derived from blood vessels and lining irregular blood-filled spaces.

Primary malignancy of the aorta is extremely rare, and Browdowski [5] reported the first case in 1873. Aortic sarcoma can be divided by the location and origin of the tumor—intima from the vascular endothelial cells (angiosarcoma), intima from the myofibroblasts or neointimal cells (intimal myofibroblastic sarcoma), media from the vascular smooth muscle cells (leiomyosarcoma), and adventitia from the fibroblasts (fibrosarcoma) [6]. Angiosarcoma can also be referred to as hemangiosarcoma or endotheliosarcoma. Epithelioid hemangioendothelioma is a benign form of the disease. In 1972, Salm [7] first established a morphological classification dividing aortic sarcoma into intraluminal, intimal, and adventitial types. Subsequently, Wright et al. [8] subdivided the intimal type into obstructive and non-obstructive types. In 2003, Chiche et al. [9] reported that the prognosis was worse for the intraluminal and intimal types due to the proximity of the lesion to the aortic lumen. Intimal angiosarcomas and intimal myofibroblastic sarcomas can be differentiated using immunohistochemical staining. Angiosarcomas express mostly endothelial proteins, such as CD31, CD34, factor VIII, and/or Fli-1 [10].

AAS can develop in all segments of the aorta, as well as in the carotid, renal, iliac, and femoral arteries. Sebenik et al. [11] reviewed 109 intimal sarcomas of large systemic arteries and reported that the most common site was the abdominal aorta (38.5%), followed by the thoracic aorta (22.9%), aortic arch (11.9%), thoracic/abdominal aorta (11.0%), and abdominal aorta/iliac/femoral artery (5.5%). The etiology of AAS is unknown, but lymphedema, radiation, trauma, inflammatory aneurysm, and Dacron graft have all been suggested to play a role [12,13].

The most common clinical symptoms are abdominal pain and vaso-occlusive symptoms, often accompanied by generalized fatigue, weight loss, and fever, which are nonspecific, making it difficult to distinguish AAS from other disease [3]. In addition, AAS is often confused with thromboembolism, aortitis, and AAA, because there are no clear imaging diagnostic criteria. Most AAS present with signs of thromboembolic disease, and sometimes with symptoms of aortic stenosis, rupture, infectious or inflammatory aneurysm, or aortitis. A clue to diagnosis is an aneurysm in a young healthy patient without combined trauma or vasculitis and suspicion of AAS should be assessed during surgery, including obtaining frozen sections. Preoperative diagnosis is very difficult because of the diverse presentation and nonspecific symptoms and imaging studies often fail to differentiate between atheroma, mural thrombus, aneurysms, and tumors [14]. The diagnosis is often made unexpectedly following pathologic examination. The diagnosis depends on a high level of suspicion from clinicians. If a patient presents with symptoms of arterial thromboembolism with a normal echocardiogram and a heterogeneous protruding aortic plaque in the absence of generalized atherosclerotic disease, AAS should be suspected.

Kamran et al. [15] suggested the following image findings as predictors of primary aortic malignancy: protrusive vegetation, nodular soft tissue component, and lack of atherosclerosis on CT; area of hypermetabolic uptake on PET;
and neovascularity and a mass-like signal distinct from the non-enhancing mural thrombus on magnetic resonance imaging (MRI). Some clues on CT are an enlarging thrombus with hyperenhancement in the arterial phase due to its high vascularity and extension of the mass into the vessel wall and/or distal embolization. MRI with gadolinium may show unusual contrast enhancement differentiating between tumors and atheromatous lesions [14]. Recently, Hossien et al. [16] reported that a new technique of finite-element multidimensional modeling with CT and MRI is useful for preoperative planning in the treatment of AAS. PET may show increased uptake in the tumor, but fluorodeoxyglucose avidity does not distinguish inflammation or infection from neoplasm.

Intraoperatively, AAS appears as a fungating intraluminal mass or a cauliflower-like gelatinous mass emanating from the intima, distinct from atherosclerotic disease. The diagnosis of AAS should be suspected and frozen sections are to be sent for analysis. The role of surgery in this highly fatal disease is uncertain, but many palliative and curative operations have been attempted, including en bloc resection, endarterectomy, simple thrombectomy, and bypass graft. Patients with embolic disease had poorer survival. Although the evidence is weak, surgical treatment for primary aortic malignancy patients has been reported to have a survival gain, with a postoperative survival rate of 16.5% and 11.8% after 3 and 5 years, respectively [9]. Surgical resection with a wide margin in invaded aorta and subsequent prosthetic graft replacement has been considered as a recommendable treatment modality based on case reports presented to date [17].

Because of the highly aggressive nature of the tumor, most patients die of metastatic disease, leading to profound cachexia, sepsis, and multisystem organ failure. The most common metastatic site is bone, followed by the lung, liver, kidney, duodenum, and peritoneum. The presence of distant metastasis at the time of diagnosis is very common (69%-83%). The mean survival was reported to be 16.7±2.4 months and the 3- and 5-year survival rates were 11.2% and 8%, respectively. The AAS reported in our case corresponds to the intimal type aortic malignancy, which has a poor prognosis and 3- and 5-year survival rates of 5.1% and 2.6%, respectively [9,18]. The role of chemotherapy or radiotherapy is not yet defined [14]. However, Fatima et al. [19] reported that complete resection of the tumor in combination with adjuvant therapy seemed to offer the best chance at survival and advocated aggressive multimodality therapy, given the young age of patients presenting with this disease.

The specific clinical scenario of AAS should be emphasized. Because of the recent trend of treating AAA using endovascular aortic aneurysm repair (EVAR), the opportunity for pathologic diagnosis after operative repair is no longer available. Some cases of AAS were diagnosed after open conversion of a complicated EVAR [13,15,18]. Endograft might cause AAS but the mechanism of action is not yet proven. Possible causes of AAS after EVAR include: 1) Dacron graft, 2) nickel and titanium alloy, and 3) coincidental and preexisting tumor at the time of EVAR. Dacron can induce or promote carcinogenesis secondary to a foreign body effect on the endothelium. Nickel/titanium alloys in the stent can also promote carcinogenesis [20]. However, in most cases, misdiagnosis of the preexisting tumor as a growing AAA lead to EVAR, and unsolved expansion of the aorta and enlarging thrombotic mass resulted in open conversion and delayed diagnosis of AAS. Some cases mimic an infected aneurysm after EVAR. Therefore, careful preoperative image analysis and postoperative CT surveillance is especially important.

**CONFLICTS OF INTEREST**

The authors have nothing to disclose.

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Concept and design: SKM. Analysis and interpretation: WKP, KLP, YSC. Data collection: YSC. Writing the article: WKP, KLP, YSC, SKM. Critical revision of the article: AH, SA, SKM. Final approval of the article: AH, SA, SKM. Overall responsibility: SKM.
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