Case Report

Awareness of a Rare Diagnosis: CIA of the Coronary Sinus (A Case Report)

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Abstract:

Interatrial communication is a common congenital heart disease in adulthood. It is subdivided into four distinct groups based on location.

Coronary sinus type is the rarest one and represents less than 1% of interatrial septal defects. Its echocardiographic diagnosis can be difficult.

We report the case of a 27-year-old female patient, admitted to the emergency department for management of a poorly tolerated supraventricular tachycardia, referred to cardiology after cardioversion.

Clinical examination revealed a SaO2 of 95%, a B2 burst at the pulmonary site. The ECG in sinus rhythm showed right bundle branch block and right atrial hypertrophy.

Echocardiography showed dilated right chambers, coronary sinus atrial septal defect with a left-to-right shunt, dysplastic mitral valve, and pulmonary pression was estimated at 85mmhg on tricuspid insufficiency flow; right ventricular function was normal.

The patient was treated with B-blocker and VKA with a good clinical course.

She is waiting a right catheterization to decide on surgical closure of the atrial septal defect.

Through this case, we wanted to underline the importance of not omitting the echocardiographic search for a defect of the roof of the interatrial septum in front of a volumetric overload of the right cavities.

Introduction

Atrial septal defect is the second most common adult congenital heart disease after aortic bicuspidia [1]. It is more prevalent in women and can be found in the setting of fetopathy in ethyllic mothers [2]. Coronary sinus atrial septal defect is an uncommon location and derives from a roof defect between the coronary sinus and the left atrium secondary to hypoplasia of the left atrioventricular fold during embryonic development [3]. It may also be associated with a syndromic grouping such as Holt Oram syndrome (Atrial septal defect, thumb anomalies and auriculo-ventricular block) or Poland syndrome (unilateral pectoral agenesis and Atrial septal defect). Some familial forms have been described in affiliation with conductive disorders [4]. The Atrial septal defect is responsible for a left-right shunt at the atrial stage depending on the pressure regime at the level of the atria and causes an overload of the right cavities. The management of the defect by percutaneous or surgical closure depends on the impact on the right cavities and the associated structural abnormalities. Through the description of this case, we have emphasized the awareness of the existence of the atrial septal defect form of the coronary sinus. The diagnostic approach being difficult, a careful evaluation of the coronary sinus roof should be considered in front of a dilatation of the right cavities.

Observation:

This is a 27-year-old female patient with no modifiable cardiovascular risk factors from a consanguineous marriage. She was born at term by an uncomplicated childbirth. In her family history, we did not find any congenital heart disease or conductive disorder. The family reports the notion of chronic bronchopneumopathy during childhood.

The patient did not report any exertional limitation or episodes of cyanosis during childhood. She consulted the emergency room for palpitations with an abrupt onset, permanent, associated with stage IV dyspnea.

The clinical examination in the emergency room showed an unstable hemodynamic state with a BP= 70/50mmhg and signs of low peripheral flow.

The ECG performed in the emergency room revealed a regular wide QRS tachycardia with a CVM of 15Ocpm suggestive of supraventricular tachycardia with conduction aberration. Electrical cardioversion was indicated in view of the clinical picture.
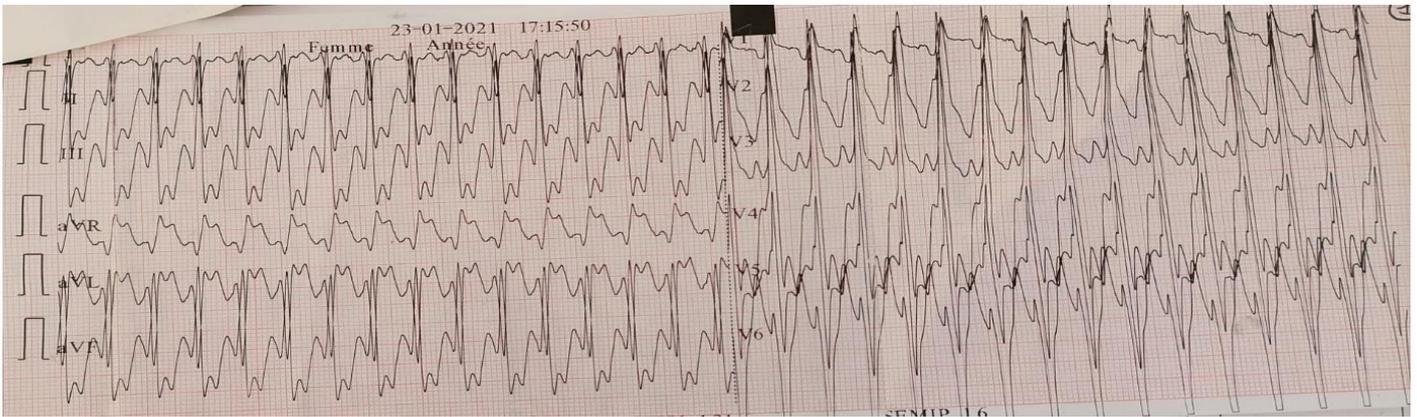
Figure 1: ECG with tachycardia

The patient was then transferred to the cardiac intensive care unit.
The examination showed :

- improved hemodynamic status with BP= 90mmhg HR=90cpm
- Height= 142cm, P=40kg, no skeletal abnormalities
- Sao2= 95%, positive Harzer sign
- Pulmonary B2 burst
- No congestive signs

ECG in sinus rhythm showed: regular sinus rhythm, right axial deviation, constant PR at 160ms, right bundle branch block.
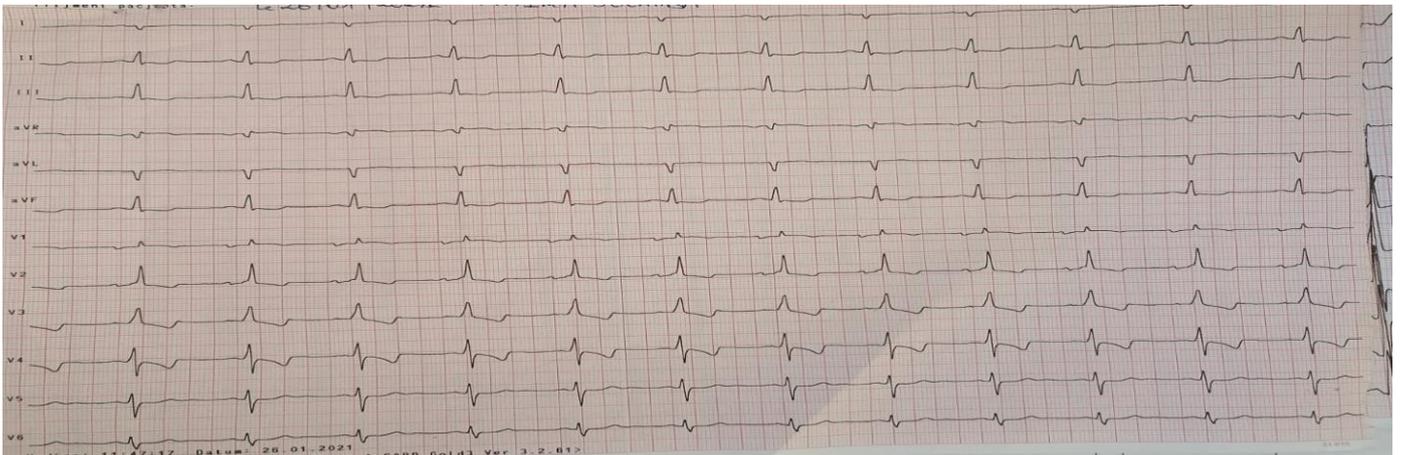

Figure 2: sinus rhythm EKG

Chest X-ray showed cardiomegaly with an ICT of 0.7, straightness of the left middle arch, supra-diaphragmatic cardiac tip, and a right overhang.

figure 3: Chest X-Ray

Echocardiography objectified:

- Situs solitus, levocardia
- Dilation of the right cavities pushing back the left cavities
- Dysplastic mitral valve with moderate leakage
- AIC of the wide coronary sinus visualized at the subcostal level with a left-right shunt
- Dilated pulmonary artery trunk with : pulmonary Vmax=1.63m/s, Pulmonary pression= 70mmhg
- Small aorta at 16mm
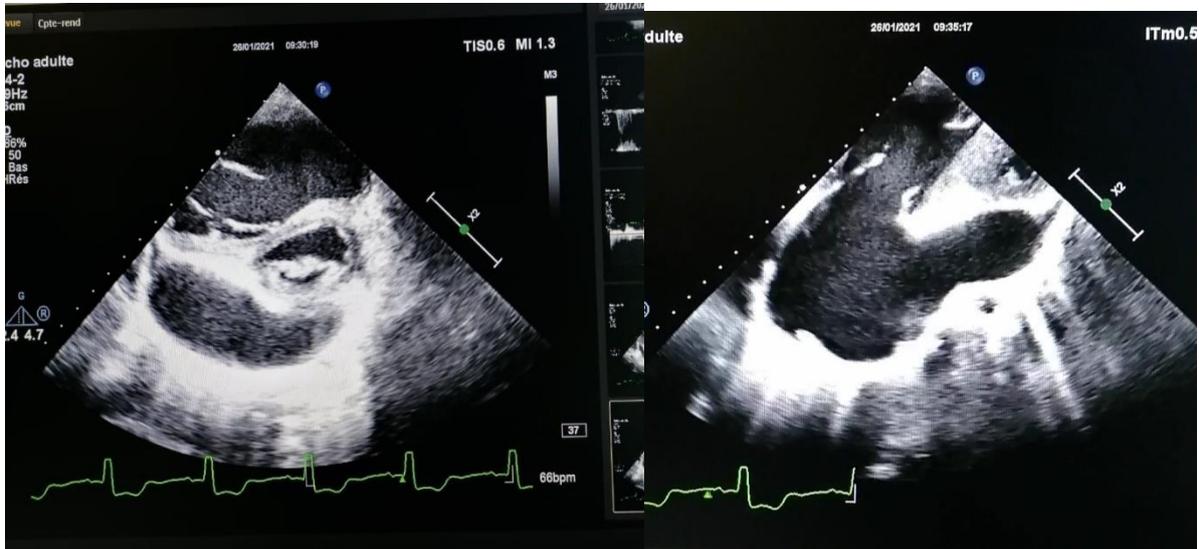- No venous return abnormality
- Dilated IVC

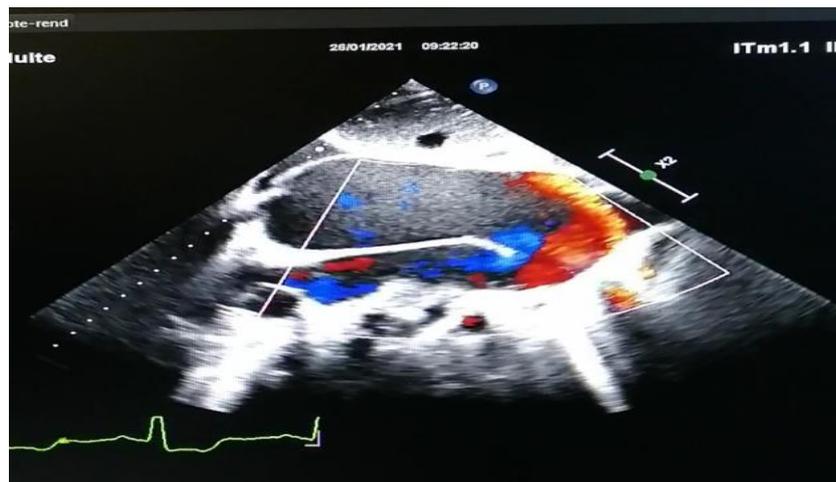**Figure 4: dilatation of the right cavities and the coronary sinus**

Figure 5: coronary sinus roof defect

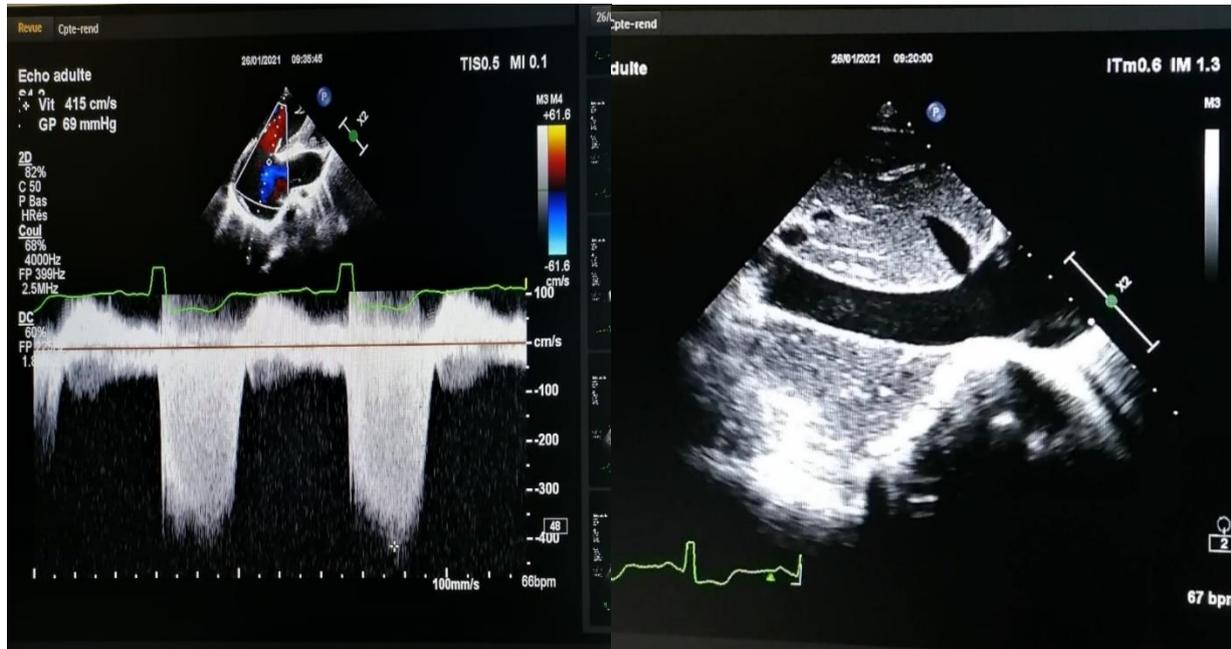
Figure 6: mitral valve dysplasia

Figure 7: Pulmonary pression and dilated inferior vena cava

The patient was put on antiarrhythmic treatment with cordarone, a low dose of b-blocker and anticoagulation with a VKA as the drug of choice.

The evolution was favorable with maintenance of sinus rhythm. The resting heart rate was 80cpm.

The patient is awaiting a right heart catheterization before surgical closure of the defect.

Discussion

Coronary sinus ASD (atrial septal defect) is a rare form of dehiscence of the interatrial septum. The non-specific clinical presentation and the low-noise evolution like the other localizations of ASD make it a pathology whose diagnosis remains essential.

The particular anatomical situation of the coronary sinus is largely responsible for the difficulty in diagnosing this form of ASD.

Apart from rare neonatal clinical forms where the shunt can be immediately right-left, the other types are revealed during childhood or in adulthood.

It is evoked at school age when a murmur is discovered by chance or when there is a history of chronic bronchopneumopathy without obvious etiology [5].

The delay in diagnosis is due to the absence of specific functional signs and a clinical tolerance compatible even with high level sports activity [6].

The late age of discovery and the absence of screening by clinical or echocardiographic examination in our case can be explained by the evolution of the pathology and by the fact that our patient lives in a rural area with limited access to care.

The volumetric overload due to the ASD generates a dilatation of the right atrium. The latter is responsible for structural and electrical remodeling that constitutes the bed of supraventricular arrhythmia [7].

Our patient was admitted for management of a poorly tolerated tachy-atrial requiring electrical cardioversion.

The incidence of these arrhythmias, in this case AF, is increased beyond 60 years of age and is estimated at 10% for non-operated patients under 40 years [8]. Some cases of isthmo-dependent right atrial flutter have been described.

This predisposition to supraventricular arrhythmias in patients with an ASD is also demonstrated by the increased occurrence of AF after surgical closure of the defect [9]. The prevalence of arrhythmia attacks in ASD is ≤ 2% in patients aged 20 years; 10-15% in subjects aged 40 years and estimated between 20-40% beyond 60 years [10].

Some studies suggest that this incidence is higher in men; with COPD; with right ventricular dysfunction and pulmonary hypertension [11].

Closure of the defect generates a decrease in mortality related to these rhythm disorders due to reverse structural remodeling [12].

Conductive disturbances are rare in patients with a ASD [13]. Sinus dysfunction has been described in subjects over 60 years of age after sinus venosus ASD repair due to the proximity of the sinus node during defect repair [13,14]. Some new techniques such as the Warden procedure have limited the incidence of sinus dysfunction secondary to closure [15].

The 1st degree atrioventricular block can also be seen in ASDs. They have been related to interatrial block by increasing the conduction time between the atria [16].

Major atrioventricular conductive disturbances have been reported in ostium primum type defects. These abnormalities may develop due to the proximity of the conduction pathways to the septal defect [17].

Some surgical procedures for closure of ostium primum septal defect may result in high-grade AVB due to damage the conductive tissue by apposition of the closure patch [18]. Factors that condition the occurrence of postoperative conductive disorder are young age; width of the defect and size of the closure patch [19].

Genetic conductive disorders have been described in association with ASD [20]. Genetic mutations have been linked to these atrioventricular blocks such as TBX5 associated with Holt Oram syndrome [20,21].

We observed a right intraventricular conductive disorder in our patient, probably related to the dilation of the right ventricle.

The lack of specificity of suggestive clinical signs gives transthoracic echocardiography a major diagnostic role. It also allows the search for associated structural abnormalities and the assessment of their impact.

Some localizations such as coronary sinus roof defects may be difficult to detect for an untrained practitioner.

Volume overload of the right cavities may be a suggestive sign. The bicaval subcostal window allowed us to diagnose the coronary sinus roof defect.

The search for venous return abnormalities, the most common in this case is the persistence of the left superior vena cava, should be eliminated [6]. Dilatation of the coronary sinus should raise suspicion of this anomaly, which gives a different twist to surgical management [22].

Our patient did not present any venous return abnormalities, especially in the suprasternal window, and this was confirmed by right catheterization.

Contrast ultrasonography has also been described as a means of diagnosing the persistence of the left superior vena cava [23].

The indication for surgical closure must be made at the appropriate time. Attention should be paid to the hemodynamic impact and the associated structural anomalies [24].

In our patient, a mitral valve dysplasia responsible for moderate regurgitation was identified.

The association of mitral damage and ASD of the coronary sinus is rarely described in the literature. In Sugimori's study, all five patients reported had mitral insufficiency concomitant with the interatrial septal defect [25].

Two lesional mechanisms of mitral regurgitation were found in this same series including prolapse of the anterior leaflet of the valve in one patient and dilatation of the annulus in the remaining patients.

Systolic pulmonary pressure on tricuspid insufficiency flow was estimated at 85 mmhg when POD was added.

Some involvement of the post-capillary was suggested in the elevation of pulmonary pression in our patient.

A study of pulmonary and systemic pressures and flows by right catheterization was proposed for our case before the indication of closure.

As pointed out in case studies reported in the literature regarding ASD closure: it appears that a Qp/Qs $\geq$ 1.5, a PVR level $\leq$ 3uW and a positive reversibility test could be contributory to a satisfactory operative outcome [25,26].

The effect of ASD closure on pulmonary pressure is sometimes difficult to predict. However, data highlight that the long-term survival of patients is similar to that of the general population (figure 8) when the defect is closed before the age of 25 years [27,28].
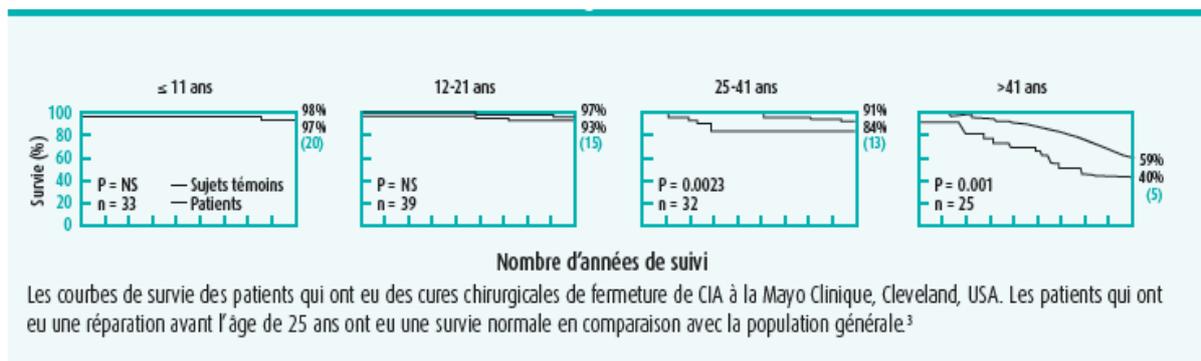

Figure 8: Survival curves of patients who underwent surgical treatment for ASD at the mayo clinic

Conclusion

Interatrial septal defects are a congenital heart disease sometimes discovered in young adults.

The late diagnosis makes it a pathology for which screening remains essential. The particular location of the ASD of the coronary sinus is important to evoke in front of a volume overload of the right cavities and a dilatation of the coronary sinus. The indication for closure will be based on the analysis of the hemodynamic repercussions, largely assisted by right catheterization.

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