

Multiple diffuse coronary cameral fistulas from the left anterior descending artery and right coronary artery to both the right and left ventricle associated with left ventricular noncompaction: A rare combination

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ABSTRACT

Diffuse and multiple coronary cameral fistulas are very rare and with very few case reports of its association with left ventricular noncompaction are published. Here, we report a 6-year-old child of multiple diffuse coronary cameral fistulas to both the right and left ventricle in association with the left ventricular noncompaction. A possible common embryological link between the two uncommon entities is also discussed.

Keywords: Multiple diffuse coronary cameral fistulas, Multiple coronary cameral fistulas, LV non-compaction

INTRODUCTION

Congenital coronary cameral fistula is a rare cardiovascular malformation, being present in 0.002% of the general population.^[1] Multiple fistulas are present in 10.7%–16% of all coronary cameral fistulas.^[2] Left ventricular noncompaction is a myocardial disorder thought to be caused by the developmental arrest of the endocardium during the embryological period. The combination of multiple coronary artery to ventricle fistulas and left ventricular noncompaction has been rarely reported in adults^[3,4] and even more rarely in children.^[5] Here, we report a rare case, in which both the right and left coronary artery was communicating with multiple fistulas to the right and left ventricles in association with the left ventricular noncompaction. We believe that this combination has not been reported. A possible common embryological link is discussed.

CASE REPORT

A 6-year-old male child was referred for the evaluation of cardiac murmur noted on routine pediatric outpatient clinic visits. Physical examination revealed no obvious syndromic features. The heart rate was 80/min with normal heart sounds with soft early diastolic murmur heard best at the apex. Electrocardiogram was normal. Echocardiography showed a dilated and ectatic left main coronary artery and left anterior descending artery, which was draining diffusely into the left ventricular cavity. The echocardiography also showed the left ventricular noncompaction. The compacted-to-noncompacted ratio in diastole was 2:1 [Videos 1 and 2]. The left ventricular function was normal. Cardiac computed tomography (CT) was done, which confirmed the echocardiography findings. Left anterior descending, circumflex and distal

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10.4103/apc.APC_37_19

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How to cite this article: Srivastava R, Azad S, Radhakrishnan S. Multiple diffuse coronary cameral fistulas from the left anterior descending artery and right coronary artery to both the right and left ventricle associated with left ventricular noncompaction: A rare combination. *Ann Pediatr Card* 2020;13:171-3.

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Submitted: 12-Mar-2019

Revised: 08-Oct-2019

Accepted: 18-Dec-2019

Published: 12-Feb-2020

right coronary artery was diffusely dilated [Figure 1a and b, respectively]. The CT showed multiple fistulous communications between the left ventricular cavity and the left anterior descending and right coronary artery. The left ventricular noncompaction was also evident in the CT [Figure 2].

Coronary angiography was done to further delineate the anatomy of this rare anomaly. The selective right coronary angiogram showed multiple diffuse coronary cameral fistulae from the right coronary artery to the right ventricle and posterior descending artery to the left ventricle [Figure 3a]. Selective left coronary angiogram showed diffusely dilated left main coronary artery/left anterior descending with multiple diffuse coronary cameral fistulae from the left anterior descending to the left ventricle [Figure 3b]. The left ventricular angiogram showed noncompaction of the left ventricle [Figure 3c].

Stress thallium test was negative for inducible ischemia. No active intervention was, therefore, indicated. The child was started on aspirin in view of the left ventricular noncompaction and placed on regular follow-up.

DISCUSSION

Congenital coronary cameral fistulas were first described by Krause in 1895. Isolated fistulas are far more commonly occurring in 90% of the cases as compared to multiple fistulae, which are present in 10.7%–16% of all congenital coronary cameral fistulae.^[2] Left ventricular noncompaction cardiomyopathy is rare cardiomyopathy characterized by increase in the noncompacted, trabeculated myocardium adjacent to compacted myocardium in the left ventricle.^[6]

The association of multiple diffuse coronary cameral fistulae with the left ventricular noncompaction is intriguing, as both of them are embryologically very closely linked. Studies suggest that myocytes project radially into the cavity and are covered by the endocardial layer. This array guarantees the best perfusion of the myocytes by increasing the contact surface between the left ventricular cavity

and the myocytes, while the coronary tree is not yet developed. Intertrabecular spaces are transformed into capillary vessels. Failure at this stage corresponds to the formation of thin elongated trabecular projections separated by deep recesses.^[7] It has been suggested that coronary artery to ventricular fistula is caused by transforming failure of sinusoidal connection to capillary and is a distinct manifestation of noncompacted myocardium.^[5] Another report provides data that indicate the ventricular coronary capillary bed forms perinatally by endocardial trapping during ventricular wall compaction.^[8] A halt at this stage of development may cause multiple diffused coronary cameral fistulae and left ventricular noncompaction. Some recent studies have suggested that ventricular noncompaction may not necessarily mean failure of compaction, and noncompaction may not be the causative factor for dysfunction or cardiomyopathy.^[9]

The combination of multiple congenital coronary artery to ventricular fistulae and left ventricular noncompaction has been rarely reported.^[3] The authors suggest that these two diseases were caused by the developmental arrest of the endocardium somewhere in between the regression of the myocardial sinusoids and the compaction of the myocardium and they may be the continuum of one disease. This combination is even more rarely reported in children. The only case we found was a 6-month old child presented with features of congestive heart failure and diagnosed with a combination of the left ventricular noncompaction and multiple diffuse coronary artery to the right ventricle fistulae.^[5]

This combination can present with varied clinical manifestations depending on the severity of each lesion. It can present as cardiac insufficiency and arrhythmia as described in patients with noncompaction cardiomyopathy. With a combination of coronary artery fistulae, the left ventricle dysfunction can become severe

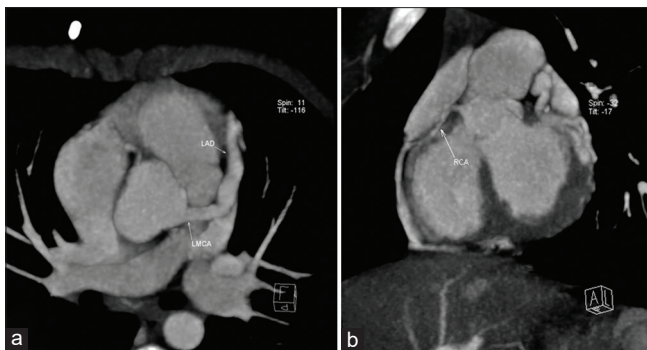


Figure 1: Computed tomography images showing (a) diffusely dilated left anterior descending, circumflex artery. (b) Dilated distal right coronary artery (shown by white arrow)

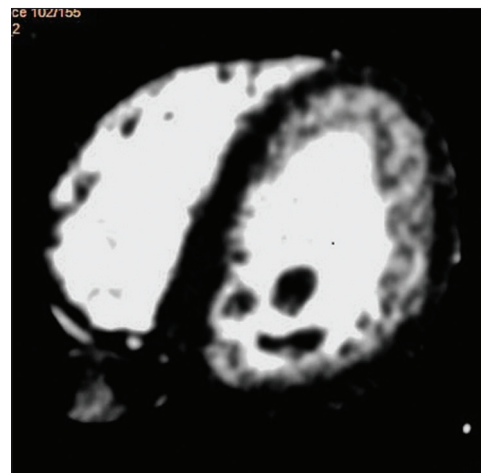


Figure 2: Computed tomography image of the ventricles in short axis showing the left ventricular noncompaction

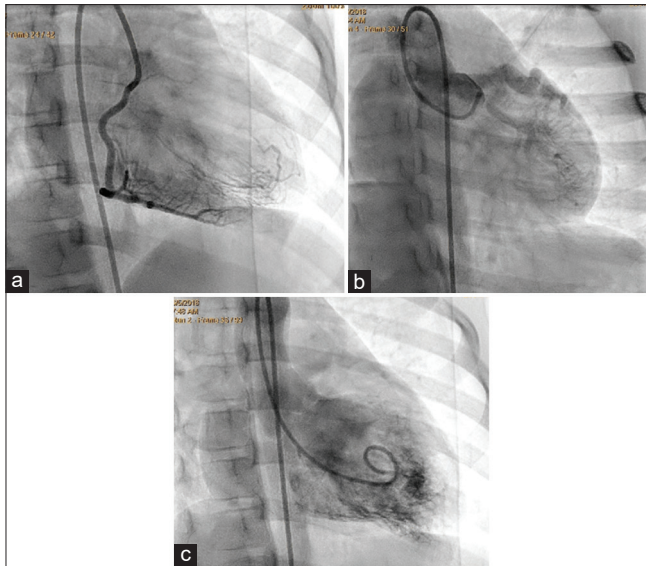


Figure 3: (a) Selective right coronary angiogram showing multiple diffuse coronary cameral fistulae from the right coronary artery to the right ventricle. (b) Selective left coronary angiogram diffusely dilated left main coronary artery and left anterior descending artery with multiple diffuse coronary cameral fistulae to the left ventricle, also note the spongy myocardium evident in the angiogram. (c) Left ventricular angiogram showing features of noncompaction: thin compacted layer and deep intertrabecular recesses

with associated myocardial ischemia due to steal from coronary cameral fistula.

The diagnosis is usually established using transthoracic echocardiography and coronary angiography. The criterion for the diagnosis of noncompaction has been summarized by Jenni *et al.*^[10] Since our patient was asymptomatic and had no features of congestive heart failure or ischemia, we did stress thallium to look for any evidence of inducible ischemia. The test turned out to be negative.

CONCLUSION

The combination of multiple coronary artery to the left and right ventricular fistulas along with left ventricular noncompaction is a very rarely seen congenital heart malformation. The presentation can vary depending on the severity of the individual lesions. The diagnosis is usually established by echocardiography and coronary angiography. The possibility of genetic etiology needs to be evaluated further.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the

patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

- Ogden JA. Congenital anomalies of the coronary arteries. *Am J Cardiol* 1970;25:474-9.
- Challoumas D, Pericleous A, Dimitrakaki IA, Danelatos C, Dimitrakakis G. Coronary arteriovenous fistulae: A review. *Int J Angiol* 2014;23:1-10.
- Dias V, Cabral S, Vieira M, Sá I, Anjo D, Gomes C, *et al.* Noncompaction cardiomyopathy and multiple coronary arterioventricular fistulae: 1 or 2 distinct disease entities? *J Am Coll Cardiol* 2011;57:e377.
- Wilhelm J, Heinroth K, Stoevesandt D, Werdan K, Plehn A. Non-compaction cardiomyopathy with diffuse left coronary artery fistulae as a rare cause of congestive heart failure. *Eur Heart J* 2013;34:12.
- Chen Y, Zhang Z, Li F, Fu L, Wu J, Zhang Y, *et al.* A case of non-compaction ventricular myocardium and multiple coronary artery-to-right ventricle fistulae. *Int J Cardiol* 2015;184:659-63.
- Jenni R, Goebel N, Tartini R, Schneider J, Arbenz U, Oelz O. Persisting myocardial sinusoids of both ventricles as an isolated anomaly: Echocardiographic, angiographic, and pathologic anatomical findings. *Cardiovasc Intervent Radiol* 1986;9:127-31.
- Sedmera D, Thompson RP. Myocyte proliferation in the developing heart. *Dev Dyn* 2011;240:1322-34.
- Tian X, Hu T, Zhang H, He L, Huang X, Liu Q, *et al.* Vessel formation. De novo formation of a distinct coronary vascular population in neonatal heart. *Science* 2014;345:90-4.
- Anderson RH, Jensen B, Mohun TJ, Petersen SE, Aung N, Zemrak F, *et al.* Key questions relating to left ventricular noncompaction cardiomyopathy: Is the emperor still wearing any clothes? *Can J Cardiol* 2017;33:747-57.
- Jenni R, Oechslin E, Schneider J, Attenhofer Jost C, Kaufmann PA. Echocardiographic and pathoanatomical characteristics of isolated left ventricular non-compaction: A step towards classification as a distinct cardiomyopathy. *Heart* 2001;86:666-71.