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Aortic Hypoplasia, an Extremely Rare Cause of Hypertension
Revista Argentina de Cardiología, vol. 82, núm. 5, octubre, 2014, pp. 425-426
Sociedad Argentina de Cardiología
Buenos Aires, Argentina

Disponible en: http://www.redalyc.org/articulo.oa?id=305332235018
In the study of refractory hypertension (HT), the importance of imaging tests to identify its etiology is well established. We describe the case of a male patient who underwent a transthoracic echocardiography and a chest CT angiography, resulting in the definitive diagnosis of the extremely rare cause of his HT.

We describe the case of a 49-year-old male patient with a history of amaurosis fugax (normal brain CT scan). During the study of refractory HT, a transthoracic echocardiography was performed, showing the bicuspid aortic valve with raphe between the right and left coronary cusps. No significant transvalvular aortic gradient was detected, with mild eccentric aortic regurgitation. Valve annulus was 25 mm and the ascending aorta was dilated (44 mm). The aortic arch was 32.6 mm, narrowing just before the beginning of the descending thoracic aorta at the level of the supra-aortic trunks (diameter 15.6 mm), with end-systolic acceleration of flow at that level but no significant gradient (Figures 1A & B). There was evidence of normal left ventricular size and thickness, with preserved systolic function.

In view of these findings, a chest CT angiography was performed, showing aneurysmal dilation of the ascending thoracic aorta (anteroposterior diameter, 44 mm), and a caliber change in the region of the aortic arch with diameter of 18 mm, and descending aorta of 22 mm (Figure 2).

All the findings were consistent with hypoplasia of the thoracic aorta, causing the patient’s refractory HT.

Coarctation of the aorta is defined as a significant narrowing of the aortic lumen that causes severe obstruction of blood flow. This process affects an isolated short segment of the aorta.

Coarctation is most often found at the junction of the ductus arteriosus and the thoracic aorta, representing a 98% of all aortic coarctations.

When the narrowing affects longer portions of the aorta, it is known as tubular hypoplasia of the aorta, an exceedingly rare anomaly (1) on which there are very few reports in the literature associating it with secondary HT. (2)

First described by Quain, (3), the etiology of this condition is not well known and congenital and acquired causes have been proposed, (4, 5) although most of the authors turn to a congenital origin.

In the case of our patient, the diagnosis of hypoplasia of the thoracic aorta was associated with the presence of bicuspid aortic valve.

Symptoms typically occur within the first three decades of life and include HT, lower extremity claudication, and mesenteric ischemia.

One of the most severe complications is the HT resulting from this condition. Hypertension is often severe and, if untreated, can lead to potentially life-threatening complications, such as stroke, heart failure or kidney failure. In our case, the patient had a history of amaurosis fugax, with no evidence of changes in the brain CT scan.

Diagnosis is made using imaging techniques, such as echocardiography or CT angiography.

### References


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Reconstructive surgery is the optimal treatment option and must be tailored depending on the characteristics of the disease. Balloon dilation has been used in the treatment of coarctation of the aorta. However, this method has not proved to be effective in patients with aortic hypoplasia affecting long segments. (6)

Fig. 1. Transthoracic echocardiography. A. Suprasternal view showing the aortic arch and the descending thoracic aorta; an evident change in diameter and narrowing just before the beginning of the descending thoracic aorta (diameter, 15.6 mm) at the level of the supra-aortic trunks are also seen. B. Suprasternal view with continuous Doppler at the beginning of the descending thoracic aorta, showing end-systolic acceleration of flow at that level but with no significant gradient.

REFERENCES


Carcinoid Syndrome

We present the case of carcinoid syndrome with cardiac involvement in a 53 year-old female patient with a history of (poorly controlled) hypertension, eclampsia, and chronic renal failure (CRF). She consults a cardiologist for palpitations and dyspnea in variable functional class. During the guided interrogation the patient referred a long-term history of flushing and a two-year history of diarrhea.