Giant thoracic aortic aneurysm. About a clinical case

Aneurisma gigante de aorta torácica. A propósito de un caso clínico

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Summary

Recognition of thoracic aortic aneurysms is important as they can have devastating complications if left undiagnosed or untreated promptly. The following is a clinical case of a 42-year-old male patient, in whom a giant aneurysm of the ascending thoracic aorta and aortic arch was diagnosed, the largest published in the literature to date. Early identification and referral for surgical intervention are vital to improve the morbidity and mortality of these patients. The objective of this article is to emphasize the current role of the different imaging methods in the detection and diagnosis of aortic pathology. Thoracic Aortic Aneurysms are increasingly being recognized on Angio-Tomography, which provides excellent detail of the aorta and its relationships with adjacent structures. In the evaluation of aneurysmal disease, cross-sectional images are particularly important in depicting the extent of disease, total aortic and arterial diameters, and the extent of mural thrombus formation.

Key words (MeSH)

Aortic aneurysm, thoracic
Computed tomography angiography
Radiography, thoracic

Palabras clave (DeCS)

Aneurisma de la aorta torácica
Angiografía por tomografía computarizada
Radiografía torácica

Resumen

El reconocimiento de los aneurismas aórticos torácicos (AAT) es importante ya que pueden tener complicaciones devastadoras si no se diagnostican a tiempo o no se tratan oportunamente. A continuación, se expone un caso clínico de un paciente de 42 años de edad, de sexo masculino, a quien se le diagnosticó un aneurisma gigante de aorta torácica ascendente y arco aórtico, el más grande publicado en la literatura hasta la fecha. La identificación temprana y la derivación para la intervención quirúrgica son vitales para mejorar la morbilidad y la mortalidad de estos pacientes. El objetivo de este artículo es enfatizar en el papel actual de los distintos métodos imagenológicos en la detección y diagnóstico de la patología aórtica. Los aneurismas aórticos torácicos se reconocen cada vez más en angio-TAC, la cual proporciona excelentes detalles de la aorta y sus relaciones con estructuras adyacentes. En la evaluación de la enfermedad aneurismática, las imágenes transversales son particularmente importantes para representar la extensión de la enfermedad, los diámetros aórticos y arteriales totales y la extensión de la formación del trombo mural.

Introduction

An aortic aneurysm is defined as an abnormal and permanent focal dilatation of the aorta, with a maximum transverse diameter that exceeds the normal range for age, sex and body surface area by 50% (1). Intrathoracic aortic intrathoracic diameters in normal adults have been published in the literature for computed tomography (CT) with respect to sex and age (2). In general terms, the upper limit values (99th percentile) for populations of Western origin are 40 mm in men and 34 mm in women. Giant thoracic aortic aneurysms are arbitrarily defined in the literature as aneurysms with a maximum transverse diameter of more than 10 cm (3).

Recognition of thoracic aortic aneurysms (TAAs) is important, as they can have devastating complications if left undiagnosed and untreated (4). These potential complications include rupture, dissection, luminal thrombus embolism, compression of an adjacent structure, and aortic valve distortion with valvular insufficiency (in aortic root aneurysms). The natural evolution of the aortic aneurysm is influenced by its expansion rate, location, complications (aortic insufficiency, rupture, dissection, thrombosis) and its underlying causes (Marfan syndrome, Ehlers-Danlos syndrome, infection, inflammation or atherosclerosis). And, above all, the annual rate of complications is influenced by the diameter of the TAA, so prophylactic surgery is recommended in patients with diameters greater than 55 mm in ascending aorta and 65 mm in descending aorta (1, 4). The following is a clinical case of a patient with a TAA that stands out for its extreme maximum transverse diameter at the time of diagnosis, with a high initial risk of complications.

Clinical Case

A 42-year-old male patient with a personal history of hypertension and smoking. He consulted the emergency department for asthenia, adynamia, fever, cough and expectoration. In the physical examination there were no alterations of note.

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Posteroanterior and lateral chest X-ray showed mediastinal widening predominantly on the left with a radiopaque image of lobulated and well-defined contours, showing vessels through it. In the lateral radiograph it occupies the anterosuperior mediastinum and part of the retrosternal clear space. The mass makes sign of the silhouette with the ascending aorta and part of the arch with which it loses its limits (figure 1).

With the diagnosis of an anterior mediastinal mass that could correspond to a lymphoma or have an aortic vascular origin, an aortic angiotomography is performed and volumetric and multiplanar reconstructions (MPR) are performed. Figure 2 shows selected images in the axial plane at two levels, sagittal and coronal, respectively, showing extensive aneurysm in the middle and distal sector of the ascending aorta and the aortic arch. Note the correlation between the mediastinal widening identified on chest radiography and the coronal reconstruction of the aneurysm on angiotomography.

The aneurysm measured 11 × 15 cm in its maximum transverse diameters. It presented a circumferential mural thrombus, with a permeable lumen of 6 cm. The proximal sector of the ascending aorta measured 4.0 cm. Figure 3, corresponding to RV reconstruction, identifies the relationship of the aneurysm with the supra-aortic trunks, which emerge from the aneurysm, with proximal dilatation of the brachiocephalic arterial trunk with a caliber of 22 mm; the rest of the supra-aortic trunks were of usual caliber. Distal to the aneurysm, the descending thoracic aorta is tortuous, of usual caliber, as is the abdominal aorta. The aneurysm determined mass effect due to compression of the trachea and bronchi sources.

The left brachiocephalic venous trunk was not observed and its topography identified collateral circulation in probable relation to chronic thrombosis.

The transthoracic echocardiogram showed: left ventricle of normal dimensions with preserved global function (LVEF 66%); remaining cardiac cavities with normal dimensions, and trivalve aortic valve, without evidence of aortic insufficiency at Doppler. Corroborates the findings of the CT angiography with respect to the aneurysm of the ascending aorta and aortic arch.

After resolution of the respiratory picture, the patient was taken to surgery which confirmed a large aneurysm of the ascending aorta and aortic arch (Figure 4a). The ascending aorta and aortic arch were replaced, from the sinotubular junction to the beginning of the descending thoracic aorta, with a 28 mm Dacron prosthesis, with reimplantation of the supra-aortic trunks (Figure 4b).

The patient had a favorable evolution and was discharged seven days postoperatively.

Discussion

The incidence of TAA has increased in recent years due to the increase in life expectancy in the population and the greater availability of diagnostic techniques. It is estimated that there are about 10 cases per 100,000 person-years worldwide (1, 5). A giant TAA is defined as an aneurysm with a maximum diameter greater than 10 cm, its incidence and prevalence are not well established in the literature highlighting that only isolated cases are published.

As for giant aneurysms of the aortic arch, 10 cm in diameter are also considered for its definition, but these are very rare. In the literature there are only a few published cases, but it should be noted that none were found with the dimensions of the aneurysm as large as in the patient described here.

Most of the published cases are of aortic arch malformations with associated giant aneurysm (6) and only one shares anatomical characteristics similar to those of the patient in this case (7).

According to clinical suspicion, the support of imaging techniques is essential for the assessment of the aorta. Chest radiography is useful to evaluate the contour, size and location of the thoracic aorta and great vessels, which, if abnormal, would warrant the performance of complementary studies as in the case described. The advent of modern CT and magnetic resonance imaging (MRI) techniques has made non-invasive high-resolution evaluation of the vascular system a reality, without the risks of the trans-arterial catheterization procedure. MRI, in spite of its known advantages, such as high contrast resolution and the fact that it does not use ionizing radiation, continues to be a less accessible method, with longer duration and high cost compared to CT. Therefore, in the evaluation of aneurysmal disease, CT images are particularly important (8). The possibility of acquiring images in multi-slice CT with excellent spatial resolution, isovolumetric reconstructions and short acquisition times, allows optimizing the use of contrast medium and obtaining angio-CT images. From the images acquired in the axial plane, the appropriate post-processes can be performed to locate and characterize the aneurysm, perform its measurements: transverse diameter in the axial plane to the vessel, diameter of the necks, length of the aneurysm, relationship with supra-aortic trunks, evaluation of mural thrombosis and signs of rupture or pre-rupture (9, 10).

Early identification and referral for surgical intervention are vital to improve morbidity and mortality in these patients. In this case, no predisposing factors for TAA formation were identified, the patient has no diagnosis of Marfan syndrome, nor family history of the same or of other connective tissue diseases. He also has no family history of TAA or sudden death. The cumulative probability of rupture or dissection increases with time and the main risk factor is the initial aortic diameter. Aortic diameter is one of the most important risk factors for rupture, dissection and aneurysmal death. For ascending aortic aneurysms larger than 6 cm, the risk of rupture, dissection or death is 15.6% (11).

It is worth highlighting the importance of mural thrombosis in a study published in 2000 that included 87 patients to monitor the progression of thoracic aortic aneurysms: the average rate of aneurysm expansion was 1.43 mm per year, which increases exponentially with increasing aortic diameter (12), highlighting that the presence of mural thrombus is associated with accelerated growth.

Conclusion

Giant aneurysms of the aortic arch are very rare. There are no published series of patients and the evidence is reduced to sporadic cases. Knowledge of the causes, significance, imaging and possible complications of aortic aneurysms is essential for a rapid and accurate diagnosis. TAAs are increasingly recognized on CT angiography, which provides excellent detail of the aorta and its relationships with adjacent structures.

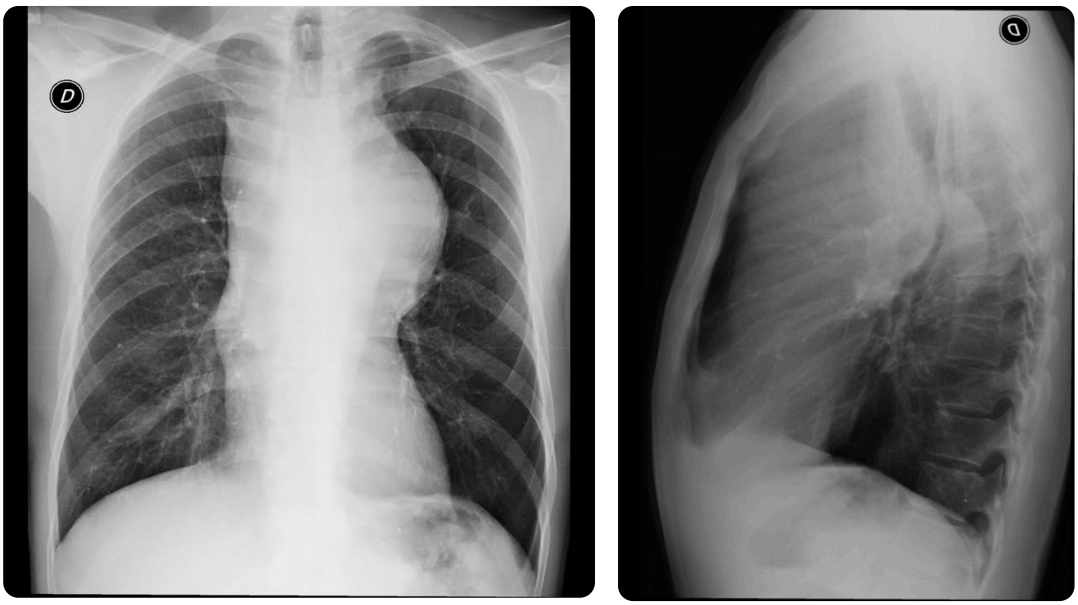

Figure 1. Chest X-ray. On the left, a posteroanterior view, in which the mediastinum is bilaterally widened with left predominance. On the right, the profile projection, showing a decrease in the retrosternal space, which may correspond in this topography to dilatation of the ascending aorta, with effacement of the aortopulmonary space.

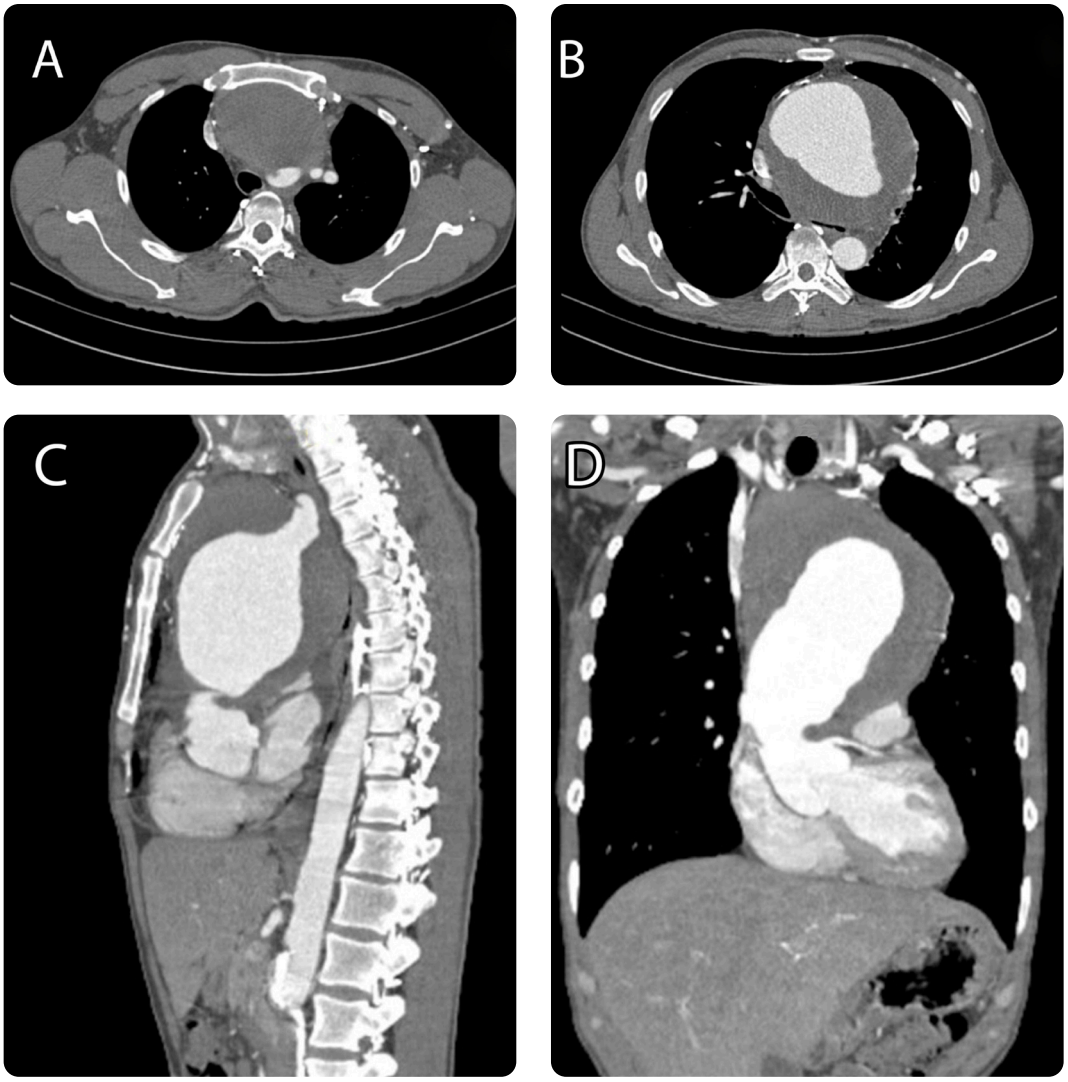

Figure 2. a). Axial angio-CT scan of the aorta showing a low density image due to thrombus in the anterior mediastinum, which displaces the trachea, the tracheoesophageal angle and the supra-aortic vessels laterally. b) Intramural thrombus surrounding the aortic lumen on its left lateral side. Descending aorta of usual appearance and diameter. c) Multiplanar reconstruction of aortic CT angiography in sagittal plane, showing the relationship of the aneurysm with adjacent structures and the emergence of the supra-aortic trunks. d) Multiplanar reconstruction of aortic CT angiography in coronal plane.

Figure 3. RV reconstruction of aortic CT angiography in a left lateral view. The emergence of the supra-aortic trunks and the normal descending thoracic aorta are highlighted.

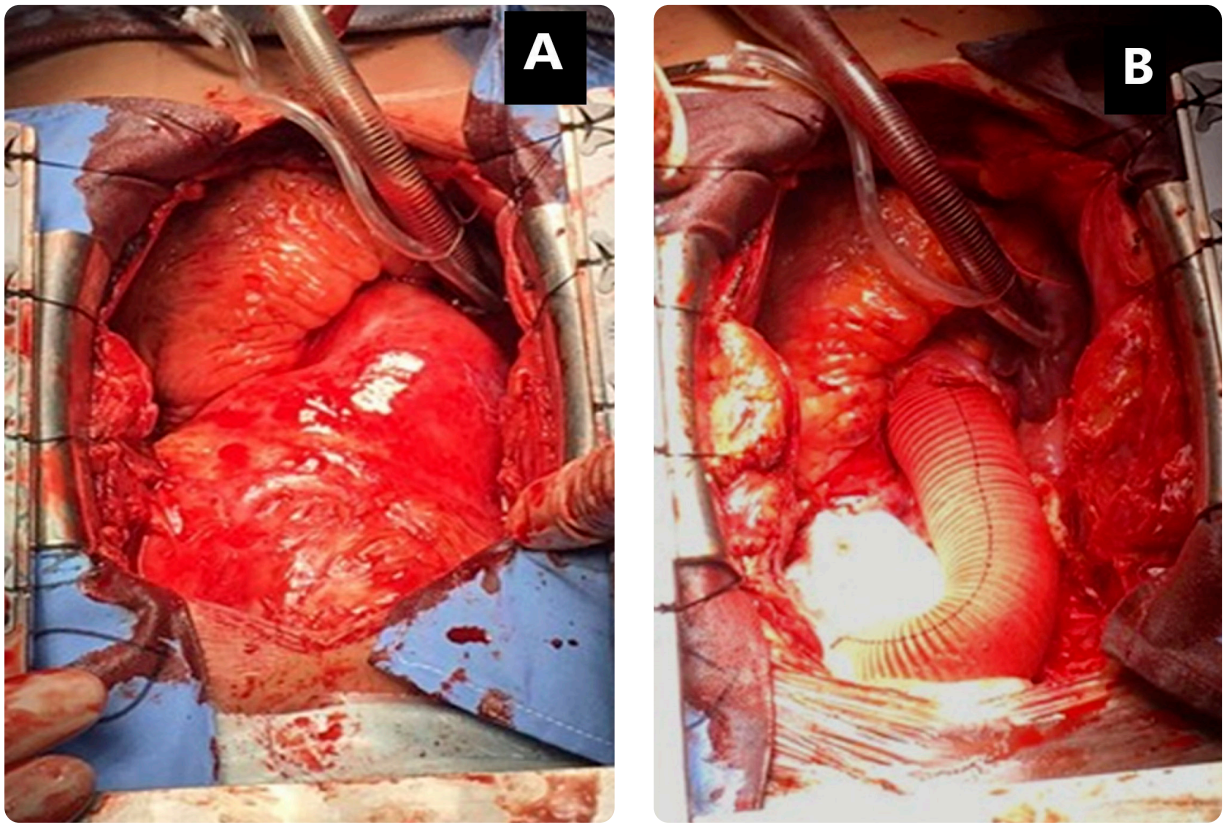

Figure 4. a) Surgical field confirming a large aneurysm involving ascending aorta and aortic arch. b) Final surgical result with prosthetic tube placement from the sinotubular junction with reimplantation of the supra-aortic trunks.

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