

**Case
Report**

A Congenital Arteriovenous Malformation Originating from the Aorta Locating in the Posterior Mediastinum

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Vascular malformations located in the posterior mediastinum are extremely rare. Most of them are found coincidentally during routine examinations. Only a small percentage of these posterior mediastinal arteriovenous malformation cases may cause symptoms such as dyspnea due to compression of surrounding tissues. Radiologic imaging can be insufficient in some cases for differential diagnosis. Because of their vascular nature, diagnostic needle biopsy may have a high risk of bleeding. Open surgical resection is a safe treatment choice under many circumstances, and it helps the diagnosis as well. In this paper, a case of a 31-year-old male is presented with an incidentally diagnosed arteriovenous malformation, originating from the descending aorta and located in the posterior mediastinum.

Keywords: arteriovenous malformation, posterior mediastinum, aorta and adult

Introduction

Arteriovenous malformations (AVMs) are abnormal connections between arteries and veins without capillary vessels. Mediastinal AVMs are rare, and they are generally linked with a history of trauma. Most of the mediastinal AVMs are asymptomatic. We have operated a mediastinal AVM in an adult without known history of chest trauma. The computerized tomography (CT) and magnetic resonance imaging (MRI) examinations have showed the

vascular nature of the mass in this case successful surgical treatment of the mass is reported in this paper.

Case Report

A thirty one year old male patient has admitted to the department of neurosurgery with the diagnosis of kyphoscoliosis. A mass was visualized in the posterior mediastinum during magnetic resonance imaging (MRI) that was done for diagnosis of kyphoscoliosis. The mass was defined as a well contoured tumor ($43 \times 23 \times 61$ mm dimensions) with isointensed areas with the muscle tissue and heterogeneous hyperintensed areas, and some peripheral hypointense areas without any magnetic signals, locating in the posterior mediastinum at the level of T6-T8 thoracic vertebrae (Fig. 1). Diagnostic needle aspiration was postponed after the case was consulted with invasive radiologists as the hypointense areas were defined as calcification and fibrosis, whereas the low signal areas were arterial vascular structures. There was no connection with the spinal chord. The CT angiographic scan that was planned to define the vascular nature of the mass revealed a direct connection with the descending thoracic aorta in the posterior mediastinum. Also at CT angiography,

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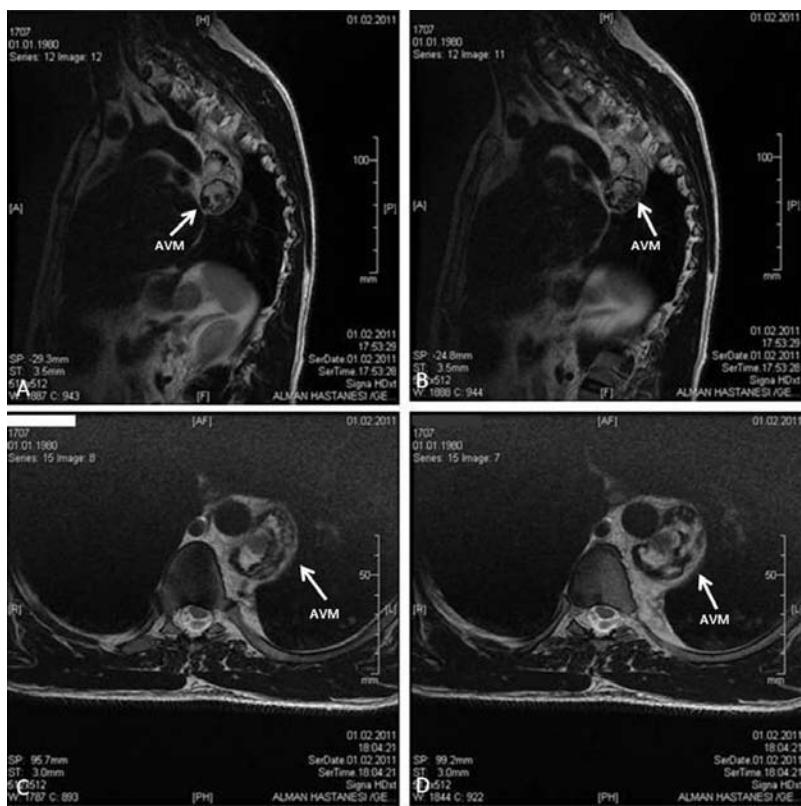


Fig. 1 The well-shaped Para aortic mass with heterogeneous hyperintense texture and peripheral hypo intensity in T2A sagittal (**A, B**) and axial (**C, D**) images.

the well contoured soft tissue was found as containing arterial vascular structure, which was hyperintense with the aorta (**Fig. 2A–2C**). There was no calcification.

There were no iatrogenic interventions, trauma or infection in the past medical history. The physical examination and laboratory tests were within normal limits. Needle biopsy was excluded due to bleeding risk. A transesophageal echocardiography (TEE) was performed to have differential diagnosis of any intra aortic pathology like dissection or aortic aneurysm, which was revealed a cystic lesion without direct connection with the aorta.

All diagnostic attempts described a tumor with vascular content. The possibility of a malignancy was not excluded, and needle biopsy was decided to have a high bleeding risk. Surgical resection was planned for definitive diagnosis and curative treatment of the tumor. Catheter intervention for embolization of the tumor was excluded, because that technique is not able to make the pathological diagnosis.

The patient was transferred to the department of cardiovascular surgery for resection of the vascular mass located in the posterior mediastinum. He was prepared for

thoracoabdominal approach and double lumen endotracheal tube was placed. Left posterolateral thoracotomy was performed under general anesthesia. After retraction of the left lung, the mass (approximately 60 mm diameter) was observed, attaching ascending aorta at 4-5-6th posterior intercostal level. A vascular connection with the aorta was detected during en bloc resection of the vascular mass. The aortic site was repaired with 4/0 pledgetted polypropylene suture material. A venous drainage was not visualized during surgery due to massive bleeding of the mass. The intraoperative diagnosis was that the extremely vascular mass could be a malign tumor. The excised tumor was sent to the department of pathology. His postoperative course was uneventful and he was transferred to the ward on postoperative day 1.

The histopathological examination showed that the excised mass (40 × 35 × 65 mm dimensions) was an arteriovenous malformation (**Fig. 3**). A postoperative CT scan has verified the en bloc excision of the mass (**Fig. 2D–2F**). He was discharged without any complication at the end of the 6th postoperative day. He had no complications and was healthy at postoperative week 6.

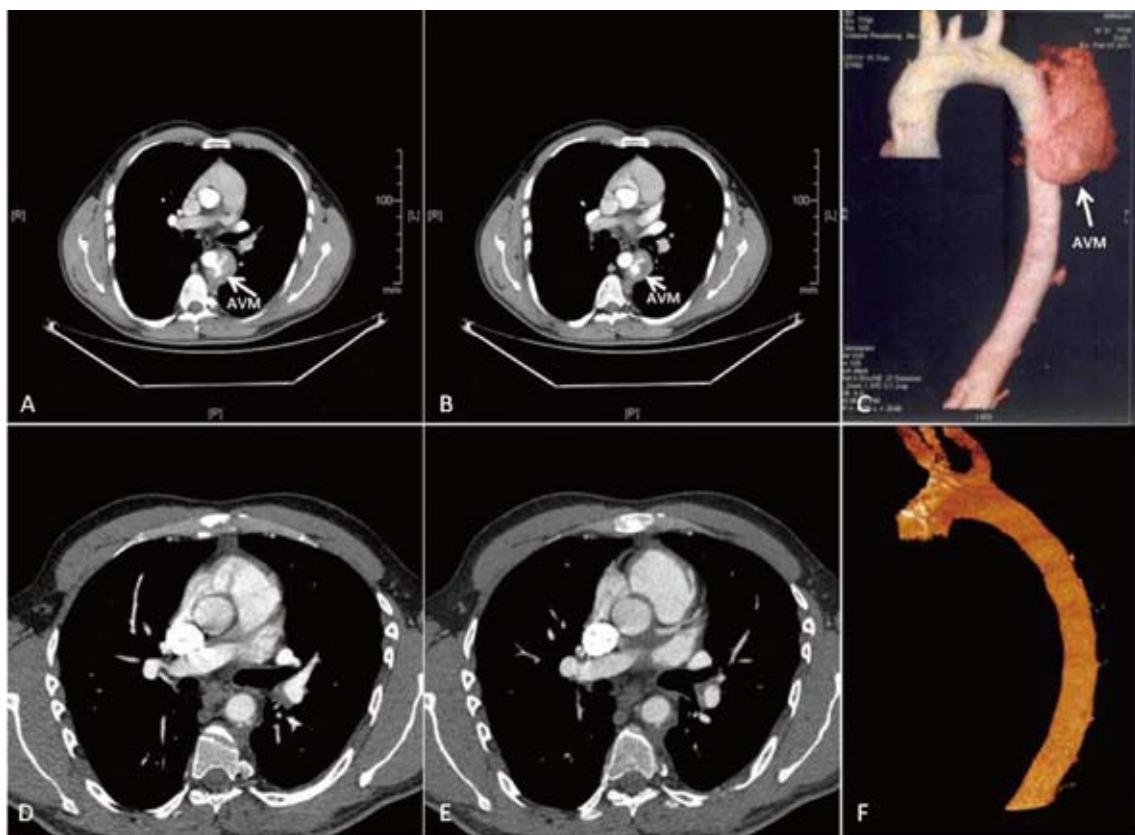


Fig. 2 Pre- and postoperative CT angiographic arterial phase scans: Well shaped AVM originating from the aorta with hyper dense areas in preoperative images (**A–C**) and en bloc of the mass without any residues in postoperative scans (**D–F**).

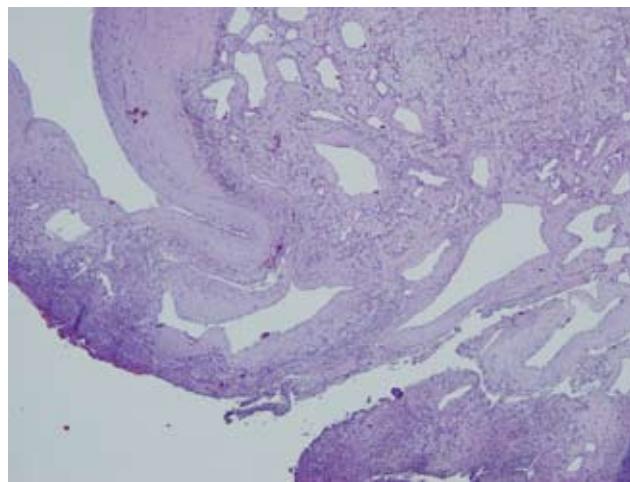


Fig. 3 Thin and thick walled dilated vascular structures in histopathological examination ($\times 4$, hematoxylin & eosin (H-E)).

Discussion

Arteriovenous malformations (AVMs) are abnormal connections between arteries and veins without capillary vessels. They can exist in any organ or tissue. The symptoms are depending to the dimensions and location. There may be one or more tortuous feeding arteries and draining veins. Mediastinal AVMs are rare, and they are generally linked with a history of trauma. Congenital mediastinal AVMs are even rarer and there are only few cases reported before.^{1–6)} Most mediastinal AVMs are asymptomatic, and they are incidentally diagnosed during routine radiological imaging. They may be symptomatic when they are enlarged or infected and have compressed surrounding tissues, which can lead to congestive heart failure.⁵⁾ Although there is a risk of hemorrhage, no spontaneous rupture has been reported.⁴⁾

CT and MR imaging tools can visualize the relationship of the mass with the surrounding tissues and mediastinal great vessels, but they may be insufficient for detection of the vascular origin. Conventional angiography

may be a useful method in this situation. Diagnostic needle biopsy may be hazardous in those cases. CT and MR have showed the vascular nature of the mass in this case, but the feeding arteries and draining veins were not visualized, which was an atypical situation for AVMs.

Other vascular malformations (hemangiomas, lymphangiomas), tumors (angiolipomas, angiosarcomas) and extra lobar pulmonary sequestration have to be kept in mind for differential diagnosis. The hemangiomas are round or lobulated, well margined, homogenous, low density masses containing 10% of flebolites in CT scans without contrast media.⁷⁾ There is a heterogeneous slow contrast media accumulation in late phase with contrast CT scan.^{7,8)} They are hypointense in T1A scans and bright hyperintense in T2A scans on MR imaging.^{8,9)} Although the mass was well margined and shaped in our case, the non-signal surrounding the vascular imaging on MR and the vascular contrast uptake in the arterial phase on the CT scan were defined significant for the diagnosis of an AVM. The T2A images were not homogeneously bright and hyperintense, which was different from hemangiomas. Angiolipomas are very rare in the mediastinum; they are generally subdermal located in the forearm. Their “dumbbell” shape is diagnostic.¹⁰⁾ The aortic tumors, such as angiosarcomas, are polypoid, mainly originated from the intima, and they can obstruct the vessel lumen and can cause embolic events.¹¹⁾ Pulmonary sequestration is manifest with recurrent infections during childhood.

Treatment choices for the AVMs are surgical resection or embolization. Silent cases can be followed conservatively without any intervention. A preoperative embolization of big tumors can decrease the amount of surgical bleeding.^{1,2,6)} Embolization was defeated preoperatively, because radiologic findings were not sufficient to exclude a malignancy. Our surgical plan was to excise the tumor and repair the aorta (primary or graft interposition).

In our case, we have performed a successful surgical

resection of an atypical AVM for histological diagnosis and definitive treatment.

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