Surgical Therapy for Angiosarcoma of the Aorta: A Case Report

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We describe the case of a 71-year-old woman presenting with abdominal angina with an intra-aortic mass at the thoracoabdominal aorta that restricted blood supply to the organs. Initially, the intra-aortic mass was suspected to be a mural thrombus; thus, endarterectomy was performed. However, postoperative histopathological examination revealed an intimal sarcoma, which relapsed locally within a few months. Additional en bloc resection of the aorta with graft interposition was performed. Despite surgical therapy, splenic metastasis was detected a few months after the second surgery; therefore, palliative care was selected for the patient.

Keywords: angiosarcoma, thoracoabdominal aorta

Introduction

Primary angiosarcoma is an extremely rare tumor with a poor prognosis, and its preoperative diagnosis is difficult.1,2) Surgical therapy is an option, involving either en bloc resection (EBR) with graft interposition or endarterectomy of the diseased part.3) However, owing to fewer reported cases, the optimal strategy for the treatment of primary angiosarcoma remains controversial. Here we present a rare case of primary angiosarcoma treated with surgical therapy, involving endarterectomy followed by EBR.

Case

A 71-year-old woman with a long-term non-steroid anti inflammatory drugs prescription was transferred to our hospital due to abdominal angina and an atypical intra-aortic mass, detected using computed tomography (CT). Echography revealed a mobile 60×20 mm mass at the thoracoabdominal aorta (Fig. 1), occluding the celiac artery. The superior mesenteric artery was largely occluded, with a peak blood-flow velocity of 3.0 m/s according to Doppler echography. CT scan image revealed an irregular mass inside the thoracoabdominal aorta (Fig. 2a), which completely occluded the celiac trunk with subsequent splenic infarction (Figs. 2b and 2c). Laboratory data showed slightly elevated levels of C-reactive protein (1.94 mg/dL) and normal white blood cell (6,430/µL), lactate dehydroase (167 U/L), creatinin phosphokinase (31 U/L), and creatinine (0.7 mg/dL). Physical examination revealed that the bilateral dorsal artery was palpable with normal ankle–brachial index (Right 0.94, Left 0.98). Initially, the mass was suspected to be a mural thrombus. Because the mass was mobile and had already caused an embolic event, emergent surgery was scheduled to prevent further thromboembolism.

Fig. 1 Echography showing a 60×20 mm intra-aortic mass (white star). The mass presented as an iso-echoic lesion, with a slightly higher-echoic lesion inside of it.
The thoracoabdominal aorta was exposed using left thoracophrenic laparotomy. The descending and supra-renal aorta were cross-clamped under partial cardiopulmonary bypass, established from the femoral artery and vein. Selective visceral perfusion to the superior mesenteric artery was established following aortotomy. The abnormal mass occupying the thoracoabdominal aorta was removed using endarterectomy. The mass was fragile, yellowish, and lipid-rich (Fig. 3a), which was unlike the usual thrombi. The possibility of an aortic tumor was considered; therefore, thorough endarterectomy was performed. The patient was discharged uneventfully on postoperative day 13. However, histopathological examination revealed an intimal sarcoma. Because endarterectomy, one of the surgical treatments for angiosarcoma, was performed thoroughly, a close follow-up was scheduled instead of additional surgical therapy. However, a follow-up CT scan image (2 months post-surgery) revealed local relapse of the tumor, which was growing and might have obstructed the aorta again. Therefore, EBR was performed. Despite surgical therapy, huge splenic metastasis was detected a few months after the second surgery, and was not detected at the time of the second surgery (Fig. 3b). Hence, palliative care was selected, and the patient died 16 months after the first surgery.

Discussion

Primary angiosarcoma is an extremely rare neoplasm, and there are only approximately 140 previously reported cases. Aortic sarcoma comprises of two subtypes, namely intimal-type and mural-type. Intimal sarcoma originates from the intima of the aortic wall, and frequently presents with thromboembolic events, whereas mural sarcoma originates from the media or adventitia, and may cause local invasion. The prognosis of angiosarcomas is generally extremely poor, with a mean survival rate of approximately 14.9 months.

The diagnosis of angiosarcoma is difficult owing to its non-specific clinical presentation and extreme rarity. The most common clinical presentation is related to the tumor embolization of the extremities or other organs, with pain, claudication, abdominal complaints, and hypertension being the frequently reported symptoms. Imaging findings are generally non-specific, and differentiating angiosarcoma from atheroma or mural thrombus is particularly challenging. In the present case, the tumor showed iso-density on CT scan, and presented as an almost iso-echoic lesion, with a slightly higher-echoic lesion on the inner side, on echography; therefore, it was difficult to rule out either thrombus or atheroma. Rapid pathological diagnosis at the time of surgery may play an important role; however, as in our institution, a department of pathology may not exist in every institution. Hence, postoperative histopathological examination remains the gold standard for diagnosis.

Despite its poor prognosis, surgery is the mainstay for treating angiosarcoma. Many reports suggest the outcomes of endarterectomy and EBR similar to those in the present study, although EBR, with a clean margin, has recently demonstrated a tendency to show better outcomes. Because the diagnosis of angiosarcoma is not confirmed preoperatively in many cases, as in the present case, the decision to perform EBR in patients with an intra-aortic mass remains a challenge; however, when preoperative diagnosis is not confirmed, endarterectomy may relieve the symptoms and help in confirming the diagnosis. In the present case, additional EBR was performed to improve the patient’s outcome, but the result was dismal. Splenic infarction was detected before the first operation, suggesting that metastasis may have already established. In such cases, additional radical therapy may not be appropriate. Given the highly malignant nature of angiosarcoma, palliative therapy to relieve symptoms should always be considered as mainstream therapy.
Conclusion

We present a rare case of primary angiosarcoma treated using surgical therapy, with an unsatisfactory outcome. Early diagnosis and prompt surgical therapy before an embolic event are desirable. Furthermore, angiosarcoma should be considered a possible diagnosis in patients with an atypical mass inside the aorta revealed by imaging.

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