

CASE REPORT

Obstructive intramural coronary amyloidosis and papillary muscle rupture

D Coisne, P Corbi, P Sosner

Heart 2003;89:e8 (<http://www.heartjnl.com/cgi/content/full/89/2/e8>)

Mitral papillary muscle rupture is usually caused by ischaemia as a complication of myocardial infarction. In a 76 year old patient with no significant disease or major cardiovascular risk factors, papillary muscle rupture was caused by obstructive intramural coronary amyloidosis, an unusual cause.

The most frequent cause of acute mitral regurgitation is ischaemic papillary muscle dysfunction or rupture complicating acute inferior myocardial infarction. Other causes described in the literature are infectious and traumatic. We report an unusual cause of papillary muscle rupture successfully operated on.

CASE REPORT

A 76 year old patient with no significant disease and no major cardiovascular risk factors was admitted urgently to a secondary hospital for acute pulmonary oedema and cardiogenic shock. Endotracheal intubation and inotropic support were required immediately. Physical examination showed a loud mitral regurgitation murmur. Except for sinus tachycardia, the ECG was normal with no ST elevation. Chest radiography showed signs of acute pulmonary oedema.

The patient was referred to our institution. Transoesophageal echocardiography showed preserved global and regional myocardial function, mild left ventricular hypertrophy, and the rupture of an anterolateral papillary muscle associated with massive mitral regurgitation (fig 1B). Coronary angiography was normal (fig 1A). The results of laboratory tests were creatine kinase 588 IU/l (normal < 200 UK/l), troponin I 22.8 ng/ml (normal < 0.5 ng/ml), and C reactive protein 21 mg/l. The patient was haemodynamically unstable and underwent mitral valve replacement with a porcine biological prosthesis (Mosaic No. 27, Medtronic Inc, Minneapolis, Minnesota, USA). Intraoperative findings confirmed the diagnosis of papillary muscle rupture caused by a focal apical necrosis. The left ventricular lateral wall did not show any signs of ischaemia. A histopathological examination confirmed the macroscopic aspect of ischaemic papillary muscle rupture. Microscopic examination showed a large zone of cardiomyocyte coagulation. Within this necrosis, microscopy showed severe inflammatory neutrophilic leucocytosis, thick arterioles, and an extracellular deposition of amyloid fibrils that bound to Congo red and exhibited apple-green birefringence under polarised microscopy (fig 1C,D)

Six months later the patient was asymptomatic. No secondary amyloidosis was found. Serum electrophoresis, renal function, and rectal and salivary gland biopsies were normal.

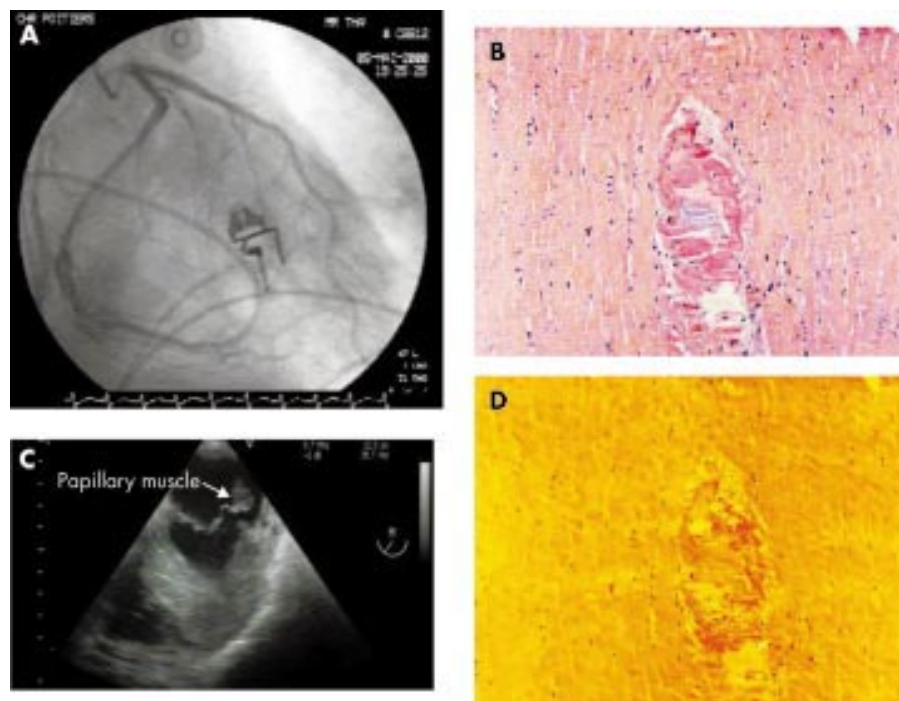


Figure 1 (A) Left coronary angiography showing the absence of epicardial stenosis in a left dominant coronary angiogram. (B) Transoesophageal echocardiography in two chamber view: note the mitral papillary muscle in the left atrium partly attached to the chordae. (C, D) Histopathological examination shows intramural deposition of amyloid substance that binds Congo red (C) and exhibits apple-green birefringence under polarised microscopy.

DISCUSSION

Causes of mitral papillary muscle rupture are usually ischaemic¹ as a complication of myocardial infarction. Non-ischaemic causes include endocarditis,² blunt chest trauma,³ and complications during invasive procedures.⁴ Few cases of “spontaneous” papillary muscle rupture have been described^{5,6} and some patients have more general disease.⁷ This is the first description of a papillary muscle rupture resulting from obstructive intramural coronary amyloidosis without any other amyloidosis location. Recently Mueller and colleagues⁸ emphasised the role of intramural coronary deposits of amyloid substances in causing ischaemic heart disease. The incidence of this clinical pathological situation seems to be very low (22 published cases). All the cases described by Mueller and colleagues were found during necropsies or after examination of explanted hearts. The clinical presentation is usually angina or heart failure. Five patients have had an acute ischaemic event but the coronary angiogram was normal. Two cases of papillary muscle rupture with no classic cause have been described previously.^{5,6} Histopathology showed focal papillary muscle necrosis, and no intramural amyloidosis was described. This new case highlights the possibility that acute papillary muscle rupture can result from an obstructive intramural amyloidosis with normal epicardial coronary angiogram and without any significant diffusion of amyloidosis disease. Vascular amyloidosis is a new cause of papillary muscle rupture and may explain the so-called “spontaneous” cases. The extent of intramural amyloid deposits can lead to large differences in the clinical presentation of this cardiac disease.

Authors' affiliations

D Coisne, P Corbi, P Sosner, Department of Cardiology, La Miletrie University Hospital, Poitiers, France

Correspondence to: Dr D Coisne, Department of Cardiology, La Miletrie University Hospital, Poitiers, France; d.coisne@chu-poitiers.fr

Accepted 17 October 2002

REFERENCES

- 1 **Iwasaki K**, Matsuo N, Hina K, *et al*. Transesophageal echocardiography for detection of mitral regurgitation due to papillary muscle rupture or dysfunction associated with acute myocardial infarction: a report of five cases. *Can J Cardiol* 2000;**16**:1273–7.
- 2 **Habib G**, Guidon C, Tricoire E, *et al*. Papillary muscle rupture caused by bacterial endocarditis: role of transesophageal echocardiography. *J Am Soc Echocardiogr* 1994;**7**:79–81.
- 3 **Cho MC**, Kim DW, Hong JM, *et al*. Left ventricular and papillary muscle rupture following blunt chest trauma. *Am J Cardiol* 1995;**76**:424–6.
- 4 **Ootaki Y**, Kozawa S, Asada T, *et al*. Rupture of the papillary muscle after percutaneous transvenous mitral commissurotomy (PTMC): a case report. *Nippon Kyobu Geka Gakkai Zasshi* 1997;**45**:1738–42.
- 5 **Morishita A**, Tsuchiya K, Hosaka S, *et al*. A case of mitral regurgitation due to partial papillary muscle rupture whose etiology was unknown. *Kyobu Geka* 1995;**48**:80–3.
- 6 **Weiss SR**, Coppola J, Gambino J, *et al*. Isolated acute papillary muscle infarction in the absence of coronary artery disease resulting in cardiogenic shock and emergent mitral valve replacement. *Cathet Cardiovasc Diagn* 1998;**43**:185–9.
- 7 **Park CW**, Shin YS, Kim SM, *et al*. Papillary muscle rupture complicating inferior myocardial infarction in a young woman with systemic lupus erythematosus and antiphospholipid syndrome. *Nephrol Dial Transplant* 1998;**13**:3202–4.
- 8 **Mueller PS**, Edwards WD, Gertz MA. Symptomatic ischemic heart disease resulting from obstructive intramural coronary amyloidosis. *Am J Med* 2000;**109**:181–8.