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Case Report

A Case of Upper Limb Arterial Embolization from Aortic Arch Intimal Sarcoma

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We report a case with aortic intimal sarcoma who presented with left upper limb arterial embolization from tumor. A 79-year-old female patient presented with paleness and left upper limb paralysis. A transesophageal echocardiogram revealed a mobile and fragile mass attached in the aortic arch. Contrast-enhanced computed tomography showed a massive irregular tumor in the aortic arch with left common carotid and subclavian artery occlusion. Total arch replacement was performed, and tumor was resected en bloc. Although the postoperative course was uneventful, multiple metastasis to the limbs was observed. The patient died 6 months postoperatively.

Keywords: intimal sarcoma, embolization, total arch replacement

Introduction

Aortic sarcoma is a rare malignant tumor that is classified by its origin as either intimal or mural.[1] Most sarcomas originate from the abdominal aorta or descending thoracic aorta. We report a case with aortic intimal sarcoma who presented with left upper limb arterial embolization from aortic arch intimal sarcoma with dissemination.

Case Report

A 79-year-old female patient with chronic kidney disease who presented with paleness and paralysis of the left upper limb was admitted to the adjacent clinic and treated with antiplatelet medication and prostaglandin production. The patient was referred to our hospital because her symptom did not improve, and thrombectomy was performed under the diagnosis of arterial thrombosis of the left upper limb. On pathological findings, the presence of distinct tumor cells was not clear, but dysplastic cells suspected of malignant tumor were found. A transthoracic echocardiogram showed no tumor or thrombus in the cardiac cavities. Since she had suffered from chronic kidney disease, we did not perform contrast-enhanced computed tomography (CT) scan. A TEE revealed a mobile and fragile mass broadly attached in the aortic arch (**Fig. 1**). Since we decided to perform total arch replacement, contrast-enhanced CT was examined under hydration, showing a massive irregular tumor in the aortic arch with left carotid and subclavian artery occlusion (**Figs. 2A, B**).

The affected aorta was resected under hypothermic circulatory arrest with antegrade cerebral perfusion through median sternotomy, and total arch replacement was performed. A soft yellowish and granular tumor protruded into the aortic wall of the resected aorta (**Fig. 3**). The postoperative course was uneventful. However, multiple sarcoma lesions were recognized in the extremities during hospitalization. The patient rejected radiation therapy and chemotherapy after the operation and was transferred to

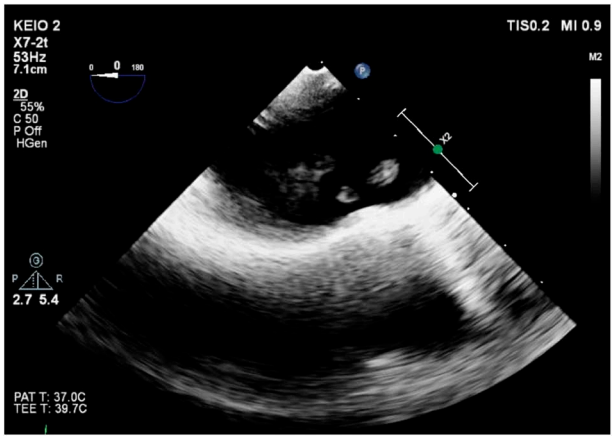

Fig. 1 Transesophageal echocardiogram shows a mobile mass broadly attached in the aortic arch.

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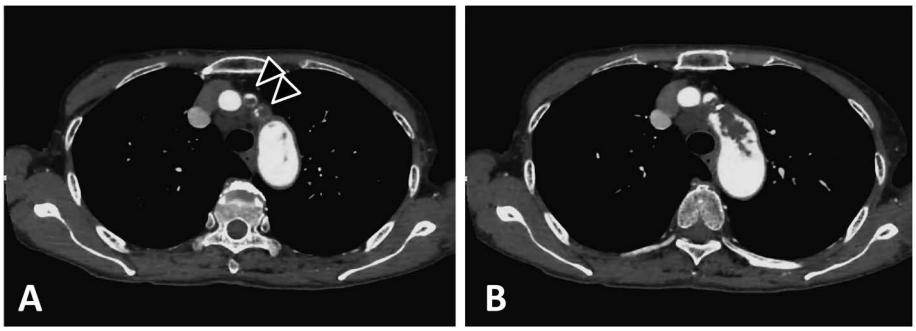

Fig. 2 Contrast-enhanced computed tomography shows an embolized left carotid and subclavian artery (**A**; arrows) and a massive irregular tumor in the aortic arch (**B**).

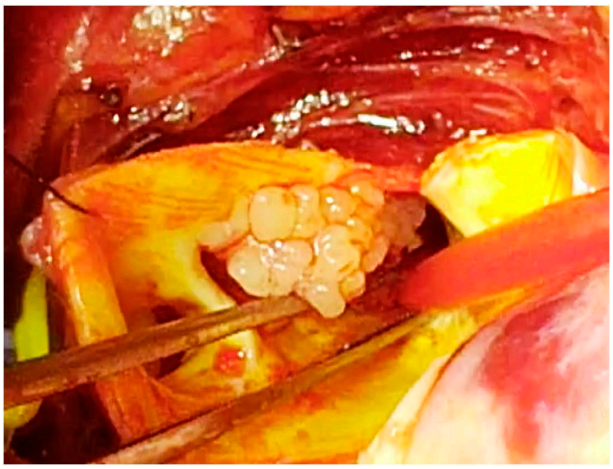

Fig. 3 Gross specimen in the aortic arch shows a yellowish and granular tumor adhering to the aortic wall.

another hospital for rehabilitation.

According to histopathological examination, the atypical spindle cells with pleomorphic nuclei had penetrated transmurally from the arterial intima to the adventitia. Further examination using magnetic resonance imaging revealed multiple metastatic lesions in the upper and lower extremities 3 months after the operation.

Discussion

Aortic sarcoma is a rare malignant tumor that is classified by its origin as either intimal or mural.[1] In this case report, a definite diagnosis of aortic intimal sarcoma was confirmed by histopathological examination. We report a similar case with aortic intimal sarcoma who presented with multiple cerebral infarction and disseminated metastatic lesions.[2]

According to the systematic review of aortic sarcoma in 2014, 165 cases have been reported since 1873, and the estimated median survival rate was 11 months in 122 patients diagnosed antemortem.[3] In terms of disease management, more prompt diagnosis and treatment consisting of surgical resection and postoperative chemotherapy or radiation therapy can possibly improve the survival rate of patients; however, its management results in palliative care because diagnosis is often made after the tumor advances to the end stage. Systemic examination is mandatory to reveal the location of metastasis. However, patients with metastatic lesions are not likely to live long even if the metastatic lesions are revealed due to its severe malignancy.

Conclusion

The prognosis of sarcoma is poor even if surgical intervention has been improved today. A multimodal approach is mandatory including chemoradiation and terminal care because clear guidelines have not been established.

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Disclosure Statement

All authors have no conflicts of interest associated with this report.

Author Contributions

Study conception: YI, AY
Data collection: YI
Analysis: YI
Investigation: YI, AY, HS
Writing: YI
Critical review and revision: all authors
Final approval of the article: all authors
Accountability for all aspects of the work: all authors

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