Case Report

Splenic rupture and haemoperitoneum in a patient with non-compaction of the left ventricular myocardium

C. L. Errando¹*, J. Tatay¹, A. Serrano-Romero², M. Gudín-Uriel², M. Revert³ and C. M. Peiró¹

¹Servicio de Anestesiología, Reanimación y Tratamiento del Dolor, ²Servicio de Cardiología and ³Servicio de Radiodiagnóstico, Consorcio Hospital General Universitario de Valencia, Avenida Tres Cruces s/n, 46014-Valencia, Spain

*Corresponding author. E-mail: c.l.errando@carloserrando.com

The anaesthetic and critical care management of blunt abdominal trauma in a patient previously diagnosed with non-compaction of the left ventricular myocardium (a rare autosomal dominant inherited disease) is reported. The management was influenced by the presence of an implanted automated internal defibrillator and treatment with anticoagulants because of the high frequency of severe arrhythmias and systemic embolism. The pathophysiology of ventricular non-compaction is reviewed briefly.

Keywords: anaesthesia, general; complications, hypovolaemic shock; complications, trauma; heart, cardiomyopathies

Accepted for publication: April 12, 2005

The spleen is the most commonly ruptured viscus after blunt abdominal trauma.¹ When splenic haematoma occurs without rupture of the visceral capsule, conservative treatment has been proposed, albeit with persisting controversy.²⁻⁴ Haemoperitoneum and haemodynamic instability are indications for urgent surgery. However, pre-existing disease or treatment may worsen the prognosis of the injuries and influence trauma management.⁵ We describe the perioperative management of a patient who suffered a traumatic splenic rupture and haemoperitoneum, and who had previously been diagnosed as having a rare and severe cardiomyopathy, non-compaction of the left ventricular myocardium (NCVM). A brief review of this rare disease is presented.

Case report

A 39-year-old man, weighing 76 kg, was admitted to the emergency department following a fall. The patient had pain and tenderness in the left hypochondrium. He was pale and sweating, but his blood pressure was 130/80 mm Hg and his heart rate was 88 beats min⁻¹. The ECG showed sinus rhythm with no specific abnormalities. A splenic rupture with intracapsular haematoma, a small hepatic tear and free intraperitoneal fluid were demonstrated using CT scanning.

Six months earlier the patient had been diagnosed as having an NCVM (also known as spongiform cardiomyopathy). There was a strong family history of cardiac problems: his mother had died suddenly at the age of 56, his father and one of his brothers had atrial fibrillation, his other brother had a cardiomyopathy and two cousins had internal automated defibrillators (IADs) implanted because of cardiomyopathy. Before the diagnosis was made, the patient suffered spontaneous paroxysmal atrial fibrillation. Under electrophysiological investigation, ventricular polymorphic tachycardia was easily induced despite concurrent treatment with amiodarone. The diagnosis was confirmed after an echocardiogram and cardiovascular MRI studies (Figs 1–3). By the time of the abdominal trauma a double-chamber IAD had been implanted and oral anticoagulant therapy (acenocumarol) had been instituted.

Initial surgical management was conservative, but 6 h after admission the patient’s general status deteriorated, with the development of severe abdominal pain, abdominal distension and a fall in blood pressure. An urgent laparotomy was requested by the surgeons.

After consultation with a cardiologist, the IAD was disconnected and external defibrillator pads were placed on the patient’s chest before anaesthesia was induced. As the coagulation profile was altered (INR 4.47) and the haemoglobin was 7.7 g litre⁻¹, two units of red blood cells and two
units of fresh frozen plasma were transfused before surgery (INR 1.80). Volume resuscitation was initiated with lactated Ringer’s solution 1000 ml and modified gelatine solution 1000 ml. After standard and invasive monitoring (radial artery cannulation and internal jugular vein cannulation with a 7F introducer), anaesthesia was induced with i.v. midazolam 0.1 mg kg\(^{-1}\) and ketamine 1 mg kg\(^{-1}\). Rocuronium was used to facilitate tracheal intubation. Anaesthesia was maintained using sevoflurane (0.75–1% end-tidal concentration) in 50% oxygen–air with controlled ventilation. An infusion of rocuronium was started, and boluses of fentanyl 100 mg were administered as required (total dose 500 μg). No significant haemodynamic alterations were observed during induction nor throughout the surgical procedure, which lasted for 120 min. The total blood loss was ~1200 ml. Splenectomy and hepatic surgical haemostasis were required. Four units of red blood cells, 4 units of fresh frozen plasma and platelets were transfused intraoperatively. After surgery, the patient was transferred to the critical care unit (CCU) and was extubated 4 h later. During the first 72 h postoperatively he needed further red cell and fresh frozen plasma transfusion but no inotropic support.

The IAD was restarted 4 h after surgery. A posteriori evaluation of the defibrillator tracing showed an automated discharge 24 h after CCU admission as a result of ventricular tachycardia. Neither clinical nor haemodynamic changes were observed at that time. After consultation with a haematologist, enoxaparin 60 mg once daily was started and oral acenocumarol was added 48 h later. The patient was discharged to the surgical ward 72 h later and discharged home 10 days after his admission to hospital.
Discussion

NCVM is a relatively unknown cardiomyopathy which consists of numerous excessively prominent ventricular trabeculations and deep intertrabecular recesses. The left ventricle is mainly affected, but the right ventricle is involved in 50% of cases. The disease originates during embryogenesis, in weeks 5–8 of fetal life, when the ventricular myocardium compacts to form the endocardial capillaries and the coronary circulation develops. The process of compaction progresses from epicardium to endocardium and from the base of the heart towards the apex. In NCVM the procedure of compaction is altered.

The prevalence of the disease remains unknown but has been estimated at 0.05–0.24%. In a series of 37,555 consecutive echocardiographic studies, Ritter and colleagues found NCVM in 17 adult patients. Although the disease seems to be very rare, some authors point out the possibility of confusion with other cardiomyopathies and the existence of subtle degrees of non-compaction. To date, three genetic forms of the disease have been described. Two forms are related to a mutation in chromosome 18, one of which is associated with other congenital heart defects presenting in childhood. The second is an isolated autosomal dominant form. Sometimes this type is related to facial dysmorphism. The third type of NCVM is linked to a mutation in the X chromosome, occurs in males and may be associated with skeletal myopathies.

Adult forms of NCVM show non-specific clinical manifestations such as heart failure, primarily from left ventricular dysfunction, chronic myocardial ischaemia, supraventricular tachyarrhythmias, conduction defects and malignant ventricular tachyarrhythmias. The prognosis of the disease is determined by its complications; the death rate and heart transplantation frequency can be as high as 50%. Despite this, the echocardiographic features have correlated well with the pathological findings in post-mortem or post-transplant specimens. Our patient showed both left and right ventricle trabeculations and blood flow through recesses, demonstrated by cardiac MRI (Figs 1–3).

Recent trends in the conservative management of blunt visceral abdominal trauma are mainly based on CT scan findings. However, the risk of delayed rupture should be borne in mind. Treatment with anticoagulants could have worsened the consequences of the abdominal trauma in this patient. In this setting, reversal of the anticoagulant is mandatory.

We could find no reports of anaesthesia in patients with NCVM to guide our anaesthetic management. The management of patients with pacemakers and internal defibrillators has recently been reviewed, and we were advised to disable the IAD during surgery. We had external defibrillator pads in place because of the high incidence of life-threatening ventricular tachyarrhythmias (as illustrated by the operation of the IAD postoperatively). For induction of anaesthesia after volume resuscitation, we chose a combination of low-dose midazolam and ketamine as this offered the lowest haemodynamic deterioration when compared with other intravenous anaesthetics (including using either ketamine or midazolam alone) in an experimental model of acute hypovolaemia. Carefully titrated doses of etomidate or thiopental are alternatives, although we would suggest avoiding propofol during haemorrhage or hypovolaemia. On the other hand, the classic arrhythmogenic (catecholamine-related) central properties of ketamine are usually balanced by other known arrhythmogenic properties (mainly in the cardiac conduction system or cardiac muscle cells) of the drug by, among other effects, decreasing excitability or increasing the relative refractory period. The overall effect in a patient with an arrhythmogenic cardiomyopathy is unknown. The choice of the neuromuscular blocking drug was directed by the emergency situation. Suxamethonium was avoided because it has arrhythmogenic properties, while rocuronium has no harmful cardiac effects and is used in our practice during rapid-sequence induction of anaesthesia.

As described in our case report, anticoagulation should be started as soon as possible because of the high probability of thromboembolic events in non-compaction.

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