DOI: 10.4274/tpa.570

Hammock mitral valve as a cause of atrial flutter: A 3-month-old infant case

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Summary

Hammock mitral valve is a very rare congenital valve disorder. It may lead to mitral stenosis or insufficiency in the pediatric age group. These patients generally present with the symptoms of congestive heart failure or sometimes sudden death may occur. Our case was a 3-month-old female infant who presented to our clinic with the symptoms of heart failure. Her cardiac evaluation revealed hammock mitral valve, left atrial dilatation, severe mitral valve insufficiency and atrial flutter. In contrast to adult patients, atrial flutter is a very rare type of cardiac aryhtmia in the pediatric age group. We present this case, because no such case with atrial flutter resulting from hammock mitral valve has been reported in the literature. *(Turk Arch Ped 2013; 48: 244-247)*

Key words: Atrial flutter, hammock mitral valve, mitral insufficiency

Introduction

Hammock mitral valve was defined by Layman and Edwards (1) in 1967 for the first time. In this anomaly, the mitral valve is bound to the anterior papillary muscle directly or by a short bond. In addition, thickening and flexion is observed in the free border of the mitral valve. This disrupted valve structure limits the movements of the valve and leads to stenosis or insufficency in the valve. Although stenosis is also observed in the mitral valve, mitral insufficiency is confronted more frequently. Mitral valve insufficiency may be observed in association with other congenital cardiac anomalies, collagen tissue disorders and metabolic or storage diseases (2,3,4,5).

Atrial flutter rhythm observed in the neonatal period or infancy is not frequently accompanied by cardiac anomalies (6). Rarely, atrial flutter rhythm may be observed in relation with cardiac anomaly. Ebstein anomaly is one of the cardiac anomalies in which atrial flutter rhythm may be observed. In addition, atrial flutter rhythm may also be observed in older children following cardiac surgery (including Fontan, Senning and Mustard) (7). In this patient, mitral valve anomaly accompanied atrial flutter rhythm. Mitral insufficiency and left atrial enlargement which developed because of hammock mitral valve were the cause of atrial flutter.

The diagnoses of left atrial enlargement developing because of hammock mitral valve and atrial flutter were made in a 3-month-old female infant who presented to our clinic with findings of heart failure. In contrast to adult patients, atrial flutter is a rare arryhthmia in infancy. This case was considered to be notable and was presented, because atrial flutter occured because of a rare congential cardiac anomaly like hammock mitral valve.

Case

A 3-month-old female infant was presented to our clinic with complaints including rapid respiration, getting tired quickly while breastfeeding and difficulty in breathing which

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started 2 weeks ago and gradually increased. In her history, it was learned that she was born at the 35th gestational week with a birth weight of 2100 g as a triplet partner. The babies had no problem after birth. In the familial history, it was learned that there was no consangineous marriage between the mother and father, the father had mitral valve prolapsus and the patient had a healthy 8-year-old sister. There was no known disease in the family.

Physical examination findings were as follows: body weight: 4.1 kg (3-10%), height: 54 cm (3-10%), heart rate: 150/min, respiratory rate: 32/min, blood pressure: 75/49 mmHg, body temperature: 36.2°C (axillary). A 3/6 pansystolic murmur was heard at the apex. The liver was palpable 3 cm below the costal margin in the middle line.

On electrocardiogram (ECG), left atrial enlargement and atrial flutter (Figure 1) were found and telecardiography revealed cardiomegaly and left atrial enlargement (Figure 2). Left ventricular systolic functions were found to be normal on echocardiographic examination (Ejection fraction: 65%, shortening fraction: 38%). Advanced enlargement in the left atrium (2.3 cm, normal: 1.5-2.1cm), left ventricular enlargement (2.91 cm, normal: 1.80-2.30 cm), thickening in the mitral anterior leaflet, short bonds and severe mitral insufficency (Figure 3A and 3B) were found.

After the patient was hospitalized in the ward, digoxin, angiotensin converting enzyme inhibitor (ACEI) and furosemide treatment was started for heart failure and atrial flutter. Afterwards, propranolol was added to the treatment. It was observed that the cardiac rate which was approximately 160/min before treatment decreased to 125/min 24 hours after the treatment was started. When the heart rate was controlled, findings of heart failure regressed. The patient's rhythm returned to normal with drug treatment. Therefore, other treatment options including cardioversion or transesophageal rapid pacemaker were not considered. It was thought that significant mitral insufficiency due to hammock mitral valve led to left atrial enlargement and atrial flutter. Therefore, it was decided that the mitral valve be repaired surgically. However, the operation was postponed to one year of age, since the too young age of the patient would affect the success of the operation. The patient

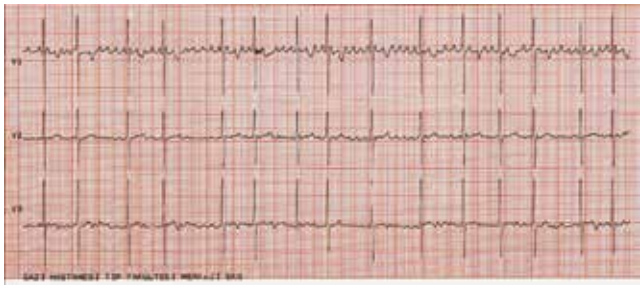

Figure 1. Atrial flutter rhythm on electrocardiogram of the patient (sawtooth)

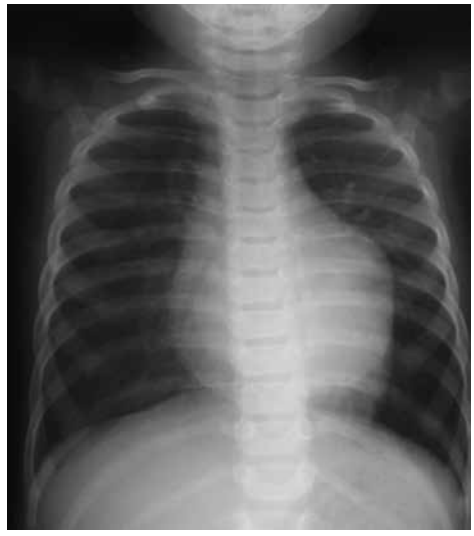

Figure 2. Left atrial enlargement on telecardiogram of the patient (double contour shadow)

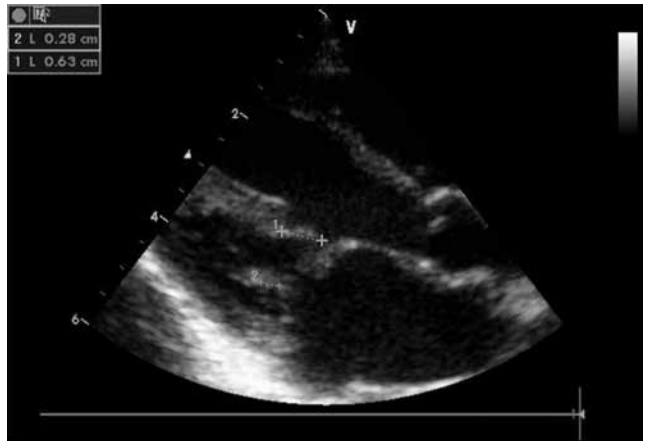

Figure 3a. Short chordae are observed on long-axis imaging on echocardiogram

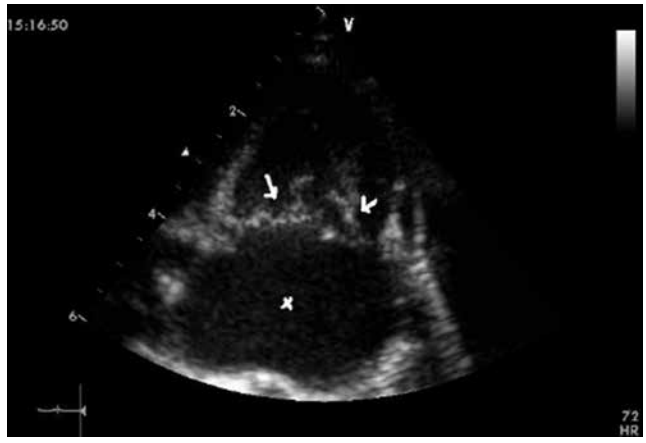

Figure 3b. Enlarged left atrium (x) and thickenning and curling (white arrows) in the mitral valve are observed on echocardiogram

was asked to come back for monthly follow-up visits and was followed up for four months without any problem. An upper respiratory tract infection at the age of seven months caused to heart failure again. The patient was hospitalized and intravenous treatment was adjusted. However, heart failure and infection could not be controlled. Respiratory failure developed in the patient and mechanical ventilation was started. Two weeks later, she was lost because of multiple organ failure, sepsis and heart failure.

Discussion

Hammock mitral valve is a congenital mitral valve disease which is observed considerably rarely in children. A short mitral valve bond or lack of mitral valve bond leads to dysfunction of the valve. The clinical picture varies according to the age of the patient and the accompanying cardiac anomaly. In congenital mitral valve diseases, findings of heart failure may occur in the early period of life. Patients may present with inability to gain weigth and frequent lower respiratory tract infections (5). In addition, left atrial enlargement arising from significant mitral failure may compress the left lung (8).

Atrial flutter is an arrythymia which is observed rarely in the newborn period and infancy and is frequently not accompanied by congenital heart anomalies. The time period of atrial flutter, the time of onset, the degree of ventricular response to atrial flutter, the time of onset of the findings are the factors which determine the clinical status (6,7). The first-line treatment in atrial flutter is transesophageal rapid pacemaker or cardioversion in patients who are not well hemodynamically (6, 9). If these therapies are unsuccessful, the ventricular rate may be controlled by digoxin. Digoxin is a very well known drug which has been used in acute treatment of atrial flutter for a long time (9). In this patient, digoxin, diuretic and ACEI treatment was started primarily to control atrial flutter and ventricular rate. Afterwards, propranolol was added to treatment. With this treatment the findings of heart failure regressed. Since atrial flutter was also controlled, other treatment options including cardioversion or transesophageal rapid pacemaker were not considered.

Hammock mitral valve is one of the congenital heart anomalies which are difficult to treat surgically. Replacement of mitral valve with mechanical valve is a considerably difficult operation in small infants. Especially small mitral valve in the infant, requirement of anticoagulant treatment after valve replacement and risks for re-replacement cause the operation to be performed in the latest possible period. In addition, the fact that the medium-term results of mitral valve replacement operations performed below the age of one year are unsuccessful supports the view that operations should be performed above the age of one. Mitral valve repair may be performed in eligible subjects instead of mitral valve replacement operation. Mitral valve replacement is preferred with a higher frequency because of low mortality risk and low re-operation risk (10,11). In our patient, a decision of mitral valve repair was made, but it was planned to perform the operation at about the age of one year, since mitral valve replacement might be necessary during the operation. A procedure like mitral angioplasty by catheter angiography was also not considered, since mitral failure was at the forefront and no prominent mitral stenosis was present. No other treatment was considered, since heart failure and atrial flutter was controlled. No problem was observed during the four-month-follow-up period, but we lost the patient because of sepsis and heart failure which developed following an intervening and uncontrollable respiratory tract infection. It was thought that left atrial enlargement arising from significant mitral failure related with hammock mitral valve led to atelectasis by compressing the left lung, prolongation of the lower respiratory infection and inability to seperate the patient from the ventilator.

Patients diagnosed with atrial flutter in the neonatal period or infancy should be evaluated in detail by echocardiography in terms of ventricular functions and structural heart disease. In cases where accompanying congenital heart anomaly is not present, it should be kept in mind that the cause of mitral failure may be mitral valve anomaly and assessment should be done accordingly. In patients with heart failure and atrial flutter arising from mitral valve anomaly who do not respond to drug treatment, surgical decision should be made even in infancy without delay.

References

1. Layman TE, Edwards JE. Anomalous mitral arcade. A type of congenital mitral insufficiency. Circulation 1967; 35: 389-395.
2. Davachi R, Moller JH, Edwards JE. Diseases of the mitral valve in infancy: anatomic analysis of 55 cases. Circulation 1971; 43: 565-579.
3. Ruckman RN, Van Praagh R. Anatomic types of congenital mitral stenosis: report of 49 autopsy cases with consideration of diagnosis and surgical implications. Am J Cardiol 1978; 42: 592-601.
4. Carpentier A. Congenital malformations of the mitral valve. In: Stark J, de Laval M, (eds). Surgery for congenital heart defects. London, Grune & Stratton 1983; 467-482.
5. Chauvaud S. Surgery of congenital mitral valve disease. J Cardiovasc Surg 2004; 45: 465-476.
6. Texter KM, Kertesz NJ, Friedman RA, Fenrich AL Jr. Atrial flutter in infants. Am Coll Cardiol 2006; 48: 1040-1046.
7. Greason KL, Dearani JA, Theodoro DA, Porter CB, Warnes CA, Danielson GK. Surgical management of atrial tachyarrhythmias associated with congenital cardiac anomalies: Mayo Clinic experience. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2003; 6: 59-71.

8. Tunaoğlu FS, Halid V, Olguntürk R, Ozbarlas N, Kula S, Sinci V. An infant with severe mitral insufficiency and collapse of the left lung due to hammock mitral valve: emergency mitral valve replacement. Anadolu Kardiyol Derg 2006; 6: 283-285.
9. Garson A Jr, Bink-Boelkens M, Hesslein PS, Hordof AJ, Keane JF, Neches WH, Porter CJ. Atrial flutter in the young: a collaborative study of 380 cases. J Am Coll Cardiol 1985; 6: 871-878.
10. Prifti E, Vanini V, Bonacchi M, Frati G, Bernabei M, Giunti G, Crucean A, Luisi SV, Murzi B. Repair of congenital malformations of the mitral valve: early and midterm results. Ann Thorac Surg 2002; 73: 614-621.
11. Stellin G, Padalino M, Milanesi O, Vida V, Favaro A, Rubino M, Biffanti R, Casarotto D. Repair of congenital mitral valve dysplasia in infants and children: is it always possible? Eur J Cardiothorac Surg 2000; 18: 74-82.