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 operable 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


