Giant aneurysm of the thoracic aorta. Answer to January e-quid

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Case study

G.H., a 76-year-old woman with no particular prior medical history was seen in the emergency room for a 5-month history of progressive dyspnea, which became stage IV dyspnea one week prior to admission. The physical examination revealed a patient who was conscious and hemodynamically stable, with signs of congestive heart failure with left pleural effusion. The laboratory tests showed renal failure. A chest X-ray was done (Fig. 1).

Figure 1. Frontal chest X-ray in a reclining position.
What is your diagnosis?

Based on your reading of the case study, which of the following would be your diagnosis?
- mesothelioma;
- pleurisy with effusion;
- giant hydatid cyst;
- aplectasis;
- pneumonia;
- thoracic aortic aneurysm;
- aortic dissection presenting as pleural effusion.

Diagnosis

Giant aneurysm of the thoracic aorta.

Imaging data

The frontal chest X-ray revealed an opaque left hemithorax (Fig. 2), with a homogeneous projection image, with no mediastinal displacement or widening of the intercostal spaces, and no air-fluid level or bone abnormalities. Analysis of that left hemithoracic opacity also showed fine arcuate calcifications in the shape of an incomplete circle to the left side of the trachea, as well as parallel linear calcifications on the left chest wall. The latter could be related to parietal calcifications from an intracavitary expansive process, such as a hydatid cyst (endemic in our field). However, their positioning, which seemed to silhouette an elongated vertical formation, was more suggestive of a vascular structure. This, therefore, favored the hypothesis of a giant aneurysm of the thoracic aorta.

The workup was completed by a Doppler ultrasound of the chest, which showed the presence of a voluminous ovoid formation occupying the entire left hemithorax, in which there was arterial flow only at the anechoic center. In view of this data suggesting a thoracic aortic aneurysm with associated parietal thrombosis, a CT-scan of the chest and abdomen was done, without injection of iodinated contrast material due to the renal failure, confirming the diagnosis (Fig. 3). That study showed a thoracic aorta with aneurismal dilatation over its entire length, whose descending segment was fusiform and extended to the diaphragm, thus occupying almost the entirety of the left hemithorax. That segment, spreading over a length of 22 cm, measured 11 cm in its largest diameter. Its wall was finely calcified in places and was slightly hyperdense, confirming the existence of parietal thrombus. Furthermore, there was also minimal homolateral free pleural effusion. Medical treatment was initiated and the patient is still being followed by a cardiovascular surgery practice.

In addition to its exceptional size, the thoracic aortic aneurysm described in this case study is unusual in that it was revealed by an opaque hemithorax, a unique diagnostic circumstance to our knowledge.

Discussion

A thoracic aortic aneurysm (TAA) is defined as a loss of parallelism of the aortic walls, resulting in saccular, fusiform, or diffuse dilatation. To be diagnosed as a TAA, that dilatation must have a diameter 1.5 times greater than the superjacent aorta. That ratio is only 1.3 in case of dystrophy or bicuspid disease. However, in practice, any aortic segment exceeding 4 cm in diameter may be considered aneurysmal [1].

The true incidence of TAA is difficult to determine because there are so many asymptomatic cases, but it is constantly increasing due to improved diagnostic methods and the aging population [2]. Clouse et al. estimated that incidence to be 10.4 cases per 100,000 population per year [3]. In 40% of cases, TAA is discovered by chance during a physical examination or on a chest x-ray performed for a different reason.

Symptomatic cases of TAA express chest pain in 37% of cases and dorsal pain in 21%. That pain is induced by compression of the mediastinal organs and chest wall, or by erosion of an adjacent bone (sternum, vertebra) [4,5]. Sometimes that pain is severe and involves a previously unaffected area or one that was previously less painful, indicating that rupture is imminent. Other symptoms of TAA are signs of congestive heart failure, thromboembolic complications leading to stroke, ischemia of the extremities, renal or mesenteric infarction. Aneurysms of the ascending aorta may be revealed by major complications such as fissuration, rupture of the pericardium or mediastinum, or secondary dissection. Aneurysms of the aortic arch and the descending aorta may be revealed by a superior vena cava syndrome, signs of compression of the aerodigestive tract such as cough, dyspnea, recurrent pulmonary infections, and dysphagia and dysphonia in case of recurrent nerve involvement [5].

Atheromatous and hereditary diseases of the elastic tissue contribute greatly to the pathogenesis of aortic dilatation. Atheromatous disease is the predominant etiology of...
TAA in 72 to 92% of cases [6]; it is almost exclusively at cause in aneurysms of the descending aorta. Hereditary elastic tissue diseases are in second place and include: Marfan’s syndrome, Ehlers-Danlos syndrome type 4, Beals syndrome, and aortoannular ectasia. Marfan’s syndrome is the most common dystrophy of the connective tissue; it is an autosomal dominant disease in 75 to 85% of cases, associated with aneurysmal dilatation of the ascending aorta in 80% of cases. In addition to its atheromatous and dystrophic origin, there are numerous other less common etiologies: infectious, syphilitic, and tubercular causes (rare nowadays); inflammatory arterial diseases: Horton’s arteritis, Takayasu’s arteritis, Behçet’s syndrome, and “primary” inflammatory aneurysms; traumatic causes and aortic dissection.

The clinical presentation of a TAA is vague or even asymptomatic; therefore imaging plays a crucial role in the morphological assessment and the assessment of operability or accessibility to endovascular treatment by placement of a stent, as well as in post-therapeutic monitoring. The chest x-ray is abnormal in 80% of cases: it may show calcified aneurysmal walls, mediastinal widening, enlargement of the aortic knob, or tracheal deviation [5]. The CT angiogram is the study of choice, useful for determining the site of the aneurysm, its largest diameter, its extent, its relationship with the aortic branches, and possible presence of other aneurysmal localizations. The MRI is the second-line study and provides the same information at the CT angiogram. It is reserved for non-dialyzed renal failure patients, young aortoannular ectasia patients, and for monitoring patients who have been diagnosed but have not undergone surgery, due to the need for repeated studies. The transthoracic echocardiogram makes it possible to examine the aortic sinus and the ascending aorta; the other segments are accessible by

Figure 3. Axial CT slices in mediastinal window at the superior (a) and inferior (b) mediastinal level, showing a calcified, aneurysmal aortic wall, progressively increasing in diameter, involving the ascending portion, aortic arch, and descending portion; the latter is correctly analyzed in the coronal (c) and oblique sagittal (d) reconstructions, the site of a giant fusiform aneurysm measuring 11 cm in its largest diameter, extending over a length of 22 cm; its wall is slightly spontaneously hyperdense in connection with an extensive parietal thrombus.
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a transesophageal approach. Arteriography is no long used for initial evaluation of a TAA, but it may be performed to locate the artery of Adamkiewicz prior to surgery. Surgery and endovascular treatment are the therapeutic options available to patients.

In conclusion, TAA is a serious condition that is rarely symptomatic. Its discovery due to an opaque hemithorax has never been reported. The different cross-sectional imaging methods, led by CT angiography, provide a thorough morphological assessment, detection of complications, and post-therapeutic follow-up.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References