Case Report

A rare case of isolated parachute mitral valve in a young girl

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Abstract

The parachute mitral valve is an extremely rare inborn malformation of the mitral valve. It is usually seen in younger children. This is a case of a 17-year-old girl who presented with high output heart failure secondary to severe iron deficiency anaemia, and her transthoracic echocardiography revealed all chordae tendineae of the mitral valve inserted into one papillary muscle in the left ventricular cavity with grade I-II mitral regurgitation and mild mitral stenosis. Other inborn heart malformations were not found. Hence, the diagnosis of an isolated parachute mitral valve was made. She has one of the rare cases of inborn malformation of the mitral valve.

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Introduction

Clinically remarkable inborn mitral valve lesions are rare and constitute approximately 0.4% of inborn heart diseases [1]. A parachute mitral valve (PMV) is a rare inborn malformation where all chordae tendineae are shorter and thicker than usual and are inserted into a single papillary muscle. It could be found either as an isolated malformation or associated with left heart obstructive malformation, which is named Shone's complex [2]. PMV limits the movement of the leaflets of the mitral valve and interrupts the blood flow into the left ventricle throughout the diastole [2].

In our case, we describe a rare case of isolated PMV (IPMV) who presented with high output heart failure secondary to severe iron deficiency anaemia.

Case report

A 17-year-old previously healthy girl presented with bilateral leg oedema and exertional breathlessness for a one month duration. Her developmental history was normal. She was severely pale and tachypnoeic. She had tachycardia, and her blood pressure was normal in both upper limbs. The heart sounds were normal, but she had a grade 2 systolic murmur at the mitral area but no radiation. Examination of other systems revealed fine crepitations in the basal lung with mild to moderate hepatomegaly suggestive of congestion.

Laboratory tests showed severe iron deficiency anaemia with low haemoglobin (1.8 g/dL) and a high eosinophil count of 680/µL. Her liver function, renal function, TSH, and serum electrolytes were normal. Her electrocardiogram and chest X-ray showed sinus tachycardia and mild cardiomegaly, respectively.

Transthoracic echocardiography (TTE) revealed a normal sized left ventricle (LVEDD: 44 mm) and left atrium. Right-sided chambers were normal and systolic functions of both sides of the heart were preserved (LVEF:60%). There was a thickened chordae tendineae

of the mitral valve attached to a single papillary muscle on the inferolateral wall of the left ventricle suggestive of PMV (Figure 1) with grade I-II mitral regurgitation and mild mitral stenosis. Other inborn malformations, including coarctation of the aorta, thickened mitral annulus, supravalvular mitral membrane, or any outflow tract obstruction, were not found. Hence, the diagnosis of IPMV was made.

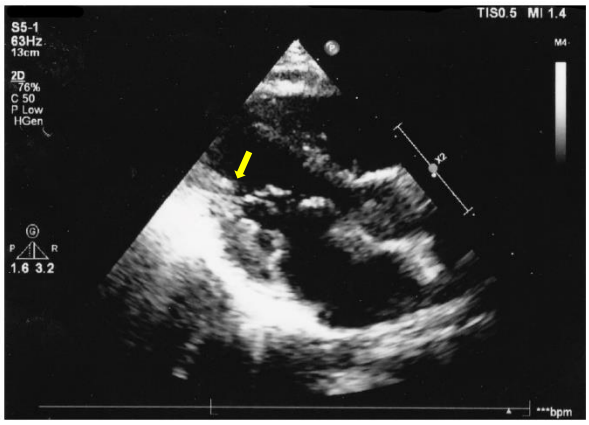

Figure 1: Transthoracic echocardiography showing chordae tendineae of the mitral valve attached to only one papillary muscle (yellow arrow) on the wall of the left ventricle

Blood was transfused to optimise the haemoglobin level to around 8g/dL and frusemide was given to alleviate the symptoms of congestion due to anaemia-induced heart failure. Iron supplements were prescribed along with worm treatment (Mebendazole 100 mg bd for 3 days). She completely recovered from heart failure symptoms on the seventh day of admission. She was discharged with haematinics. She was regularly followed up in the medical clinic. At the follow-up visit of the third month, a repeat TTE was done, and her haemoglobin was 12.1 g/dL. From the PMV point of view, she was asymptomatic. However, she needs regular annual echocardiogram assessment which was arranged. High eosinophil count with an absence of bleeding history, such as heavy menstrual bleeding and per rectal bleeding, suggested that worm infestation was the possible aetiology of iron deficiency anaemia. We planned further investigation to find the cause of iron deficiency if she fails to respond to haematinics and worm treatment.

Discussion

PMV is referred to as all chords converging and inserted into one papillary muscle, commonly seen in the paediatric age group [3]. In most instances, PMV is a part of Shone's complex, which includes PMV, subaortic stenosis, coarctation of the aorta, and supravalvular mitral ring [2]. This condition is a persistent embryonic circumstance caused by interrupted lamination of the posterior and anterior portions of the trabecular ridge between 5th and 19th weeks of gestational age, driving them into one papillary muscle [4].

Substantial haemodynamic effects leads to early presentation and diagnosis in the paediatric age group. Therefore, undiagnosed adult cases are very rare with only a few incomplete forms of PMV have been detected in adults [5,6]. The complexity and severity of malformations may be the deciding factor in a variety of manifestations. Thus, adult patients with undiagnosed PMV present with a spectrum of symptoms from accidental detection to heart failure. In our case, the heart failure was due to severe anaemia rather than PMV *per se*, which was supported by the recovery of heart failure after treating the anaemia and the findings of the valvular lesion in our patient were not severe enough to cause heart failure. The high output-heart failure is usually caused by severe anaemia, as in this case, arteriovenous fistula, beriberi, hyperthyroidism, and Paget disease.

In a systematic review by Hakim et al, only 9 adult patients with PMV have been described till 2010 [6]. As in our case, an isolated case of PMV in a twenty-nine-year-old woman was described in 2016 [9]. In 2018 Bade et al reported 2 cases of severe inborn mitral stenosis associated with severe pulmonary hypertension secondary to PMV [10].

According to the available literature, there is no association between PMV and iron deficiency anaemia. Despite the presence of PMV, our patient tolerated anaemia which indicates the severity of PMV in our patient is minimal.

Currently, two-dimensional TTE is the diagnostic tool, whereas transoesophageal echocardiography (TOE) is confirmatory in difficult cases [7]. A pseudo-parachute mitral valve where all chordae are converged to major papillary muscle and the other papillary muscle being atrophied can be differentiated from true PMV by careful study of TTE but often need TOE. Our patient's PMV finding was quite clear with TTE, and further study of TOE was not required.

PMV is mostly associated with mitral stenosis and infrequently with mitral regurgitation. The below minimal hemodynamically significant PMV requires no intervention [6]. However, such patients must be carefully followed because PMV has progressive evolution leading to unstable haemodynamics [5,8].

Mitral valve replacement or repair is required only in haemodynamically considerable stenosis or regurgitation of PMV [11]. In our case, as the patient was asymptomatic specific treatment was not necessary. However, annual surveillance should be carried out.

Conclusion

This case report has brought out awareness of an IPMV malformation.

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Consent

Written consent was obtained from the patient and parent for the publication of this study.

Patient perspective

I feel very happy that doctors from Sri Lanka and other countries will learn something from my case. I was very sick when I got admitted to Teaching hospital Jaffna. I had leg swelling, breathlessness, and poor exercise capacity. My relatives and neighbours asked my mother to show me to a doctor. I was anxious and thought that I got a bad disease.

I got to know from doctors that I had low haemoglobin due to fewer iron nutrients. They gave blood and iron tablets. They also told that my heart valve had some abnormality, but which was not the cause of my symptoms. My mother and I was advised by doctors that I needed a regular clinic to follow up and that my heart should be scanned every year. To prevent worms, I learnt hygienic habits from health staff.

I feel very happy and normal now. I became active after treatment. I want to thank my doctors and ward staff for what they did for me.

References

1. Nadas AS. Pediatric *Cardiology*. 3rd ed. Philadelphia: Saunders; 1972.
2. Hone JD, Sellers RD, Anderson RC, Adams P Jr, Lillehei CW, Edwards JE. The developmental complex of "parachute mitral valve," supravalvular ring of left atrium, subaortic stenosis, and coarctation of aorta. *Am J Cardiol.* 1963;11:714-25.
3. Schiebler GL, Edwards JE, Burchell HB, Dushane JW, Ongley PA, Wood EH. Congenital corrected transposition of the great vessels: A study of 33 cases. *Pediatrics* 1961;27 (5) Suppl: 849-88
4. Oosthoek PW, Wenink AC, Wisse LJ, Gittenberger-de Groot AC. Development of the papillary muscles of the mitral valve: Morphogenetic background of parachute-like asymmetric mitral valves and other mitral valve anomalies. *J Thorac Cardiovasc Surg* 1998;116:36-46
5. Schaverien MV, Freedom RM, McCrindle BW. Independent factors associated with outcomes of parachute mitral valve in 84 patients. *Circulation* 2004;109(19):2309-2313. DOI: 10.1161/01.CIR.0000129269.98460.C8
6. Hakim FA, Kendall CB, Alharthi M, Mancina JC, Tajik JA, Mookadam F. Parachute mitral valve in adults—a systematic overview. *Echocardiography* 2010;27(5):581-586. DOI: 10.1111/j.1540-8175.2009.01143.x.
7. Yesilbursa D, Miller A,Nanda NC, et al: Echocardiographic diagnosis of a stenotic double orifice parachute mitral valve with a single papillary muscle. *Echocardiography* 2000;17:349 352. DOI: 10.1111/j.1540-8175.2000.tb01149.x.
8. Marino BS, Kruge LE, Cho CJ, Tomlinson RS, Shera D, Weinberg PM, et al. Parachute mitral valve: morphologic descriptors, associated lesions, and outcomes after biventricular repair. *J Thorac Cardiovasc Surg* 2009; 137(2): 385-393 e4. DOI: 10.1016/j.jtcvs.2008.09.016
9. Toufan M, Mahmoudi SS. Isolated parachute mitral valve in a 29 years old female; a case report. *J Cardiovasc Thorac Res* 2016;8(1):43-45. DOI: 10.15171/jcvtr.2016.08.
10. Bade AS, Patil V, Shaikh S, Khemani H, Singh G, Bansal NO. Parachute mitral valve with late presentation: rare case reports. *Int J Res Med Sci* 2018;6:2850-2. DOI: 10.18203/2320-6012.ijrms20182935.
11. Yuan SM. Parachute mitral valve: Morphology and surgical management. *Turk Gogus Kalp Damar Cerrahisi Derg.* 2020;28(1):219-226. DOI: 10.5606/tgkdc.dergisi.2020.18041.