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Case Report

Isolated Right Ventricular Non-compaction in a Neonate Born to a Mother with Gestational Diabetes: A Case Report

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ABSTRACT

Background: Ventricular non-compaction, which is a rare congenital cardiomyopathy, results from an arrest in normal endomyocardial embryogenesis. It is characterized by a pattern of prominent trabecular meshwork and deep inter-trabecular recesses. The clinical manifestations include systolic and diastolic dysfunctions, heart failure, ventricular arrhythmias, and cardioembolic events.

Case report: Here we present the case of a neonate (38 weeks, weighing 2580 gr) born to a mother with gestational diabetes (GDM) with isolated right ventricular non-compaction (IRVNC) and systolic and diastolic right heart dysfunction due to meconium aspiration syndrome and tension pneumothorax. A 4-month follow-up demonstrated a complete improvement in the newborn.

Conclusion: It is of paramount importance to consider RVNC cardiomyopathy a differential diagnosis of cyanosis and respiratory distress due to meconium aspiration syndrome in neonate s born to mothers with GDM. Apart from RV cardiac dysfunction, RVNC could cause such complications as pulmonary hypertension and tension pneumothorax. It can be also spontaneously improved in GMD.

Keywords: Cardiomyopathy, Gestational diabetes, Newborn, Right ventricular non-compaction, Tension pneumothorax

Introduction

Ventricular non-compaction, which is a rare congenital cardiomyopathy, results from an arrest in normal endomyocardial embryogenesis. It is characterized by deep trabeculations and intertrabecular recesses in the ventricular wall which give the myocardium a "spongiform" appearance (1). Non-compaction cardiomyopathy usually occurs due to the endomyocardial morphogenesis which leads to a failure of trabecular compaction in the developing myocardium (2). Nonetheless, the left ventricle is usually involved in this disorder and the involvement of the right ventricle has been reported in children (3-5).

The current study reported on a neonate born to a mother with gestational diabetes (GDM) with meconium aspiration. Before the commencement of the study, the informed written consent was obtained from the neonate's parents. During the admission, he was diagnosed with isolated right ventricle non-compaction without particular evidence of LV involvement.

Case report

A 1 day-old boy was admitted to the neonatal intensive care unit (NICU) with cyanosis and respiratory distress due to meconium aspiration syndrome immediately after birth.

He was the virginally born neonate of a mother with GDM at 38 weeks (gestational age) GA with the Apgar scores of 5 and 7 at 1 and 5-minutes, respectively. He was not born to a

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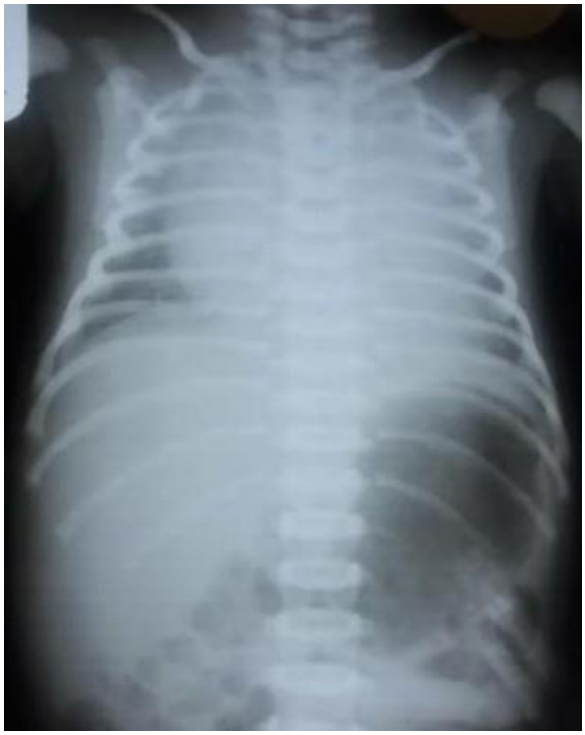

Figure 1. Chest radiograph in AP view demonstrating cardiomegaly with pulmonary vascular marking and no pneumonia aspiration or meconium aspiration

consanguineous couple and there was no positive family history of congenital heart disease. On admission to hospital, he weighed 2580 gr with a height of 50 cm and a head circumference of 36 cm. The vital signs were as follows: the heart rate=167 beats/min, respiratory rate=75 breaths/min, blood pressure= 67/29 mmHg, and oxygen saturation= 80-85% on room air.

According to the cardiovascular examination, he had regular pulses, equal femoral pulses, the apex beat at the 4th intercostal space midclavicular line, normal heart sounds, and 2/6 degrees of holosystolic murmur on the lower left sternal border. Except for mild hepatomegaly in the abdomen, other physical examinations were normal. On the first day, his chest radiography was indicative of cardiomegaly. He needed continuous positive airway pressure ventilation, oxygen therapy via NIPPV, and received surfactant therapy.

On the second day, chest radiography repeated as the O2 saturation decreased, and the need for the fraction of inspired oxygen (FIO2) increased to 70%. The radiography showed a massive right pneumothorax; consequently, a chest tube was inserted for the patient. After 5 days, the air leak improved and he had an oxygen saturation level of 95% (Figure 1a, b). Electrocardiogram showed an extreme right axis deviation (RAD) and right ventricular enlargement (Figure 2).

The echocardiographic findings demonstrated prominent trabecular meshwork with deep intertrabecular recesses in the right ventricle wall, a dilated right ventricle with right ventricular non-compaction, normal left ventricle systolic and diastolic function (figures 3, 4), an ejection fraction of 58%, a right to left shunt through the foramen ovale, and a PDA with the left to right shunt (Systolic blood pressure: 37 mmHg, Diastolic blood pressure: 12mmHg). There was moderate tricuspid regurgitation with a peak pressure gradient of 35 mmHg, and pulmonary artery pressure was obtained at 45 mmHg. The right Systolic function was not acceptable. Tricuspid annular plane systolic excursion

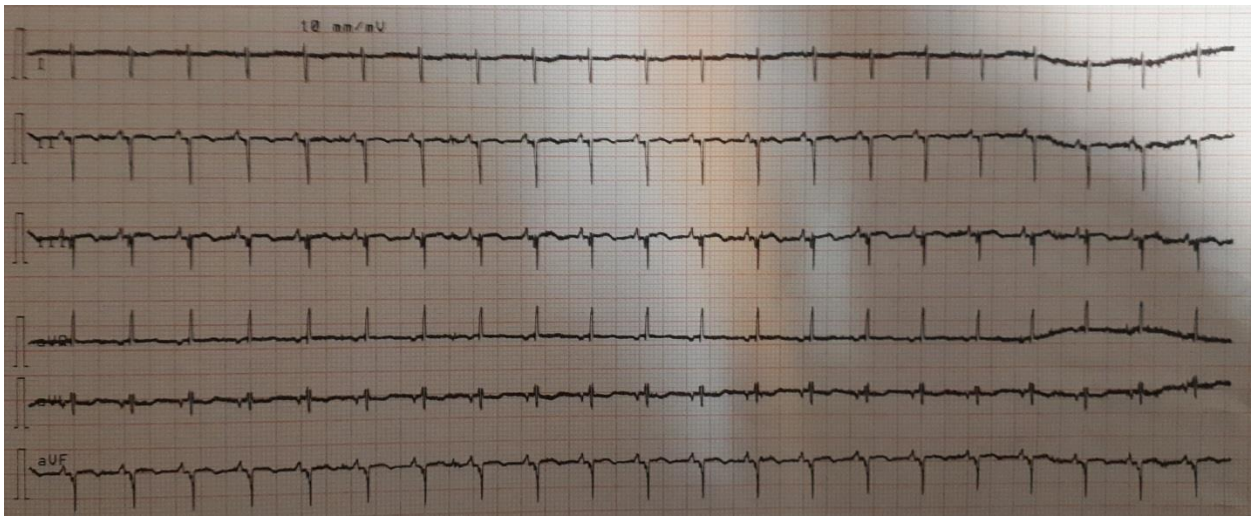

Figure 2. Electrocardiogram showing an extremely right QRS axis and right ventricular enlargement and on ST elevation or arrhythmia

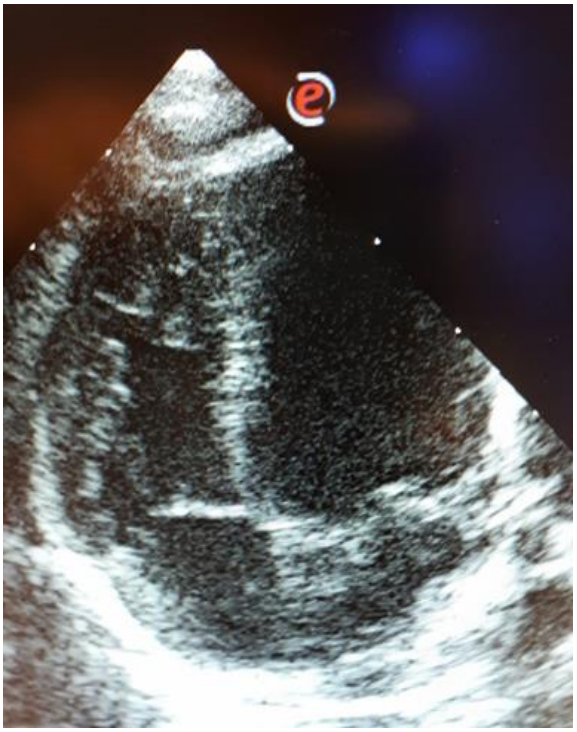

Figure 3. (a) Echocardiography (apical four-chamber view) showing accentuation of the dilation and multiple, prominent muscular trabeculation in the RV

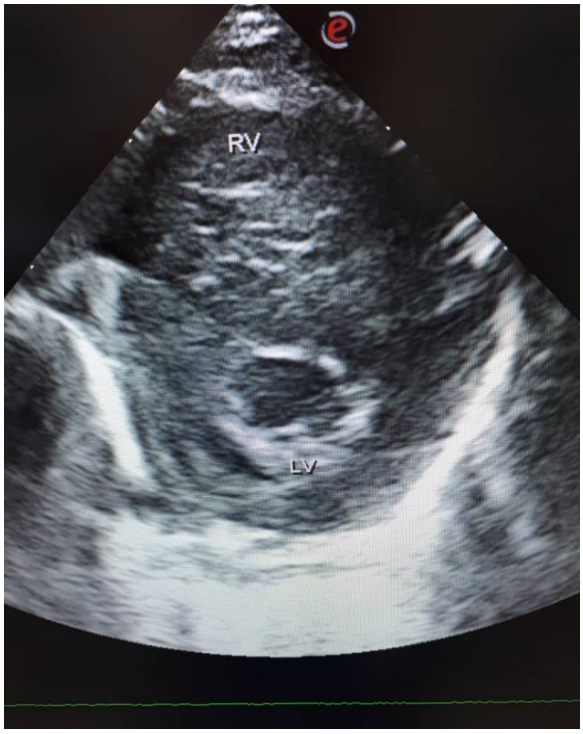

Figure 4. Echocardiography (Subxiphoid short-axis view) demonstrating RV non-compaction

(TAPSE) was reported as 5 mm (Sm=0.01 ms), diastolic function was evaluated with ratio peak mitral inflow velocities at early (E) and late (A) diastole (E/A ratio = 2.87), and diastolic function was disordered. To treat right heart failure, he received diuretic therapy and inotrope medications, such as furosemide (1 mg/kg/day) and milrinone drip (0.5 mcg /kg/min). On the 10th day, the echocardiogram showed that TAPSE increased to 9 mmHg, and left ventricular ejection fraction was 69%. Finally, after 18 days, he was discharged while he was healthy and breastfeeding well with no respiratory distress.

4 months later, at the follow-up visit, the patient looked well and reached a weight of 5.9 kg at the age of 3 months. The echocardiogram demonstrated mild enlargement dimension of the right ventricle, TAPSE of 14 mm, E/A ratio=1.43 with mild tricuspid regurgitation (pressure gradient=18 mmHg), and a normal left ventricular ejection fraction (65%). The right ventricle showed evidence of RVNC cardiomyopathy, intertrabecular recesses in the RV free wall, and RV enlargement. 6-month follow-up showed total improved RVNC and normal reference values for systolic and diastolic parameters.

Discussion

Ventricular non-compaction is a genetic heterogeneous cardiomyopathy characterized by intertrabecular recesses and prominent trabeculations protruding from ventricular walls. Non-compaction is suggested to be the result of an arrest in the normal myocardial compaction during the gestational age of 5-8 weeks. Nevertheless, the pathophysiology of ventricular dysfunction is not clear. It seems that arrhythmias result from fibrosis caused by subendocardial ischemia and coronary microcirculatory dysfunction (CMD) (6). Since the pathogenic non-compaction cannot be easily differentiated from normal trabeculations, it is hard to accurately measure the prevalence of RV involvement.

Imaging is the base of morphological diagnosis for ventricular non-compaction, and echocardiography is the main diagnostic tool for the diagnosis. Moreover, cardiac magnetic resonance imaging (MRI) has received special attention to confirm the diagnosis and search for possible complications (6, 7). Non-compaction cardiomyopathy can co-occur with congenital cardiac malformations, such as septal defects, aortic stenosis, and aortic coarctation. This disorder is called isolated when there are no other congenital diseases. The hereditary pattern is also

considered a male predominance.

Heart failure can be the presentation of non-compaction cardiomyopathy in infants and neonates, while children and adolescents may present this disorder with cardiac failure, stroke, or arrhythmia. It can sometimes be asymptomatic and identified incidentally, similar to the patient reported in the present study; nonetheless, this patient had the clinical manifestation of right heart failure. Based on recent studies on the gene localization in this disorder and other case reports, the location and type of the mutation are not correlated with the disease phenotype or severity in isolated ventricular non-compaction (8). There was also no abnormality in the echocardiographic screening of first-degree relatives in the current study.

According to other studies, atrial fibrillation, Complete Right Bundle Branch Block, and ventricular arrhythmia can be observed in RVNC (1). Moreover, the prominent trabecular meshwork on the luminal surface of the ventricle was presumed to increase the probability of an ectopic pacemaker and re-entrant pathways (9). In the present study, there were no ventricular or atrial arrhythmias. In other studies, certain criteria were defined for the diagnosis of RVNC, including the transmural thickness of the non-compacted RV greater than 75% and dilation of the RV (10).

In a study conducted by Yun et al. (9), just one in four patients had a normal-volume RV. In another study, it was suggested that the diagnosis should be made only when the accentuation of the trabecular meshwork is present in certain areas and RV is dilated (10). The diagnosis of IRVNC in this study was based on these four criteria which were detected in the infant: 1. prominent and deep trabeculations and inter-trabecular recesses in the lateral wall and apex, 2.direct blood flow from the ventricular cavity into the deep inter-trabecular recesses, 3. two-layered structure of the ventricular wall with an end-systolic ratio of non- compacted-to-compacted layer exceeding 1.4 (in infants), and 4. coexisting with no cardiac malformation (11).

In this case, the parents well cooperated and permitted the cardiologist to completely evaluate the potential complications and performed all the required examinations. The neonate was recovered without any organ damage and central nervous system complications. However, there was a limitation during the diagnosis of this case since it was not possible to perform a cardiac MRI due to the age of the patient. Ultimately, there is no agreement on the exact treatment of IRVNC; nonetheless, diuretics, digoxin, angiotensin-converting enzyme inhibitors, carvedilol and calcium channel blockers help control the disorder (12).

Conclusion

As evidenced by the obtained results, we can conclude that it is of paramount importance to consider RVNC cardiomyopathy as a differential diagnosis of cyanosis and meconium aspiration syndrome in a neonate born to a mother with GDM. Apart from RV cardiac dysfunction, RVNC could cause such complications as pulmonary hypertension and tension pneumothorax. It can be also spontaneously improved in GMD.

Acknowledgments

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Conflicts of interests

The authors declare that they have no conflict of interest regarding the publication of the present article.

References

1. Maron BJ, Towbin JA, Thiene G, Antzelevitch C, Corrado D, Arnett D, et al. Contemporary definitions and classification of the cardiomyopathies: an American heart association scientific statement from the council on clinical cardiology, heart failure and transplantation committee; quality of care and outcomes research and functional genomics and translational biology interdisciplinary working groups; and council on epidemiology and prevention. Circulation. 2006; 113(14):1807-16.
2. Weiford BC, Subbarao VD, Mulhern KM. Noncompaction of the ventricular myocardium. Circulation. 2004; 109(24):2965-71.
3. Kaur J, Aziz BB, Jaafar R. Isolated right ventricular non-compaction in a newborn. Progress Pediatr Cardiol. 2019; 52:57-9.
4. Maheshwari M, Gokroo RK, Kaushik SK. Isolated non-compacted right ventricular myocardium. J Assoc Physicians India. 2012; 60:56-7.
5. Saglam M, Saygin H, Kozan H, Ozturk E, Mutlu H. Noncompaction of ventricular myocardium involving the right ventricle. Korean Circ J. 2015; 45(5):439-41.
6. Aggarwal S, Kalavakunta J, Gupta V. A case of isolated right ventricle noncompaction with ST-Elevation chest leads. Heart Views. 2016; 17(1):30-4.
7. Boughrarou R, Mansouri B. Noncompaction of ventricular myocardium: a magnetic resonance imaging study of 50 patients. Eur Soc Radiol. 2016; 10:C-0430.

8. Markiewicz-Loskot G, Moric-Janiszewska E, Loskot M, Szydlowski L, Weglarz L, Hollek A. Isolated ventricular non-compaction: clinical study and genetic review. Europace. 2006; 8(12):1064-7.
9. Yun H, Zeng MS, Jin H, Yang S. Isolated noncompaction of ventricular myocardium: a magnetic resonance imaging study of 11 patients. Korean J Radiol. 2011; 12(6):686-92.
10. Fazio G, Lunetta M, Grassedonio E, Gullotti A, Ferro G, Bacarella D, et al. Noncompaction of the right ventricle. Pediatr Cardiol. 2010; 31(4):576-8.
11. 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(6):666-71.

Lim S, Ahn J, Lee D, Park M. Isolated right ventricular noncompaction accompanied by right ventricular failure. Korean J Med. 2015; 88(1):69-73.