Case report - Vascular thoracic

The rupture of descending thoracic aorta due to the necrosis of aortic intimal sarcoma

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Abstract

Aortic intimal sarcoma is rare and the prognosis is very poor. We experienced a case of ruptured aortic intimal sarcoma in the descending aorta. A 69-year-old man underwent an emergency operation for the rupture of descending aorta. The postoperative course was uneventful. The histological examination of aortic wall showed aortic intimal sarcoma. The patient developed a local recurrence and abdominal dissemination of the tumor three months after surgery. We report the case and discuss about the diagnosis and treatment of thoracic aortic intimal sarcoma.

Keywords: Aortic diseases; Vascular neoplasms; Aortic rupture

1. Introduction

Primary malignant tumor of the aorta (PMTA) is rarely seen and the prognosis is poor. The most common growth pattern of PMTA is intimal type, which often forms intraluminal polyps and develops arterial embolism. However, aortic rupture due to PMTA is extremely rare. We report a rupture case of PMTA and discuss about the diagnosis and treatment of this rare disease.

2. Case history

A 69-year-old man visited a local hospital with two episodes of sharp back pain. His vital signs were stable. However, chest X-ray showed a right pleural effusion, and chest computed tomography (CT) revealed that the descending aorta ruptured into the right pleural cavity. A fusiform aneurysm with a diameter of 50 mm was found at the level of the 9th thoracic vertebrae (Fig. 1).

The patient was transferred to our hospital for emergency surgery. In the operating room, a chest tube was placed in the right pleural space, and 1400 ml of bloody fluid was drained. Then, the patient was positioned in the right lateral decubitus position. A left thoracotomy was made through the 5th intercostal space. A large hematoma was found around the descending aorta. Atherosclerotic finding inside the aneurysm was minimal. Instead, there was an intimal erosion (about 1 cm) on the right side of the aorta at the level of the 9th thoracic vertebrae, which was suspected as a rupture site. The aneurysm was resected and reconstructed with a woven polyester straight graft (Hemashield; Boston Scientific Corp, Natick, Mass; size = 24 mm). Proximal and distal anastomoses were performed in an end-to-end fashion. Histological examinations of the aneurysm wall showed malignant tumor cells in all layers of the aortic wall. The tumor growth was most evident in the adventitia (Fig. 2).

Tumor cells were seen on the surgical margins of both proximal and distal sides. Immunohistochemical staining showed a positive finding for smooth muscle actin and vimentin, and negative finding for smooth muscle myosin, factor VIII, CD31, CD34, D2-40, keratin and symaptophysin. The proliferation marker MIB-1 (Ki-67) stained about 20% of the malignant cells. These findings were compatible with sarcoma of the aortic intima.

3. Discussion

PMTA is rarely seen. The male/female ratio is 9:5, and the mean age is 59.5 years [1]. The favorite site of PMTA is
the thoracic descending aorta [1]. PMTA is categorized by not only histological pattern but also growth pattern. Salm [2] categorized aortic tumor into three growth pattern; intimal, polypoidal (intraluminal), and adventitial (or mural). The intimal and polypoidal types may have a tendency to extend along the intimal surface and frequently form intraluminal polyps, which may cause embolism. The adventitial type primarily involves media and/or adventitia. Most of the reported PMTA are the intimal type. In our case, although the tumor was mainly found in the media and adventitia, tumor cells extended along the intimal surface with thrombus formation. The histopathological diagnosis was intimal sarcoma. Some intimal sarcomas have been reported to have myofibroblastic differential growth pattern [3, 4], which was also seen in our case.

The prognosis of PMTA is poor, and survivals after diagnosis range from 8 to 14 months [1]. Intimal type sarcoma has the most aggressive behavior, leading to death within months [3]. Importantly, PMTA is barely suspected or diagnosed at the early stage because the symptoms are non-specific and PMTA is a very rare disease. In most cases, PMTA is diagnosed by histological examination following surgery for arterial embolism, aneurysm or rupture [1]. Rupture cases are less frequently seen than embolism cases. Cantena and colleagues reported a rupture case [5]. They suspected a PMTA intraoperatively and performed segmental resection of the descending aorta and a wedge lung resection. There was no recurrence or metastasis during a 21-month follow-up. Clear surgical margins are important for survival after surgical resection of PMTA. We could not achieve clear surgical margins since we did not suspect PMTA when we saw an erosive change inside the aneurysm. Although preoperative diagnosis is not easy on imaging studies, a PMTA should be suspected and an extended resection might be necessary when an atypical lesion or erosion is found inside the aorta during surgery.

References

eComment: Sarcomas of the great vessels. Is there a role for chemotherapy?

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We read with great interest the article by Tanaka et al. regarding a case of aortic intimal sarcoma [1]. We would like to highlight the role of adjuvant chemotherapy in a setting of multimodality treatment. Sarcomas of the great vessels previously diagnosed during surgery or autopsy are rare and highly lethal. This represents predominantly female patients between the age of 22 and 81. The prognosis is poor with a mean survival of 12 months after onset of symptoms and one and two years survival rates of 22% and 7%, respectively [2].

The mainstay of treatment for sarcoma is surgical resection as this remains the only potentially curative modality. Adjuvant chemotherapy and radiation can be considered following surgical excision although their role remains undefined. An ~20% response rate can be expected with a combination chemotherapy regimen involving an anthracycline and an alkylating agent; however, the value of this regimen in the adjuvant setting for intimal aortic sarcoma is unclear [3].

According to the literature, there are two cases with satisfactory survival. The first patient underwent resection followed by adjuvant chemotherapy [4]. The second one was found inoperable during surgery and underwent a two-drug combination chemotherapy consisting of ifosfamide and epirubicin with long-term survival [5].
In our institute, we had a similar unresectable case of primary pulmonary artery sarcoma. The patient underwent the same chemotherapeutic scheme and is still alive after six months, with a marked regression of the tumor. In conclusion, we think that intensive chemotherapy is worth trying in unresectable patients.

References


