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An Extreme Pan-Biventricular Infantile Hypertrophic Cardiomyopathy: About a Scarce Case

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Abstract

Hypertrophic cardiomyopathy (HCM) is one of the common cardiomyopathies in cardiology practice known by a characteristic involvement of the left ventricle. We report a rare and intriguing case of severe HCM with right ventricle involvement in a child.

Keywords

Hypertrophic cardiomyopathy; Infant; Right ventricle.

Abbreviation

HCM: hypertrophic cardiomyopathy; DCM: dilated cardiomyopathy; ECG: electrocardiogram; SAM: Systolic anterior motion; TM: Time Motion.

Introduction

Hypertrophic cardiomyopathies in children cover a much greater semiological, sonographic and etiological diversity than that of adults. In pediatrics, emphasis should be placed on etiological investigation. These patients must be cared for in highly specialized centers with expertise not only in cardio pediatrics but also in metabolic pediatrics and genetics.

Case

A 12-year-old child in school, an only son, was hospitalized in our department for exploration of fatigue associated with increasing dyspnea on exertion. During the interrogation there was no notion of sudden death in the family at a young age. On examination his BP was 11/7 cm Hg with a regular pulse of 70bpm. The patient had no craniofacial malformation, skin pigmentation or visceromegaly. There was a mid-thoracic systolic murmur of 4/6. The ECG showed a regular sinus rhythm with electrical hypertrophy of the left ventricle (LV). Echocardiography showed, in addition to a very diffusely hypertrophied left ventricle with an indexed LV mass of 180 g/m² and a thickness of the inter ventricular septum of 48 mm (figure 1), a right ventricle also hypertrophied with a thickness of its wall free that exceeds 10 mm. The left atrium was moderately dilated at 38 ml/m². The cavity of the left ventricle was reduced to a small opening in the shape of a banana due to the phenomenal hypertrophy of the walls. In TM mode the patient had a systolic anterior motion (SAM) without mid-systolic collapse of the aortic valve (figure 2). On Doppler, there was a maximum spontaneous left intra ventricular gradient of 100 mm Hg (figure 3). Based on the echocardiography data, we concluded that there was pan-bi-ventricular obstructive HCM. The patient was put on metoprolol and isoptine but he kept the intra-LV gradient high. Her 24-hour rhythm holter showed flaps of unsustained ventricular tachycardias. A myomectomy with defibrillator implantation was offered to the student's parent but refused. We were satisfied with medical treatment, implantation of a defibrillator and regular clinical/telemetric follow-up which has so far been without any particular events. As part of an integral care of our patient, a genetic, metabolic and echocardiographic investigation of the patient and his family was carried out but the results remained negative (Figure 1-3).

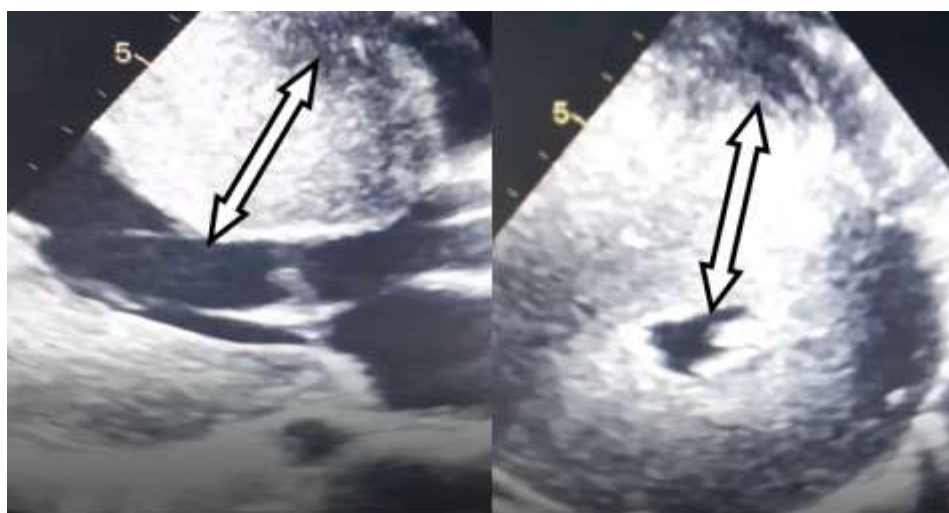


Figure 1: Heavily thickened LV.

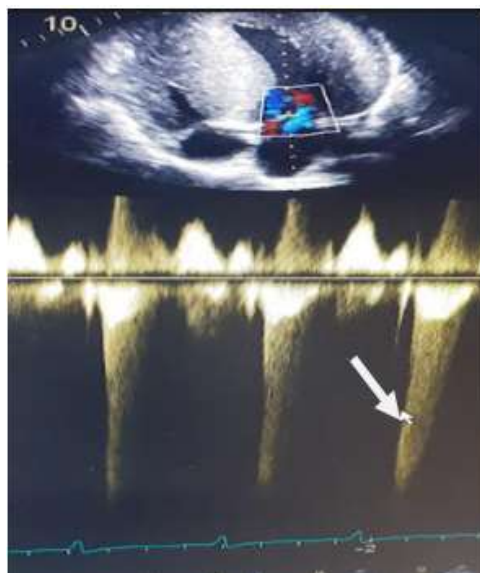


Figure 2: Banana shaped LV.

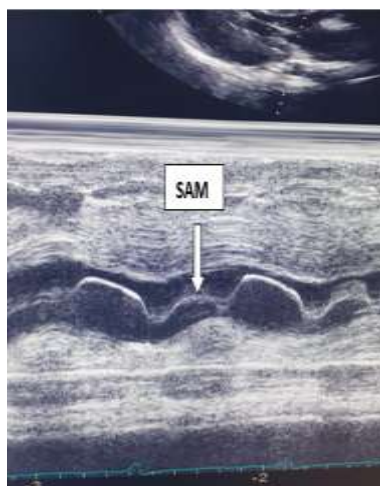


Figure 3: SAM LV-gradient, Hypertrophied RV.

Discussion

A thickness of the LV wall larger than 15 mm on imaging (> 13 mm in relatives' adults) or well above two standard deviations above the anticipated mean in children retains a positive diagnosis of HCM in adults and/or a probationer (first patient diagnosed as having HCM within a family). Abnormal loading conditions should not account for this rise in wall thickness. Twenty-five to thirty percent are the result

of an unknown cause [1]. Metabolic diseases, mitochondrial cardiomyopathies, neuromuscular diseases, malformation syndromes, infiltrative heart disease, endocrine heart disease, and heart disease secondary to therapy such as ciclosporin, hydroxylchloroquine, or steroids account for only 5 to 10% of genetic or non-genetic causes [2]. Indeed, the difference between adult and child HCM is due to the fact that most child hypertrophies are primitive, have a poor prognosis, and are diagnosed after a primitive infantile syndrome that includes neuromuscular signs or metabolic impairment, as well as a potential risk of DCM secondary to HCM [2]. In our patient, the massive form [3] of the HCM was notable, as is the biventricular involvement, which is unusual given that the HCM is traditionally associated with LV hypertrophy. In the literature, the occurrence of RV involvement is poorly known, and it is frequently connected with lentiginosis, which is not the case in our child [4,5].

Conclusion

Biventricular HCM is an extremely rare pathology in the cardiology setting. It is often secondary to a metabolic or genetic pathology. Our case remained idiopathic unless it was mitochondrial cardiomyopathy which will require a myocardial biopsy which is not often available in all centers.

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