Embolectomy Plus TEVAR in Advanced Stage Aortic Angiosarcoma with Acute Limb Ischemia: A Case Report

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ABSTRACT

Objective: To report the case of primary aortic angiosarcoma with bilateral adrenal metastasis coming with acute limb ischemia, including the management and pathological report. **Case presentation:** A 54 year-old female came to the hospital because of her back pain. CTA of the whole aorta revealed the posterior mediastinal mass invaded the aorta, bilateral adrenal mass and renal infarction. She was sent for endoscopic ultrasonography (EUS) with fine needle aspiration (FNA) at the mediastinal mass and adrenal mass. During the hospitalization, the patient developed acute limb ischemia on her right leg. CTA of the both legs showed absence of the distal run-off from the right common iliac artery. We performed femoral embolectomy and TEVAR. Pathological report showed malignant spindle cells and the immunohistochemistry suspected the angiosarcoma of the aorta. **Conclusion:** Primary aortic angiosarcoma is a very rare disease. There is no consistent approach to the management due to its rarity.

Keywords: Acute limb ischemia; adrenal metastasis; aortic angiosarcoma; TEVAR (Siriraj Med J 2017;69: 306-309)

INTRODUCTION

Primary angiosarcoma of the aorta is very rare. These cases are usually first seen with symptoms of aortoiliac occlusive disease or acute thromboembolism or other complications from the tumor. Most of the cases can be diagnosed during autopsy or pathological examination from the tumor thrombus removed from the vessels. No consistent approach to the management of patients with primary mediastinal sarcomas has been delineated because of sparse data.[1] We describe and report the case of primary aortic angiosarcoma with bilateral metastasis because of the rarity of the disease.

CASE PRESENTATION

A 54 year-old female came to the Police General Hospital, and complained of back pain for 2 months. The pain was located at the paravertebral area on the middle of the back, without radiated pain to another area. It was not related to her position and movement. She had no history of dysuria or abdominal mass. Her neurological test showed normal with full motor power and negative straight leg raising test. She was sent to do a chest X-ray and electrocardiography (EKG) because of hypertension. The EKG appeared normal, but widening mediastinum was seen in chest X-ray. CT angiography of the whole aorta was performed (Fig 1).

The CTA revealed lobulated peripheral enhancing isodensity mass 5.31 x 5.32 x 6.20 cm. at aortopulmonary region, abut inferior and left lateral aspect of aortic arch, which had invaded to aortic arch and descending thoracic aorta, and caused a narrowing of left main pulmonary artery. There was a filling defect within descending thoracic aneurysm, which was suspected to be tumor thrombus. Subcarinal lymphadenopathy was seen and measured

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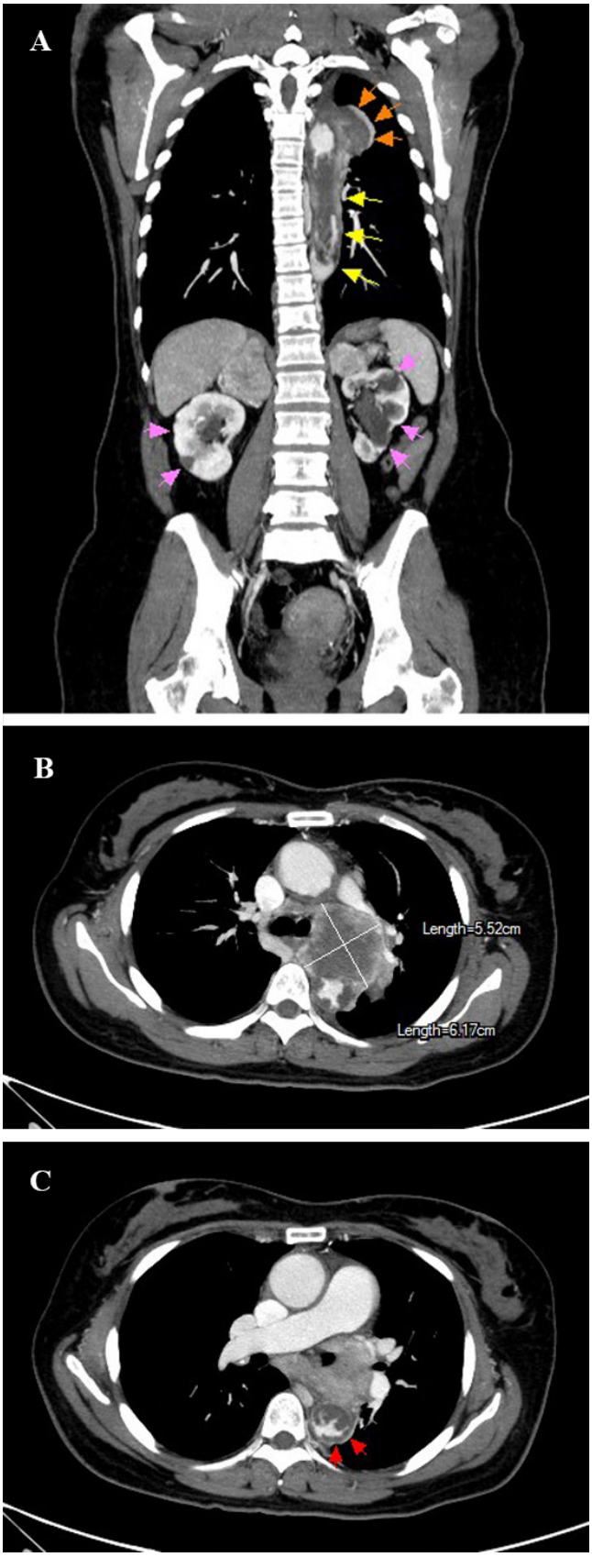


Fig 1. CTA of the whole aorta **A.** Mediastinal mass with aortic invasion (orange arrow), tumor thrombus at the descending aorta (yellow arrow) and bilateral renal infarction (pink arrow) **B.** Size of the mediastinal mass **C.** Tumor thrombus at the descending aorta (red arrow).

2.60 x 1.33 cm. Another two lobulated heterogenous enhancing masses 4.86 x 3.77 cm. and 4.78 x 3.87 cm. at bilateral adrenal glands were demonstrated. The patient was hospitalized on February 27, 2017 for further work-up.

During admission, she underwent the EUS with FNA at the mediastinal and adrenal mass for two times. The ultrasound showed a hypervascular mediastinal mass which had encased and invaded the aorta, soft in consistency with harder consistency for adrenal mass. The first pathological result was atypical round cells, but limited interpretation due to inadequate tissue yielded. Repeated biopsy showed spindle cell tumor suspicious for sarcoma with bilateral adrenal metastasis.

Six days after the second biopsy; the patient experienced pain with numbness at her right foot for 6 hours. The pain score was 10 out of 10. Her right leg was cold and mottled skin from the knee to toe, absent of pulse at right popliteal artery (PA), dorsalis pedis artery (PDA) and posterior tibial artery (PTA) (Fig 2). Inaudible doppler signal was demonstrated at right PA, DPA and PTA. The laboratory findings were: a hematocrit of 37.9%, a hemoglobin of 12 g/dl, white blood cells of 9,700 /mm^3, BUN of 12 mg/dL, a creatinine level of 0.9 mg/dL, PT 12.9 sec, APTT 50.2 sec, INR 1.20. Electrolytes were normal including HCO_3 28.7 mmol/L. The patient was sent for CT angiography of the lower extremities.

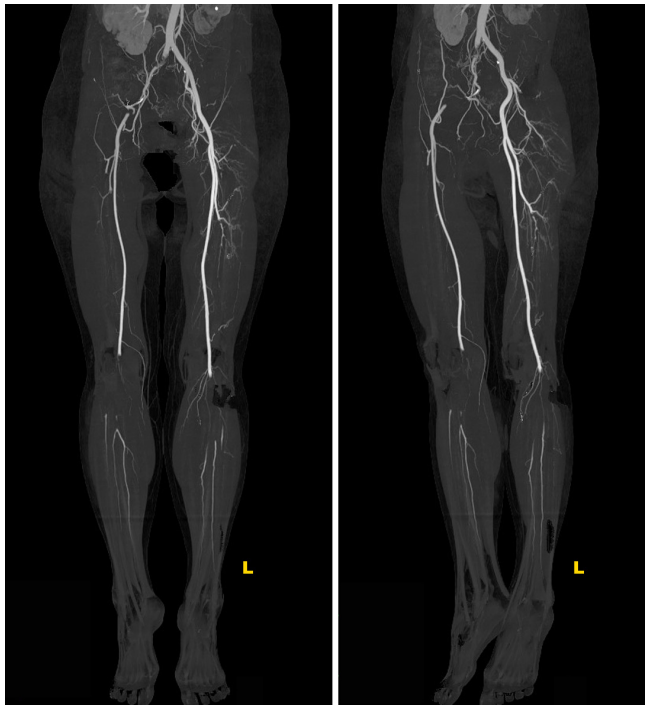


Fig 2. CTA of the both legs showed the defect and poor distal run-off.

The CTA showed the total occlusion of the right common iliac artery (CIA) and external iliac artery (EIA) with reconstitution flow from collateral to right common femoral artery (CFA), total occlusion of right PA, bilateral tibio-peroneal trunks and anterior tibial artery (ATA) with poor distal run-off (Fig 2). She was diagnosed of acute limb ischemia (Rutherford IIb), so we immediately set up an operation for embolectomy and TEVAR.

Reconstruction imaging from CTA showed proximal neck diameter of 26 mm, distal 23 mm. The total length of the lesion was 200 mm. Femoral artery diameter was 8.43 mm. We planned to deploy 2 stent grafts, 150 mm each with the overlap of 50 mm using Valiant Captivia Thoracic Covered Stent Graft Delivery System (VAMF3228C150TE and VAMC3026C150TE; Medtronic Vascular, Santa Rosa, California) as shown in Fig 3.

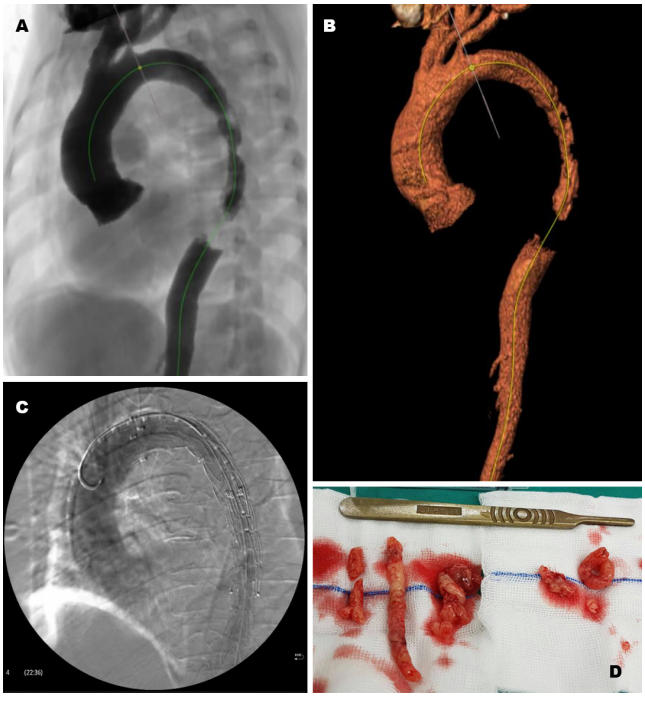


Fig 3. A,B) CTA demonstrated image of the lesion at descending aorta **C)** Fluoroscopic image of descending aorta after the stent graft was deployed **D)** Tumor thrombi.

After administration of general anesthesia and skin preparation, open arteriotomy was done at both groins (bilateral common femoral arteries). Tumor thrombus at right CFA was seen and removed with No.4 Fogarty balloon. Angiography of both lower extremities was done and showed the filling defect at bilateral popliteal artery. Embolectomy was done until good backflow was demonstrated. After finishing with the lower extremities, then aortography was done and filling defect was seen at descending aorta, distal to left subclavian artery. Self-expandable covered stent graft (32 to 28 mm and 30 to 26 mm) was placed just distal to left subclavian artery. Post-dilatation balloon was inflated. Complete aortography and angiography showed no tumor thrombus left. Distal pulses were fully palpated until DPA and PTA.

Third day post-operative, the patient was transferred from intensive care unit with a good clinical outcome, and the distal pulses were good after that. The patient can walk longer distance without pain. The pathological result from the tumor thrombus revealed malignant spindle cell tumor, suspicious of angiosarcoma. Tumor cells were negative of Caldesmon, and AE1/AE3, SOX10, S100, CD34 and EMA. CD117 and CD31 activities were diffusely positive. Fli-1 and thrombomodulin activities were focally positive. The counselling was done between oncologist and patient's relatives explaining the risk and benefit. They refused the chemotherapy and chose the palliative treatment.

About 2 weeks after the cover stent placement, the patient got worse. Supportive treatment and pain control were used. Twenty four days post-operative, the patient had a cardiac arrest and was under chest compression for 20 minutes. The patient's relatives wanted to stop CPR, so the CPR was stopped. Cause of death was not clearly known, with suspected cause of natural disease.

Discussion

Angiosarcomas are most often presented as cutaneous lesions, typically in the scalp or face of an elderly. However, angiosarcomas can occur in any anatomic site, including the deep soft tissue, breast, visceral organs, and bone.[2] Primary aortic angiosarcoma is a very rare disease. Primary malignant disease of the aorta was first described by Brodowski in 1873. He described the tumor as a fibrosarcoma of the aorta.[3] From that time till 1998, there were only 87 cases report of primary angiosarcoma and 70% of the cases had distant metastasis.[4] It is found two times more common in men than women, with age range from 48-85 years.[5] Distant metastases is common at the time of diagnosis. Because of the rarity of the disease, there is no prospective study to compare the management.[6] A recent journal suggested that complete resection of the tumor in combination with adjuvant therapy (chemoradiation) seems to offer the best chance of survival.[6,7]

In our case, the patient presented with mediastinal mass and adrenal metastasis. She was hospitalized and waiting for the histopathological report. The oncologist had a plan to consult for TEVAR before neoadjuvant chemotherapy session, after histopathological report became available. The report did not come on time, and she developed the acute limb ischemia caused from the

tumor thrombus. At that moment, we performed femoral embolectomy and TEVAR to control the tumor and to prevent the further emboli. Unfortunately, the patient and her relatives decided to refuse chemotherapy and went through the palliative treatment. The patient has a cardiac arrest and died on day 24 postoperatively.

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