Multidetector Computed Tomography in the diagnosis of aortic pseudocoarctation. Two case reports and literature review

Martín Mela, Valeria Carrozza, Diego Haberman, Adriana Martínez, Enrique Gurfinkel.

INTRODUCTION

Aortic pseudocoarctation is an elongation and kinking of the thoracic aorta which typically compromises the distal portion of the aortic arch and the proximal descending portion, closely related to the insertion site of the ligamentum arteriosum (1, 2).

It is usually asymptomatic and diagnosis is generally incidental on imaging scans (chest x-rays, computed tomography -CT- or Magnetic Resonance Imaging -MRI-) indicated for other reasons.

No serious complications occur in its natural course, and therefore it does not require surgical treatment (3).

We report two cases diagnosed by multidetector computed tomography angiography (MDCT angiography), which had been performed for various indications, with aortic pseudocoarctation not being the initially suspected diagnosis.

We performed a literature review and update of this condition.

PATIENTS AND METHODS

Scans were performed with a multislice scanner of 64 rows of detectors (Toshiba, Aquilion®), using a radiation dose modulation software (Sure Exposure), with no electrocardiographic gating at effective doses of 6 and 7 mSv.

Images were obtained using a 0.5-mm slice thickness, 0.3-mm reconstruction interval, 0.828 pitch factor and a tube rotation time of 0.5 seconds.

Intravenous non-ionic contrast agent (Xenetix 350, Temis Lostaló®) was injected by infusion pump (Medtrad, Stellant®) at a dose of 1 ml/kg and a flow rate of 4 ml/sec. Multiplanar and three-dimensional (3D) reconstructions were performed at workstations (Vitrea, Vital Images ®).

Case 1

Fifty-nine year-old male patient with a diagnosis of severe aortic stenosis and scheduled valve replace-.
Aortic pseudocoarctation

Regular follow-up Doppler ultrasound scans showed a significantly restricted valve opening with severe calcifications of leaflets and commissures. Although bicuspid valve was suspected because of the extent of calcifications, this could not be demonstrated by trans-thoracic Doppler imaging. No other cardiovascular structural abnormalities were found and cardiac catheterization remained as a pending issue.

The patient had a history of drug-controlled hypertension.

Peripheral pulses were symmetrical, with no differences between upper and lower limbs.

Laboratory tests were within normal values. As part of a routine pre-surgical test, a chest x-ray was performed, which showed widening of the superior mediastinum which was assumed to be of vascular origin.

Given the clinical suspicion of thoracic aortic aneurysm, a chest MDCT angiography was performed, showing and elongation and kinking of the aorta involving the transition between the distal region of the aortic arch and the proximal descending portion, with no evidence of significant stenosis of arterial lumen with characteristics of pseudocoarctation (Figs. 1, 2, 3, 4 a, b, c).

The patient underwent aortic valve replacement surgery. Progress was good, with no complications, and the patient was discharged 72 hours after surgery.

Fig. 1: Aortic pseudocoarctation. MDCT angiography with 3D reconstruction. Left lateral view. Kinking in the proximal descending portion of the aorta, at the site of insertion of the ligamentum arteriosum (arrow).

Fig. 2: Aortic pseudocoarctation. MDCT angiography with sagittal reconstruction and MIP (maximum intensity projection) technique. The arrow points towards aortic kinking.

Fig. 3: Aortic pseudocoarctation. MDCT angiography with sagittal reconstruction showing kinking in the distal aortic arch. Severe calcifications in the aortic valve, pointed by arrows.
Case 2

Seventy-six year-old male patient who presented at our department to undergo MDCT angiography indicated for follow-up of known ascending aorta aneurysm.

Relevant medical history included aortic valve replacement with metal valve implantation in 2001. The reason for replacement had been severe aortic stenosis from bicuspid valve. Pre-surgical testing included a standard angiography, reporting severe aortic valve stenosis, mild dilatation of the ascending aorta and tortuosity of the proximal descending aorta.

Subsequent postoperative follow-up was performed by transthoracic Doppler ultrasonography, not reporting evidence of pseudocoarctation.

In the last Doppler scan, because of a poor acoustic window, proper measurement of the diameter in the tubular portion of the ascending aorta could not be performed. The patient’s primary cardiologist decided to complement the study with a computed tomography scan.

The MDCT angiography performed for follow-up of the ascending aorta diameters showed postoperative changes in the aortic valve, dilatation of the ascending aorta and elongation of the thoracic aorta at the site of the ligamentum arteriosum, with characteristics of pseudocoarctation (Figs. 5 and 6).

DISCUSSION

Aortic pseudocoarctation is a relatively rare congenital anomaly defined as a tortuous elongation of the distal segment of the aortic arch and the proximal descending aorta portion, posterior to the origin of the left subclavian artery. It is not associated with significant

---

**Fig. 4 a, b, c: Aortic pseudocoarctation. MDCT angiography showing axial images in three consecutive slices in the cranio-caudal direction. The arrows point towards aortic kinking, showing the characteristic morphology of this anomaly.**

**Fig. 5: Aortic pseudocoarctation. MDCT angiography with oblique reconstruction and MIP technique showing pseudocoarctation, pointed by arrow. Absence of collateral circulation.**

**Fig. 6: Aortic pseudocoarctation. MDCT angiography with 3D reconstruction. Left lateral view. Calcifications on the anterior aspect of the proximal descending aorta at the site of insertion of the ligamentum arteriosum.**
Aortic pseudocoarctation

luminal narrowing and there is no collateral circulation.

It was first described in the literature in 1951 by Dotter, Steinberg and Souders, who were part of three working groups. They introduced the term currently used to designate this condition, which, albeit known, was associated with other entities and had no name of its own (1, 4, 5).

The exact etiology is unknown. It has been proposed that the cause is a failure of fusion of the embryologic dorsal segments, which would result in the development of this anomaly (6). Another proposed theory makes reference to fibrosis of the ligamentum arteriosum, which would cause traction of the proximal descending aorta, approximating the adjacent pulmonary artery, with the consequent associated aortic tortuosity (7).

This condition usually has no associated clinical symptoms, and patients generally have normal blood pressure levels, with preserved and symmetric peripheral pulses.

The association of pseudocoarctation with other cardiovascular abnormalities is common, and similar to those associated with true coarctation.

Smith and Edwards propose that both conditions would have a common histopathologic substrate. Malformations include bicuspid aortic valve, subaortic stenosis, atrial and ventricular septal defects (8-10).

A study conducted in 1986 retrospectively analyzed 12 cases of pseudocoarctation. In 9 patients, associated cardiovascular disease was confirmed (7 cases of valve disease, 1 atrial septal defect and 1 ventricular septal defect) and the clinical symptom was cardiac murmur. In the other 3 patients, there were no associated anomalies and diagnosis resulted from incidental imaging findings (1).

In the absence of typical signs or symptoms of this condition, imaging studies are essential for diagnosis. Chest x-ray may show unspecific widening of the superior mediastinum. MDCT angiography and MRI are highly effective to show the typical morphology of this disease, and to assess other related cardiovascular abnormalities.

Both techniques have advantages and disadvantages inherent to the imaging approach. MDCT angiography is an extremely fast method. Patient preparation, and technical programming takes approximately 5 minutes, with a 10- to 15-second apnea. From this perspective, different MRI sequences are used in multiple planes determining a longer test. Furthermore, is well known the impossibility of performing MRI to patients with claustrophobia, implanted pacemakers

Figure 7: True coarctation. MDCT angiography with MIP reconstruction. Focal narrowing of the proximal descending aorta with collateral circulation that bypasses the obstructed segment. Patent ductus observed as associated anomaly. Arrowhead.

Figure 8 a, b, c: True coarctation. Axial MDCT angiography. showing axial images in three consecutive slices in the cranio-caudal direction. (a) The arrows point collateral circulation of significant development, typically absent in pseudocoarctation. (b) At the exact site of coarctation, descending aorta with severe narrowing (arrow). (c) Aorta has recovered its diameters (arrows). Arrival of a collateral vessel that has bypassed the site of narrowing (arrowhead) is observed.
or other ferromagnetic objects. However, CT has an essential limitation in the use of radiation. The use of contrast agents provides highly valuable information, in both methods (MRI and CT), but iodine used in CT may have more frequently adverse effects.

In our report, diagnosis was done by MDCT angiography. In one case the imaging study had been indicated for mediastinal widening and suspected aneurysm, in the other one it was indicated for follow-up of aortic root diameter.

Differential diagnosis with true coarctation–where there is a focal narrowing of the proximal descending aorta–should be made.

Depending on the degree of stenosis at the level of the coarctation, a significant pressure gradient may be observed between the proximal and distal portions of the stenosis, with development of collateral circulation [1, 2] (Figs. 7 and 8 a, b, c).

In imaging studies, differential diagnosis should also be made with aneurysms or vascular rings.

Pseudocoarctation remains usually stable and asymptomatic over time, and therefore does not require surgery. It should be monitored by imaging studies, since, with time a low percentage may develop aneurysmal dilatation distal to the site of pseudocoarctation, related to turbulent flow [2].

CONCLUSIONS

MDCT angiography makes it possible to study in detail the characteristics of pseudocoarctation as well as associated cardiovascular anomalies. It also allows an accurate differential diagnosis between this condition and true coarctation or other aortic anomalies.

References