SAS Journal of Medicine
Abbreviated Key Title: SAS J Med
ISSN 2454-5112
Journal homepage: https://saspublishers.com

OPEN ACCESS

Medicine and Pharmacy

Congenital Coronary Ectasia and Bicuspid Aortic Valve: A Case Report with Review of the Literature

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DOI: 10.36347/sasjm.2022.v08i04.014 | **Received:** 07.03.2022 | **Accepted:** 13.04.2022 | **Published:** 21.04.2022

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Abstract **Case Report**

Coronary ectasia is a rare clinical condition whose etiology is dominated by atherosclerosis. In a small percentage of patients, it may be of congenital origin. Its mechanisms are still poorly understood and its management is still non standardized due to its rarity. We report a case of congenital coronary ectasia discovered in a patient with aortic valve stenosis caused by congenital bicuspid aortic valve; this association being frequent in congenital coronary ectasia. The aim of this report is also to review the clinical features of coronary ectasia, its diagnosis and treatment.

Keywords: Congenital, Bicuspid Aortic, Literature.

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INTRODUCTION

Coronary ectasia is a rare condition [1, 2], and its congenital origin is even less common. It is usually asymptomatic but can be revealed by an acute or chronic coronary syndrome. Its pathophysiology is still incomplete, its diagnosis is relatively simple and relies on coronary angiography, but its management is a major challenge as there are no consensual therapeutic approaches due to its rarity which impedes large randomised trials.

We report a case of congenital coronary ectasia discovered incidentally during the preoperative work-up of symptomatic aortic stenosis (stress angina) caused by bicuspid aortic valve. This case report is intended as a didactic opportunity to review the literature on the complex management of coronary ectasia.

CASE PRESENTATION

We report the case of a 53-year-old man with a history of primary biliary cholangitis treated with ursodeoxycholic acid for 2 years, polyclonal hypergammaglobulinemia treated with immunosuppressants and cardiovascular risk factors such as android obesity, type 2diabetes and recent hypercholesterolemia.

He has been followed since 2005 for bicuspid aortic valve of incidental discovery and progressive worsening. The patient was asymptomatic until one month before admission to hospital when he presented with CCS class II stress angina without other associated signs.

Cardiovascular examination revealed a harsh systolic murmur at the aortic focus radiating to the neck vessels indicating aortic valve stenosis. Peripheral pulses were present and symmetrical without murmurs along the vascular axes. The rest of the somatic examination was unremarkable.

The ECG showed a regular sinus rhythm with HBAG without repolarisation disorders, and the chest X-ray showed signs of pulmonary venous hyperpressure.

Transthoracic echocardiography confirmed the presence of a tight aortic valve stenosis on bicuspid aortic valve (Figure 1A and 1B)(Vmax: 4.3 m/s, mean Gd: 47 mmHg) with preserved LVEF at 53%. The left ventricle was undilated with moderate hypertrophy. The ascending aorta was of normal calibre.

As part of the preoperative workup for congenital valve disease, coronary angiography revealed ectatic coronary arteries with significant first diagonal stenosis and an intermediate lesion of the distal right coronary artery (Figure 2A, 2B).

The decision of the medico-surgical staff in relation to this coronary ectasia associated with congenital bicuspid valve was to replace the valve with a mechanical prosthesis. The minimal extension work-up (ETSA and EDMI) was normal.

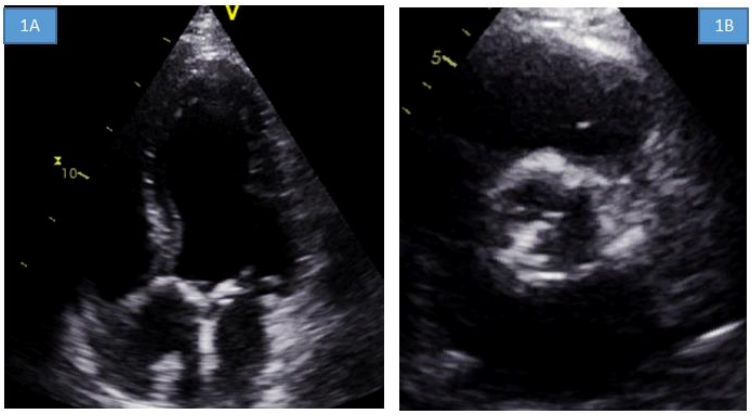

Fig-1A,B: bicuspid aortic valve 5 cavities and small axis cut

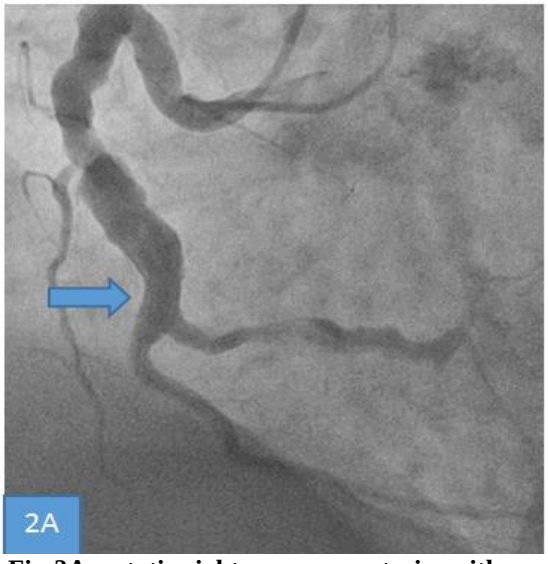

Fig-2A: ectatic right coronary arteries with an intermediate distal lesion

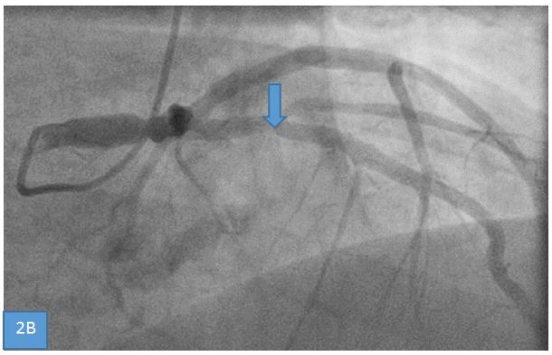

Fig-2B: ectatic coronary arteries with significant first diagonal stenosis

DISCUSSION

Coronary ectasia is the dilation of a coronary segments to a diameter at least 1.5 times that of the adjacent normal coronary artery and a length of dilation exceeding more than one third of the length of the normal coronary artery [3]. It may be either diffuses, affecting the entire length of a coronary artery, or localised. It may involve one or more coronary arteries and develop on healthy or pathological coronary arteries. Classically, four types are distinguished [4]:

- Type 1: diffuse ectasia of two or three vessels
- Type 2: diffuse ectasia in one vessel and localised disease in another vessel
- Type 3: diffuse ectasia in a single vessel
- Type 4: localized or segmental disease

This rare condition is found in 3-8% of coronary angiograms and in 0.22-1.4% of autopsy series. In the CASS registry, only 4.9% of coronary artery ectasia was found, mainly in the right coronary artery (68%) and in the proximal part of the anterior interventricular artery (60%) [5]. It is attributed to atherosclerosis in over 50% of cases. Other acquired etiologies of coronary ectasia include Kawasaki disease, which is very common in children, mycotic or septic emboli, Marfan's syndrome, arteritis due to polyarteritis nodosa, Takayasu's disease or systemic lupus erythematosus [6].

In our case, a coronary ectasia was discovered incidentally during the preoperative work-up of a patient with symptomatic narrowing of the aorta due to bicuspidism. As bicuspidism is a congenital malformation of the aortic valve, its association with diffuse coronary ectasia strongly suggests a congenital origin of the coronary ectasia. Furthermore, when congenital coronary ectasia is present it is frequently associated with aortic bicuspidity.

The pathophysiology of congenital coronary ectasia is still a mystery. Atherosclerosis usually leads to a decrease in the lumen of the vessel; however, there is evidence of remodelling of the medial and external limiting membrane of the arterial wall in both directions. Expansion of this membrane may explain the coronary dilation in atherosclerosis. Expansive

remodelling is facilitated by enzymatic degradation of the tunica media and associated chronic inflammation [7]. Clinically, most patients are asymptomatic. However, the mode of revelation may be an acute coronary syndrome.

However, the mode of onset can be an acute coronary syndrome or a chronic coronary syndrome as in our patient. Indeed, coronary dilatation leads to a decrease in blood flow velocity favouring stasis and the formation of intracoronary thrombi that can embolise distally [8, 9]. Coronary angiography coupled with intravascular ultrasound is the gold standard in the evaluation of coronary ectasia. They allow the diagnosis and characterisation of anomalies [10].

There is still no consensus on the management of coronary ectasia due to the rarity of cases. The treatment approach is individualised taking into account the patient's clinical and anatomical risks. When coronary ectasia occurs in pathological arteries (atherosclerosis), anti-ischaemic medical treatment including control of cardiovascular risk factors is indicated. Surgery may be proposed when optimal medical treatment fails. Thrombolysis, administration of anticoagulants (heparin, glycoprotein IIb/IIIa receptor blockers) is often beneficial in acute coronary syndromes associated with coronary ectasia. Primary angioplasty is also an alternative provided that a good calibre stent is used to avoid stent embolisation. Many authors recommend chronic anticoagulation; however, there are no randomised trials showing its benefit in coronary ectasia. The expected benefit must outweigh the risk of bleeding [11, 12].

Patients with coronary ectasia with recurrent complications are also candidates for surgery. This involves ligation of the proximal and distal segments of the ectatic vessel and replacement with a bypass graft. No attempt to repair the ectatic vessel is recommended as the results are poor.

In our case, the treatment of coronary ectasia was respected but the patient benefited from an aorto-coronary mono-bridging in combination with aortic valve replacement with a prothesis.

CONCLUSION

Congenital coronary ectasia is a rare condition with poorly understood mechanisms and complex management. This case report reminds us of the unmet need for randomised trial data to better understand and manage this condition.

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