CASE REPORT

Aortic coarctation with great juxtaductal calcified aneurysm. A rare presentation in childhood

José Daniel Espinoza-Hernández, MD, FACS [a], Rogelio Marroquín-Romero, MD [b], Carlos Bernardo Villarreal-Rubio, MD [b] , Ulises Venegas-González, MD [a], and Manuel Alfredo Osuna-Izaguirre, MD [c]

[a] Servicio de Cirugía Cardiotorácica, Hospital General Regional # 1 del Instituto Mexicano del Seguro Social, Tijuana, Baja California, MEXICO
[b] Servicio de Anestesiología Cardiovascular, Hospital General Regional # 1 del Instituto Mexicano del Seguro Social, Tijuana, Baja California, MEXICO
[c] Servicio de Cardiología Pediátrica, Hospital General Regional # 1 del Instituto Mexicano del Seguro Social, Tijuana, Baja California, MEXICO.

A female 11-year-old, was diagnosed with aortic coarctation and a great juxtaductal calficied aneurysm. Successful surgical repair through a left thoracotomy was performed and with left heart bypass. The coarctation and aneurysm was removed. A 20-mm Dacron graft Hemashield (Meadox Medicals) was interposed. The postoperative course was uneventful, there was adequate control of blood pressure. She was discharged 7 days after operation. It is very rare the association of calcified aneurysm and coarctation in childhood.

Key words: **Aortic Aneurysm; Aorta/Aortic, Coarctation; Calcification.**

Una paciente femenina de 11 años de edad fue diagnosticada con coartación aórtica y un gran aneurisma yuxtaductal calcificado. Se realizó una reparación quirúrgica exitosa a través de una toracotomía izquierda y con el uso de un Bypass izquierdo. La coartación y el aneurisma fueron removidos y interpuso un injerto Dacron Hemashield de 20 mm (Meadox Medicals). La evolución postoperatoria transcurrió sin incidentes. Hubo un control adecuado de la presión arterial. Fue dada de alta 7 días después de la operación. Es muy rara la asociación de aneurisma calcificado y coartación aórtica durante la infancia.

***Palabras clave*: Aneurisma Aórtico; Aorta/Aórtico; Coartación; Calcificación.**

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Most patients with aortic coarctation are diagnosed and undergoing corrective surgery at an early age. However, there are patients who undergo surgery until late childhood or adulthood. Thoracic aortic aneurysms are mostly related to coarctation described in adult patients or postoperative of aortic coarctation [1].

Surgery on the thoracic aorta without spinal cord protection for traumatic rupture of the aorta or thoracic aneurysm is associated with cord ischaemia up to 24% of patients [2]. The child with coarctation, however, usually develops a good collateral circulation. Because of this fact, the incidence of cord damage in surgery for aortic coarctation is 0.3 – 1.5%. Nevertheless, in later childhood it is more common, with a maximal incidence of 4% in the 11-16-year-old age group. It has been reported that after 15 minutes of aortic occlusion, despite of the quickest anastomotic techniques, the patient may be at high- risk of paraplegia[3].

The use of shunts kind of left heart bypass and hypothermia have been shown to be protective for the spinal cord during other types of surgery on the descending aorta. Left heart bypass to protect the spinal cord in patients undergoing operations on the thoracic aorta was first described in 1957 by Cooley and Gerbode [3,4].

Left heart bypass has the advantage of collecting blood coming from a low pressure system; a cannula is usually placed in the left atrium to drain the blood toward the centrifugal pump, which, in turn, takes back the blood throughout a cannula placed in the descending aorta. The central venous pressure must be maintained above 12 mm Hg. Body temperature greater than 35C and arterial medium pressure above 65 mmHg. The heart beats and perfuses the upper portion of the body while the bypass protects the distal circulation [4].

We present a case of juxtaductal calcified aneurysm and coarctation in childhood successfully operated on by using the left heart bypass.

CLINICAL CASE

This case is about an 11-year-old girl with no history for calcium metabolism disorders or any other traumatic event.

Corresponding author: Dr. José Daniel Espinoza-Hernández
email: jdehcardiotx@gmail.com

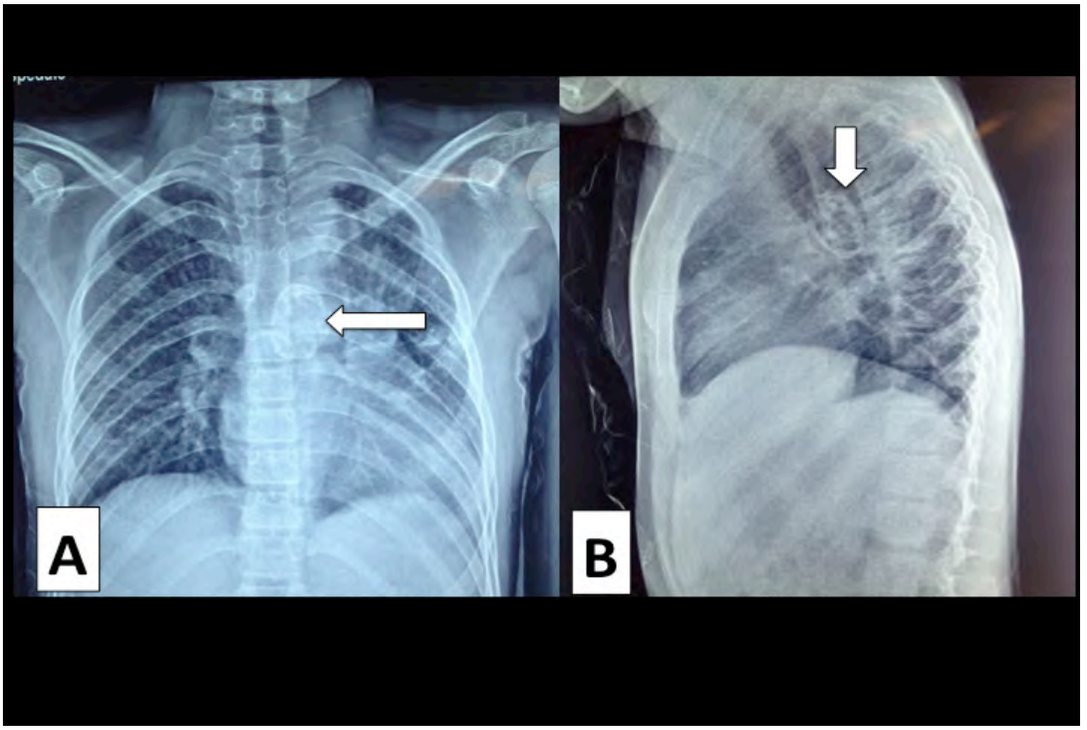

Figure 1. Chest Roentgenogram showing a calcified aneurysm in mediastinum.

Physical examination revealed severe systemic hypertension with arm-cuff pressure 190/90 mmHg and leg-cuff pressure of 80/40 mmHg. Carotid thrill was present, and a systolic murmur was found throughout the precordium. Both femoral pulses were importantly diminished. A chest roentgenogram with oval calcified image sizing 4.0 cm x 3.0 cm in the mediastinum slightly on the left side (**Fig.1**). Transthoracic echocardiogram showed a juxtaductal aortic coarctation with normal left ventricular ejection fraction.

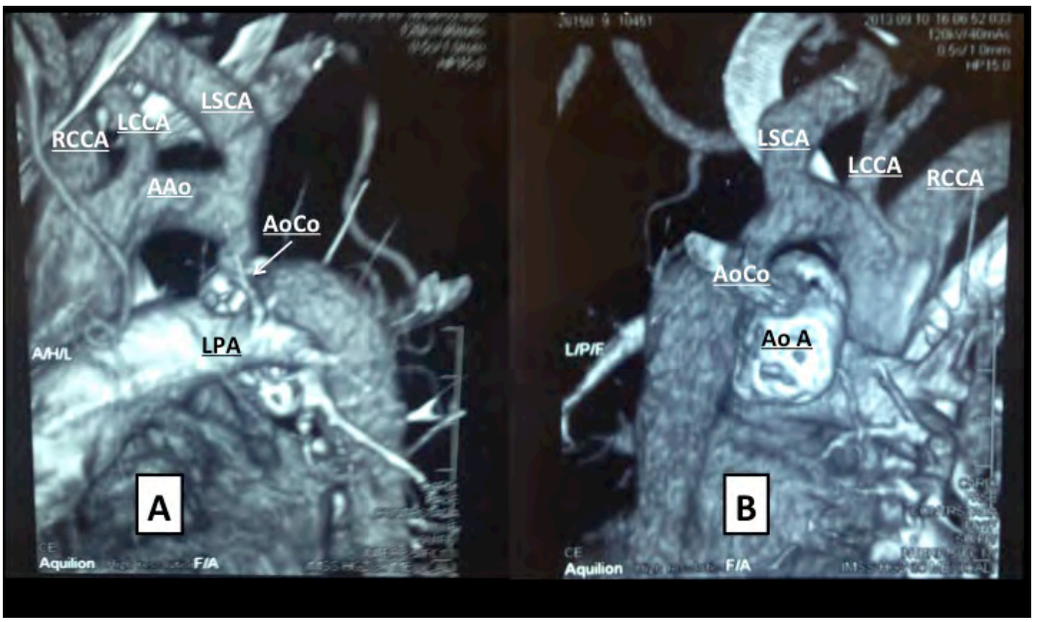

Figure 2. Preoperative 3D CT scan views. (A) Frontal view showing mild hypoplasic arch and coarctation. (B) Posterior view showing the great aneurysm. AoA=Aortic Arch; AoCo= Aortic Coarctation; LCCA= Left Common Carotid Artery; LPA= Left Pulmonary Artery; LSCA=Left Subclavian Artery; RCCA= Right Commom Carotid Artery.

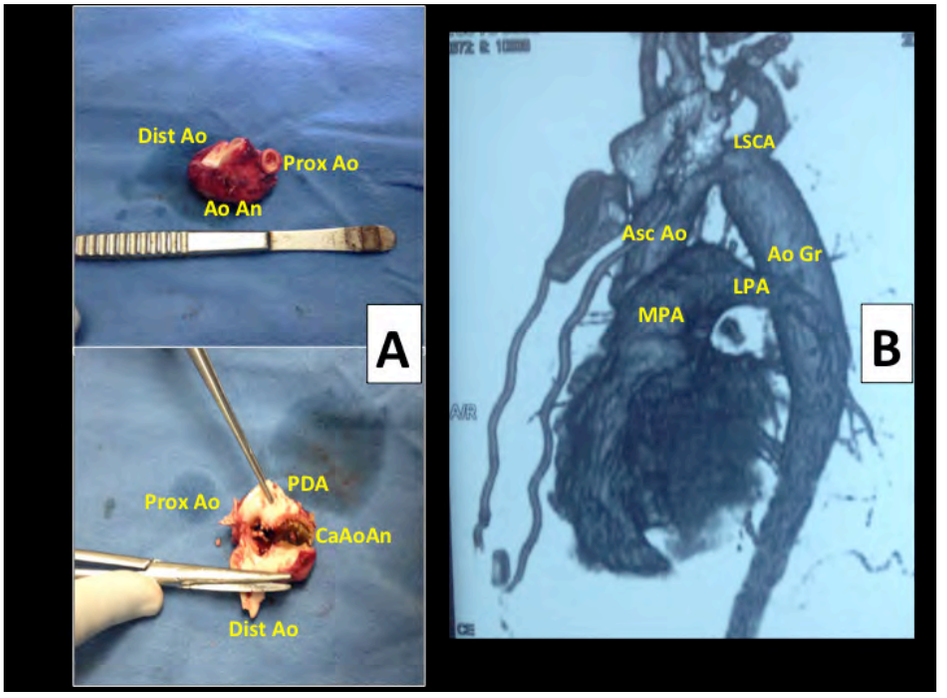

Figure 3. (A) Surgical specimen. Ao An= Aortic aneurysm; CaAoAn= Calcified Aortic Aneurysm Dist Ao= Distal Aorta; Prox Ao= Proximal Aorta. (B) Image of graf tinterposition in the descending aorta. AoGf= Aortic Graft; LPA= Left Pulmonary Artery; LSCA= Left Subclavian Artery; DAo= Descending Aorta.

A spiral CT-scan of the chest was consistent with aortic coarctation. In addition, it revealed a calcified fusiform aneurysm of the ductus arteriosus, whose dimensions were 4.0 x 3.5 cm (**Fig 2**).

With all this above, surgical plan was carried on. A left thoracotomy was performed. The descending aorta was cannulated and the left atrial appendage was used for venous return. Left heart bypass for repair of aortic coarctation was instituted by means of a centrifugal pump. The proximal aorta was clamped between the left carotid and the subclavian arteries. The distal aorta was clamped near the diaphragm and the ductus arteriosus was clamped in the pulmonary end. A transverse aortotomy was performed all along the involved aorta until above and below of coarctation and the ductus arteriosus. Thus, the total aneurysm was removed (**Fig. 3A**). After closing the pulmonary segment of the ductus arteriosus, a 20-mm Dacron graft Hemashield was placed between both aortic ends.

The postoperative course was uneventful, with an adequate control of blood pressure. The patient was discharged at seventh day after operation. The echocardiography at 3 postoperative weeks showed an adequate continuity of thoracic aorta. A mean gradient across the graft of 5 mm Hg was reported, with a peak-to-peak gradient of 12 mm Hg. Left ventricular ejection was 0.50. The CT-scan showed the graft in proper position (**Fig. 3B**). The histopathologic examination of the aneurysm showed thinning of the wall at the expense of breakage and loss of elastic fibers and reduced muscle tissue replaced by a basophilic not fibrillar matrix, and severe calcification.

COMMENT

The association between coarctation of the aorta and aneurysm is well known. The aorta adjacent to the site of maximal narrowing may become aneurysmal as a result of hemodynamic and prolonged effects, aortic dissection, or development of a mycotic aneurysm. The overall prevalence of aneurysm is about 10% by the end of the second decade of life [1].

When an aortic aneurysm is present, resection of the segment of aorta involved, along with the coarctation, is required, and continuity is established with a prosthetic graft [1-3]. Atherosclerotic changes in the coarctated aorta are frequently found in adolescents and adults and may impose technical challenges for the repair. First, the fixed aorta is not suitable for the end to end repair, and the friable vessel wall makes the anastomosis difficult and less hemostatic. Second, the aortic cross-clamping may dislodge debris into the circulation and cause distal embolization [2].

When exist association between coarctation and aneurysm the use of left heart bypass with a centrifuge pump (Biomedicus) is helpful in the management of blood pressure and may reduce the incidence of ischemia damage to the spine [3-4].

It isextremely rare the association of calcified aneurysm and coarctation of the aorta in childhood. Life expectancy may be prolonged by the resection of the aortic aneurysm, and quality of life may be improved by better control of hypertension as well as the recovery of the left ventricle ejection fraction.

Special mention deserves the need of using the left bypass in these cases when paraplegia is a risk. Therefore, it must be thoroughly evaluated in patients with risk factors.

As a conclusion, the relationship between aortic coarctation with aneurysm of the ductus arteriosus and / or descending aorta, has a low incidence in childhood. The usefulness of the left bypass in these rare cases reduces the possibility of spinal and / or organic injury, for this reason in hospitals with low volume of descending aortic surgery, the safety shown of the left bypass must be exploited.

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