

Azygos Vein Aneurysm Forming a Mediastinal Mass

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Aneurysm of the azygos vein is a rare cause of mediastinal mass that is normally detected radiologically in asymptomatic patients. In some cases a diagnosis is made by noninvasive methods, such as computed tomography and magnetic resonance. However, in the present case imaging revealed what appeared to be a solid mass. When the mass had been excised by thoracotomy, the histopathologic diagnosis was aneurysm of the azygos vein.

Aneurisma de la vena ácigos como masa mediastínica

El aneurisma de la vena ácigos es una causa muy infrecuente de masa mediastínica y habitualmente se trata de un hallazgo radiológico en pacientes asintomáticos. Aunque en algunas ocasiones se puede llegar al diagnóstico por métodos no invasivos, como la tomografía computarizada y la resonancia magnética, en nuestro caso la imagen obtenida se asemejaba a una masa sólida. Se intervino a la paciente mediante toracotomía y se procedió a la exéresis de la tumoración. El diagnóstico histopatológico fue de aneurisma de la vena ácigos.

Key words: *Azygos vein. Aneurysm. Mediastinal mass.*

Palabras clave: *Vena ácigos. Aneurisma. Masa mediastínica.*

Introduction

Although solid masses in the posterior mediastinum can include neurogenic tumors, schwannomas, neurofibromas, ganglioneuromas, and cystic lesions, vascular causes should not be forgotten. We describe a case with a confirmed diagnosis of azygos vein aneurysm and discuss the possible etiology, pathogenesis, diagnostic methods, and treatment of this entity.

Case Description

The patient was a 49-year-old female homemaker with no relevant medical history, history of substance abuse, or respiratory symptoms. A chest radiograph before an appendectomy in February 2004 revealed a posterior mediastinal mass, and the case was referred to the pneumology department for analysis. Physical examination, biochemical analysis, blood test results, spirometry, and electrocardiography were completely normal. Contrast-enhanced computed tomography of the thoracic and abdominal regions revealed a well-defined oval-shaped retrotracheal mass, 2.7 cm 2 cm, in the posterior

mediastinum extending to the carina. The mass made minimal contact with the right margin of the esophagus (Figure 1). We observed no axillary or mediastinal lymph node enlargement suggesting malignancy, and no abnormalities of the pulmonary parenchyma were detected. The upper abdomen presented no abnormal images. Nor were enlarged retroperitoneal lymph nodes detected.

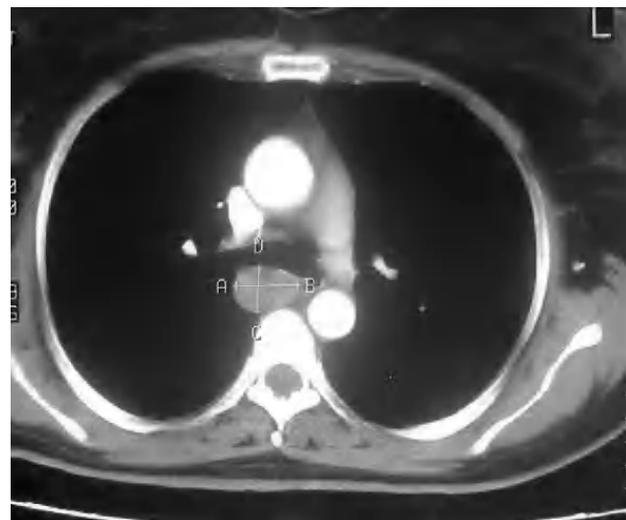


Figure 1. Contrast-enhanced computed tomography scan of the thorax showing a well defined mass in the posterior mediastinum. The lesion was a high-density (54 Hounsfield units) mass measuring 2.7 2 cm.

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The location of the mass might have suggested a cystic lesion, but the density of 54 Hounsfield units was too high for that interpretation. To differentiate between a cystic lesion and a solid tumor, we requested magnetic resonance imaging. A well defined lobulated right paravertebral mass, approximately 3 cm × 3.5 cm × 1.5 cm, was seen at the D5-D6 level; T1 weighted images showed a signal intensity isointense to muscle, and T2 weighted images showed a hyperintense signal with hypointense zones in the lower region. No cortical erosion or spinal canal involvement was observed. Heterogeneous enhancement of the lesion was seen after injection of intravenous gadolinium (Figure 2). These findings seemed to point in the direction of a neurogenic tumor as the first diagnostic possibility.

With this suspicion in mind we referred the patient to the thoracic surgery department for evaluation. Surgical excision by thoracotomy was scheduled and subsequently revealed an azygos vein lesion, which was resected in block followed by ligation at its proximal and distal ends as well as ligation of the tributary intercostal veins. There were no complications. Histopathology confirmed an aneurysm of venous vascular structure. The patient's postoperative course was satisfactory.

Discussion

Aneurysms are persistent, abnormal vascular dilations. Most reports describe arterial aneurysms and they are said to have a predilection for certain locations. Reports of venous aneurysms are scarce in the literature and their pathology and natural history are yet to be defined.

The etiology of venous aneurysms has been insufficiently studied. Such aneurysms can be classified into 2 types: primary and secondary. Secondary venous aneurysms can be attributed to trauma, infection, venous valvular insufficiency, and arteriovenous fistulas resulting from increased blood flow, which can be caused by certain heart diseases or pulmonary vein abnormalities that can lead to obstruction of blood flow to the right chambers.¹ Our patient reported no history of trauma and there was no sign of heart failure or liver disease.

The rarity of azygos vein aneurysm is evident. Dilation of the azygos system, on the other hand, is common, and it arises principally as a result of an increase in central venous pressure. There are other causes, such as portal hypertension, thrombosis, agenesis and obstruction of the inferior vena cava, pregnancy, and states of high flow. In such cases dilation is directly related to increased blood flow since the azygos and hemiazygos veins form a collateral system. The most common cause of azygos vein aneurysm is reported to be inferior vena cava aplasia.^{2,3} Nevertheless, no venous anomalies or dilation of the azygos system were found in our patient.

The pathogenesis of aneurysms is also unknown. Mention has been made of the onset of a venous hypertrophic process or endothelial phlebosclerosis.⁴ Other authors support the hypothesis of congenital weakness or degenerative changes in the venous wall due to abnormalities in the connective tissue.⁵ This hypothesis is based on the demonstrated relevance of those factors in the genesis of arterial aneurysms. In

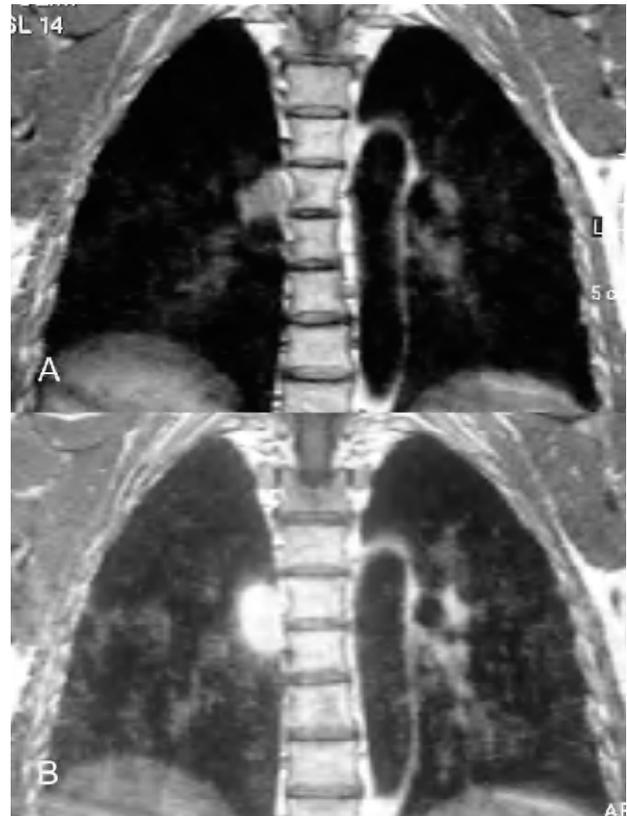


Figure 2. Magnetic resonance image of the chest (cardiac collimation). The T1 weighted image with no gadolinium (A) shows a paravertebral mass with a signal intensity isointense to muscle. The T1 weighted image following administration of gadolinium (B) shows intense heterogeneous contrast enhancement.

endothelial hypertrophy, there is proliferation of connective and elastic muscular tissue in the intimal layer of the venous wall as a response to mechanical stress. Endothelial venosclerosis is a degenerative process characterized by loss of connective tissue and smooth muscle cells. Schatz and Fine⁶ considered these changes comparable to those responsible for arteriosclerosis. Pathology of resected veins have demonstrated the absence of the longitudinal muscular layer of the adventitia and, in other cases, fibrosis of the intima with an atrophied middle muscular layer.⁶

More cases of venous aneurysm have been reported among women than men although the difference is not statistically significant; nor is the difference significant for age or location of the aneurysm. Most patients are asymptomatic and the lesion is detected in chest radiographs. In the present case, retrospective identification of the cause of the azygos vein aneurysm was not possible; however, since the patient's medical history, physical examination, and radiographs ruled out trauma and increased pressure of the azygos system, we assumed the origin to be congenital.

The cardinal venous system develops during the third and fourth weeks of gestation. The system consists of the anterior and posterior cardinal veins, which unite to form the common cardinal vein. The posterior cardinal

veins are replaced by 2 pairs of additional veins—the subcardinal and supracardinal veins—which feed into a portion of the inferior vena cava and the azygos system. A segment of the right supracardinal vein forms an anastomosis with a section of the superior vena cava (which in turn branches off from the anterior cardinal vein); thus, the azygos vein drains into the right atrium through the superior vena cava.^{7,8} Accordingly, the azygos vein can be considered the confluence of 3 embryonic veins: the right supracardinal vein (azygos vein), the right posterior cardinal vein (branching off from the azygos), and the right anterior cardinal vein (superior vena cava). This confluence is considered to be an anatomically vulnerable point at which aneurysms may develop. It has been postulated that aplasia and hypoplasia of the superior vena cava can affect drainage of the azygos vein⁹; however, in the present case, the vena cava was observed to be unaffected.

Veins are composed of 3 layers: the intima, media, and adventitia. During vasculogenesis the first layer to develop is the intima; the media develops once a stable vascular pattern is in place (through interaction of the epithelium and the mesenchyma). In case of congenital dilation, the venous layer most affected is the media, in contrast to acquired dilation (varicose veins), where endothelial fibrosis is observed. Detection of abnormalities in these veins requires histologic study.¹⁰

Radiographs of patients with azygos vein aneurysm may show a prominent azygos vein or an abnormal density in the mediastinum. In a chest radiograph, the size of the aneurysm usually changes as the patient breathes, stands, or performs a Valsalva maneuver. This is of great importance in avoiding potentially dangerous diagnostic tests, such as fine needle aspiration or mediastinoscopic biopsy.¹¹ In the past, venography was employed for diagnosis, but computed tomography and magnetic resonance are currently the noninvasive techniques for evaluating vascular abnormalities and they can help rule out other causes of azygos vein dilation. In the case we report, blood flow in the venous aneurysm was slow, such that the magnetic resonance image was similar to that of a solid mediastinal mass. A similar observation was reported for another case.¹²

Among reported complications arising from venous aneurysms are the following: thrombophlebitis, pulmonary embolism, venous rupture or obstruction, and pressure effects on adjacent structures. Surgical

treatment of a venous aneurysm is indicated to prevent such complications or as a response to chest pain or dyspnea or an increase in the size of the aneurysm. The greatest risk of surgical treatment is pulmonary embolism resulting from manipulation of the aneurysm. With this risk in mind, physicians can take a conservative approach to managing asymptomatic patients or those in which aneurysm size does not increase. Despite the risk, some authors, such as Pasic et al,¹³ recommend prophylactic surgery to treat venous aneurysms of the mediastinum. Surgery was indicated in the case we report because diagnostic images were interpreted as suggesting a neurogenic tumor. Therefore, even though azygos vein aneurysm is rare, it should be considered in the differential diagnosis of a mediastinal mass.

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