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Aorto-Left Ventricular Tunnel, Description of a Case Emphasizing the Differential Diagnosis with other Pathologies

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ABSTRACT

Aorto-Left Ventricular Tunnel (ALVT) is a rare congenital heart disease (CHD) that is worth studying, specifically when considering the differential diagnosis in other pathologies such as coronary artery fistulas (CAF), sub arterial VSD with aortic insufficiency, ductus arteriosus and the like. We present a case of a child who was asymptomatic until the age of 12 and was diagnosed of ALVT during the study of a heart murmur. We found two interesting associations: a unique coronary artery and a sub aortic VSD. After attempting to close the tunnel and VSD, aortic regurgitation was detected, which required reintervention. Authors highlight the importance of the echocardiogram and catheterization to reveal details of the malformation as well as the need for constant follow up after surgery to monitor the possibility of later complications.

KEYWORDS: Aorto – left ventricular tunnel, Coronary artery fistula, Ductus arteriosus, Ventricular septal defect (VSD) with aortic insufficiency, Subarterial VSD

BACKGROUND

ALVT is a rare pathology; until 2018, a set of over 200 cases, with an incidence of 0.03- 0, 4% of CHD, had been published [1]. In spite of its low frequency, in 1988, Hovaguimian put forward a classification [2]. However, even when Edwards had also included this concept in his publication in 1957 [3], the first description of this pathology is attributed to Levy who in 1963 coined it as aortic–left ventricular tunnel [4]. It was found that the diagnosis could be made in the uterus and in postnatal life by means of echocardiography [5,6]. Clinical findings along with the echocardiogram play an outstanding role during the diagnosis [7-10]. In around 90% of patients, the tunnel connects to the left ventricle and, in most cases, the aortic entrance it is located above the sinus of the aortic valve [11,12]. Similarly, as this study presents, the association with coronary artery malformations has been reported as frequent [13], and the majority of them can cause heart failure in the first month of life or in the neonatal period. Notwithstanding, as shown in this study, some patients can be asymptomatic until a later point in their lives, or even until adulthood [11,12]. We consider that although this is a rare disease, it is important to examine it through the differential diagnosis of other pathologies.

DESCRIPTION OF THE CASE

This is the case of a 12 year old boy who was sent to our service by a heart murmur that had been heard when the patient was 6 years old.

Cardiac Evaluation

Asymptomatic patient with normal pulses, mild cardiac hyperactivity with a thrill in the upper portion of the left sternal border, loud second heart sound and continuous murmur IV/VI in the upper portion of the left sternal border. Furthermore, there was a different systolic murmur in the mid-portion of the left sternal border. The systemic pressure was 125/45. The EKG showed biventricular enlargement with predominant on the left ventricle (Figure 1). The Thorax X-Ray showed enlargement of the left ventricle, dilation of the ascending aorta, and increased lung blood flow. A diagnosis of VSD with aortic valve insufficiency was made.

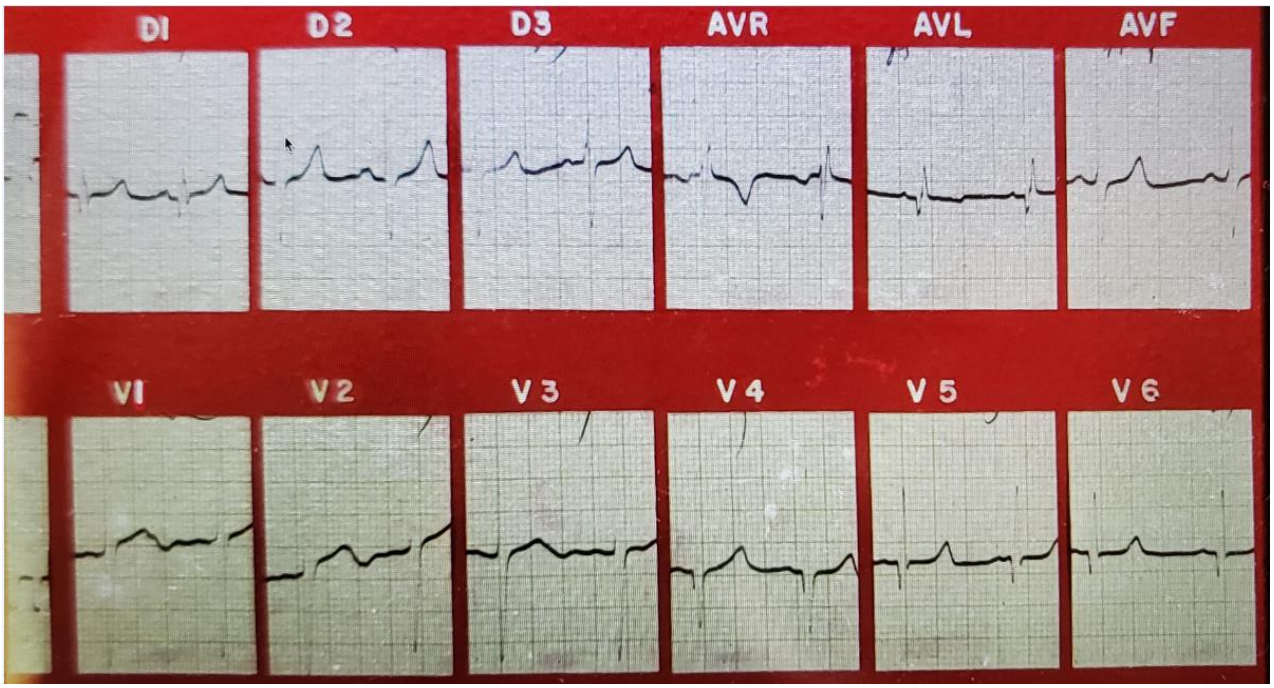

Figure 1: *EKG of the patient that shows right and left ventricular enlargement but mainly of the LV.*

During the echocardiogram, turbulences in the outflow tract of the left ventricle were found and there was a subaortic VSD. It was striking to see that we did not find dilation of coronary arteries (as is common in CAF) and only one coronary artery was visualized.

With the diagnosis of CAF and perimembranous VSD we proceed to catheterize. During the catheterization procedure we found an ALVT (Figure 2A), there was only 1 coronary artery and a subaortic VSD without pulmonary hypertension. The Tunnel was draining into the left ventricle at the upper portion of interventricular septum (Figure 2A). The patient was sent for surgery.

During the surgery, the hole of the tunnel was found in the lateral portion of the sinus of the left coronary valve from which the only coronary artery emerged. The entrance to the tunnel was closed with a patch and the VSD was also closed (Figure 3). After surgery, an important aortic insufficiency was detected, which led to carry out a revision and a subsequent aortoplastia. After this, the recovery of the patient was uneventful.

DISCUSSION

The frequency of ALVT is too low: 0.03 to 0.4 of all CHD. So, it could be considered an exotic CHD that is much more frequent in males than in females [7,11,12]. Despite its low rate of recurrence, this pathology is important in the differential diagnosis with other CHD, mainly the CAFs [14,15], which was the first diagnosis of our case and the subarterial VSD with aortic insufficiency secondary to prolapse of the right coronary valve (PRCV) to the right ventricle through the VSD [16] (Figure 2B).

Figure 2A

Figure 2B

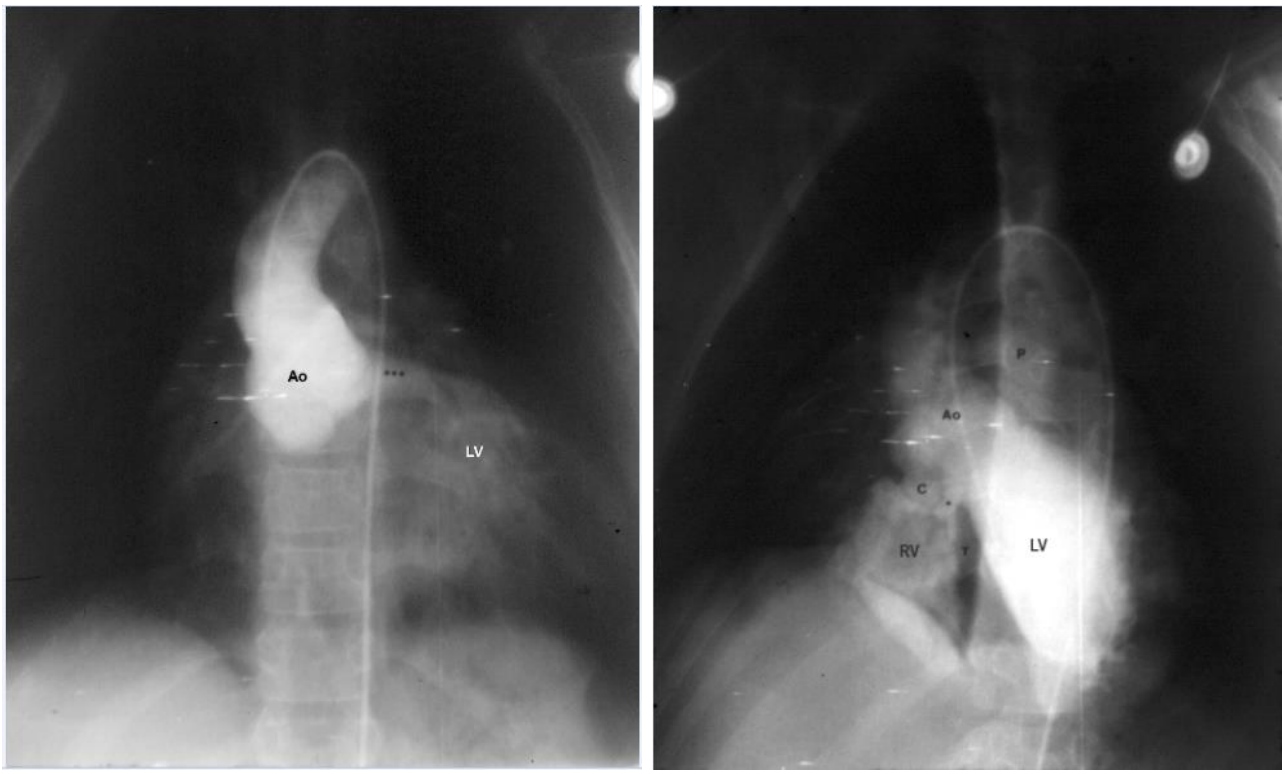

Figure 2. 2A: Aortogram showing the ALVT (Asteriscs), arising from the left coronary sinus and draining into the left ventricle (LV). 2B: Left ventricular angiogram of one of our patients, that shows a subaortic VSD with prolapse of the right coronary valve (c) to the right ventricle through the VSD (asterisc). LV: Left ventricle; RV: Right ventricle; Ao: Aorta; P: Pulmonary trunk. The asterisc shows the subaortic VSD

Considerations on differential diagnosis

Clinical aspects

Although patients with CAF and patients with ALVT can be asymptomatic for a long time, many of them present heart failure during the neonatal period. Conversely, patients with sub arterial VSD and PRCV with aortic regurgitation commonly show the prolapse of the valve and the aortic insufficiency after 5 years of age. In this particular case, it is noteworthy that even though heart murmur was heard at the age of 6, this patient continued asymptomatic and was sent for cardiac evaluation when he was 12. Likewise, other interesting findings concern the origin of the tunnel, since, while in the majority of cases the origin of the tunnel is above the aortic sinus, in our case, the origin is in the same left sinus [17] where the only coronary artery is. As mentioned previously, coronary malformations associated with ALVT are frequent, but in this case, it had a rare association: the subaortic VSD which encumbered the clinical diagnosis [16,18].

In the differential diagnosis, regarding the clinical evaluation, the characteristics of a heart murmur are of utmost importance. In CAFs as well as in ALVT (as in our case) a continuous murmur can be heard. Nevertheless, in ALVT the diastolic component is more noticeable. In relation to the diastolic murmur, which can be notorious in ALVT, it is important to highlight the difference with the diastolic murmur of the sub aortic VSD with aortic regurgitation. In this pathology the diastolic murmur always decreasing. Initially it is very soft and increase progressively with the time. Finally, concerning the continuous murmur, we need to consider the differential diagnosis with the murmur of ductus arteriosus. In this pathology although the murmur is continuous, the diastolic component is short, and there is a reinforcement at the end of the systole (Gibson´s murmur) [19].

In the differential diagnosis the echocardiogram and the catheterism play a vital role [8-11]. Figure 2 shows two cases: Figure 2A shows the angiogram corresponding to our case of ALVT, but, in another projection, the subaortic VSD was clearly demonstrated and Figure 2B that shows the angiogram of one of our patients with sub aortic VSD with prolapse of the right coronary valve and aortic regurgitation; anyway, it is crucial to acknowledge the echocardiogram as the most important tool in diagnosis including prenatal diagnosis [5,6,8-12]. The prenatal diagnosis is of great value because allows to plan the management of the patient after delivery.

Moreover, several publications pinpoint continuous postoperative care as critical because of aortic insufficiency, progressive dilation of the ascending aorta or a reopening of the tunnel could appear [20-22].
Finally, another possibility of treatment different from surgery is the Trans catheter closure with amplatzer device [23].

Figure 3A

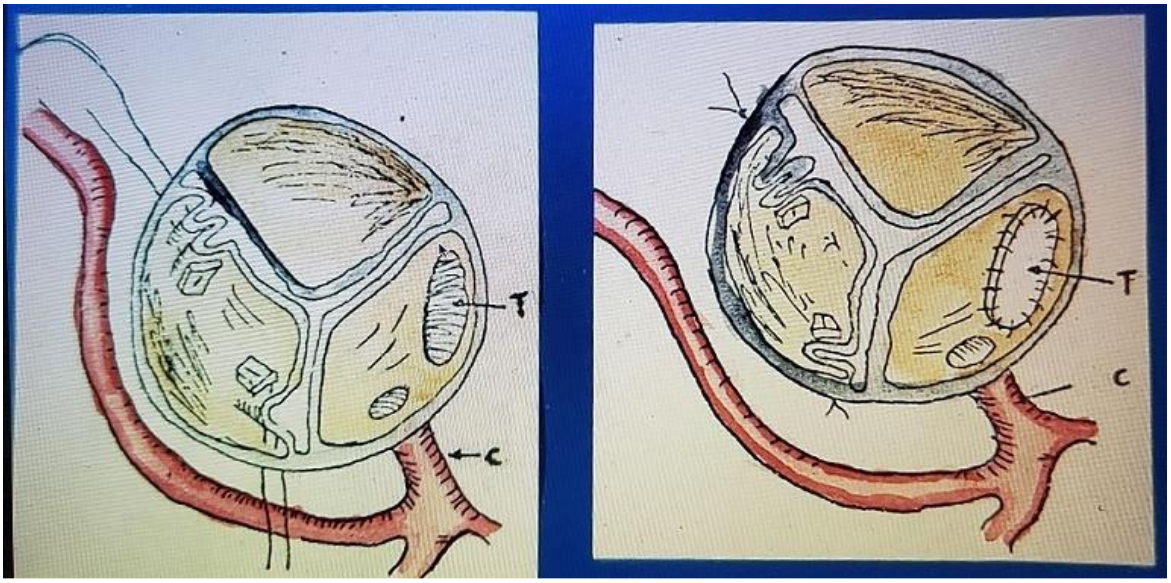

Figure 3B

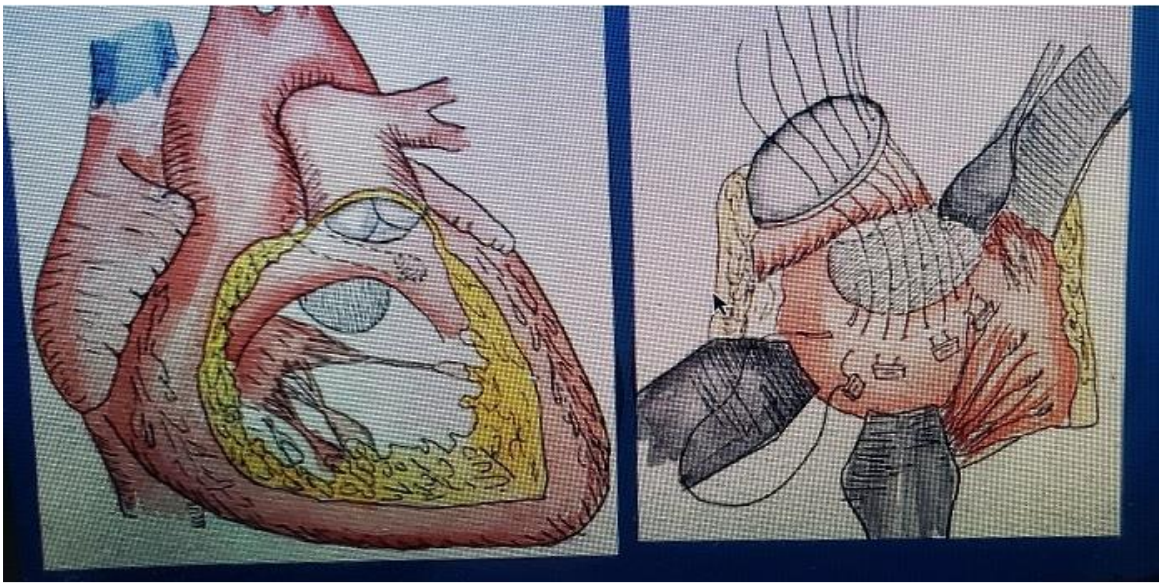

Figure 3: *Drawings illustrating the surgical treatment: 3A: The figure illustrates the closure of the aortic entrance of LAVT with a patch (arrow). 3B: The figure illustrates the closure of the VSD with a patch, T: Tunnel entrance; c: Coronary artery*

CONCLUSION

- Although ALVT is a rare pathology; considering the differential diagnosis of other pathologies becomes imperative
- Our case has two interesting associations: The malformation of the coronary arteries as only one coronary artery was found, which is a frequent association, and the subaortic VSD that is a rare association
- The echocardiogram and the catheterization are very important tools in diagnosis of ALVT
- After surgery, patients need continued follow-up due to the frequency of later complications.

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