Eur J Pediatr (2009) 168:741–743
DOI 10.1007/s00431-008-0818-7

SHORT REPORT

# A neonate with the rupture of mitral chordae tendinae associated with maternal-derived anti-SSA antibody

**Akiko Hamaoka · Isao Shiraishi · Masaaki Yamagishi · Kenji Hamaoka**

Received: 8 June 2008 / Accepted: 7 August 2008 / Published online: 29 August 2008


**Abstract** Acute mitral insufficiency due to the rupture of chordae tendinae from papillary muscles is a rare but sometimes fatal condition in neonates and infants. Here, we report a 21-day-old neonate with sudden onset of mitral insufficiency due to rupture of the chordae tendinae. The baby was successfully rescued by appropriate diagnosis and emergency surgical operation with reconstruction of the ruptured chordae using autologous pericardium-reinforced mattress suture. Intraoperative findings revealed fibrous scar formation at the distal end of the posterior papillary muscle, indicating a remnant of a certain inflammatory reaction. Transplacental passage of maternal anti-SSA antibody could account for the fetal inflammation at the ruptured chordae and the papillary muscle.

**Keywords** Mitral insufficiency · Rupture of chordae tendinae · Fetal inflammation · Anti-SSA antibody

A. Hamaoka (✉) · I. Shiraishi · K. Hamaoka
Department of Pediatric Cardiology and Nephrology,
Graduate School of Medical Science,
Kyoto Prefectural University of Medicine,
Kawaramachi-Hirokoji, Kamigyo,
Kyoto 602-8566, Japan
e-mail: ahamaoka@koto.kpu-m.ac.jp

M. Yamagishi
Department of Pediatric Cardiovascular Surgery,
Graduate School of Medical Science,
Kyoto Prefectural University of Medicine,
Kyoto, Japan

## Case report

A male neonate was born at 40 weeks gestation with a body weight of 2,708 g. During pregnancy, his mother showed high titer of anti-SSA and anti-SSB antibodies (118 U/ml and 53.2 U/ml, respectively) without any specific symptoms. He suddenly presented with paleness and dyspnea. On admission, his body temperature was 37.2°C, heart rate was 188 bpm, and respiratory rate was 70/min with retraction. Cardiac auscultation revealed a systolic regurgitation murmur with a third sound. A blood test showed an elevated WBC (24,800/μl) and normal CRP levels (0.23 mg/dl). An arterial blood gas examination demonstrated severe metabolic and respiratory acidosis. The serum level of anti-SSA antibody was markedly elevated (over 500× normal). Electrocardiograms showed elevated R-waves and inverted T-waves in the right precordial leads and a prolongation of the PQ interval (0.16 s). A chest X-ray showed an enlargement of the heart (cardiothoracic ratio 58%) and severe congestion of the lungs. Two-dimensional (2-D) echocardiography demonstrated a prominent prolapse of the posterior leaflet of the mitral valve with ruptured chordae tendinae (Fig. 1a). Further observations revealed a high echoic lesion at the top of the posterior papillary muscle (arrow in Fig. 1b). Severe mitral regurgitation was also detected by a Doppler color-flow study. An acute rupture of the chordae tendinae was suspected and an emergency surgical operation was, therefore, undertaken. The intraoperative findings confirmed a laceration of the chordae tendinae from the posterior papillary muscle (Fig. 1c,d). The distal portion of the posterior papillary muscle was pale and atrophic, thus, suggesting fibrous scar

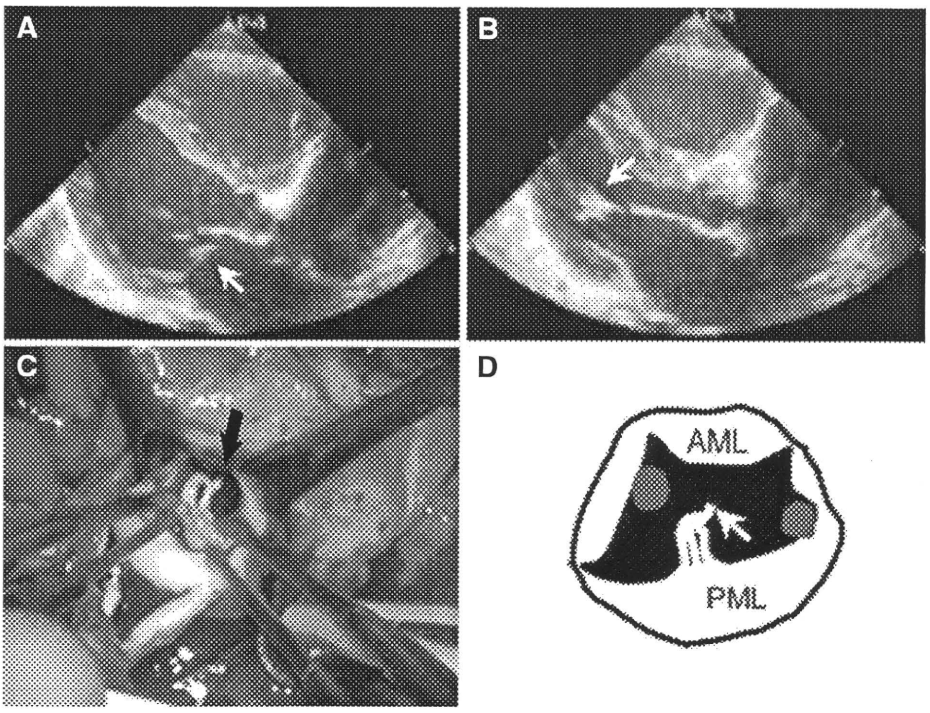


**Fig. 1** **a**, **b** Two-dimensional echocardiograms of the patient before the repair of the ruptured mitral chordae tendinae. **a** A prominent prolapse at the posterior leaflet of the mitral valve is indicated (*arrow*). **b** A high echoic lesion at the distal portion of the posterior papillary muscle is noted (*arrow*). **c** The intraoperative findings demonstrate a rupture of the chordae tendinae (*arrow*) from the posterior papillary muscle. **d** A schematic drawing of the lacerated chordae tendinae (*arrow*) and the mitral valve. AML = anterior mitral leaflet; PML = posterior mitral leaflet

formation after certain inflammation. The lacerated chordae tendinae was anastomosed to the intact portion of the papillary muscle with an autologous pericardium. Mitral annuloplasty was also performed to reduce the mitral regurgitation using a reinforced mattress suture. An intraoperative endocardial biopsy specimen could not be obtained because the scar tissue was located in a critical portion of the papillary muscle repair. After the operation, echocardiograms showed a normal left ventricular function (EF=0.69, FS=0.33), without any significant regurgitation of the mitral valve. The serum level of anti-SSA antibody normalized 6 months after birth. Since undergoing surgery, the patient has been growing well for 2 years. Only the first-degree atrioventricular block (PQ interval 0.20 s) remains.

## Discussion

A rupture of the mitral chordae tendinae in children has been considered to be a very rare condition. However, more cases may remain undiagnosed and untreated than those previously reported because the disease is sometimes fatal without appropriate treatment. The known etiology includes rheumatic fever [1], bacterial or viral endocarditis and myocarditis [2], myxomatous mitral valve [3], blunt chest trauma [5], Kawasaki disease [6], myocardial infarction [7], and Marfan's syndrome [11]. An interesting finding in this patient is the fibrotic scar formation observed at the chordae tendinae and the posterior papillary muscle. This finding strongly suggests the occurrence of some type of inflammation during the fetal period.

Transplacental passage of anti-SSA and anti-SSB antibodies is a well known cause of congenital complete atrioventricular block in neonates [4]. Recent studies have clarified that these antibodies initially bind to the L-type calcium channel of the fetal cardiomyocytes and induce calcium dysregulation, apoptosis, inflammation, and subsequent sinoatrial dysfunction and atrioventricular block [9]. Left ventricular dysfunction, such as dilated cardiomyopathy and endocardial fibroelastosis, has also been reported as late-onset cardiac complications [8]. A neonatal case with a chordal rupture associated with maternal-derived anti-SSA antibody has been reported. Fibrotic scar formation at the papillary muscle, which was very similar to our case, was speculated to be the cause of the rupture in that case [10]. Although a histological study was not performed in our case, the injury of the papillary muscle due to anti-SSA antibody is thought to be the likely cause of the rupture. The persistent PQ interval prolongation also supports the presence of cardiomyocyte injury with anti-SSA antibody.

In conclusion, a rupture of the mitral chordae tendinae is rare. However, it should be recognized as a serious complication of maternal-derived anti-SSA antibody-related cardiac involvement in neonates and infants.

## References


1. Anderson Y, Wilson N, Nicholson R, Finucane K (2008) Fulminant mitral regurgitation due to ruptured chordae tendinae in acute rheumatic fever. J Paediatr Child Health 44:134–137
2. Baird CW, Constantinos C, Lansford E, Pigula FA (2007) Mitral valve chordal rupture masquerades as endocarditis. Pediatr Cardiol 28:297–299
3. Barber JE, Ratliff NB, Cosgrove DM 3rd, Griffin BP, Vesely I (2001) Myxomatous mitral valve chordae. I: mechanical properties. J Heart Valve Dis 10:320–324
4. Buyon JP, Clancy RM (2005) Neonatal lupus: basic research and clinical perspectives. Rheum Dis Clin North Am 31:299–313
5. Grinberg AR, Finkielman JD, Piñeiro D, Festa H, Cazenave C (1998) Rupture of mitral chorda tendinea following blunt chest trauma. Clin Cardiol 21:300–301
6. Mishima A, Asano M, Saito T, Yamamoto S, Ukai T, Yoshitomi H, Mastumoto K, Manabe T (1996) Mitral regurgitation caused by ruptured chordae tendineae in Kawasaki disease. J Thorac Cardiovasc Surg 111:895–896
7. Moursi MH, Bhatnagar SK, Vilacosta I, San Roman JA, Espinal MA, Nanda NC (1996) Transesophageal echocardiographic assessment of papillary muscle rupture. Circulation 94:1003–1009
8. Villain E, Coastedoat-Chalumeau N, Marijon E, Boudjemline Y, Piette JC, Bonnet D (2006) Presentation and prognosis of complete atrioventricular block in childhood, according to maternal antibody status. J Am Coll Cardiol 48:1682–1687
9. Wahren-Herlenius M, Sonesson SE (2006) Specificity and effector mechanisms of autoantibodies in congenital heart block. Curr Opin Immunol 18:690–696
10. Weber HS, Myers JL (1994) Maternal collagen vascular disease associated with fetal heart block and degenerative changes of the atrioventricular valves. Pediatr Cardiol 15:204–206
11. Weidenbach M, Brenner R, Rantamäki T, Redel DA (1999) Acute mitral regurgitation due to chordal rupture in a patient with neonatal Marfan syndrome caused by a deletion in exon 29 of the FBN1 gene. Pediatr Cardiol 20:382–385