Non-operative management of a splenic laceration in a patient with the Proteus syndrome

W Ceelen, J De Waele, M Kunnen, B de Hemptinne

Abstract
An adult patient with the Proteus syndrome sustained a grade III splenic laceration after falling off a horse. Clinical features of this rare disorder include subcutaneous and visceral hamartomatous tumours. The patient also suffered from chronic intravascular coagulation associated with extensive haemangiomatosis (Kasabach-Merritt syndrome). Considering the visceral anomalies and abnormal coagulation, a non-operative approach was preferred despite considerable transfusion requirement, and the patient successfully underwent embolisation of the splenic artery. This is the first reported case of splenic injury in a patient with Proteus syndrome.

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Keywords: splenic injury; embolisation; Kasabach-Merritt syndrome; Proteus syndrome.

The spleen is the most frequently injured intra-abdominal organ following blunt abdominal trauma. In recent years, a less aggressive approach toward splenic laceration is accepted as being as safe as urgent splenectomy in selected cases. Attempts have been made to identify and test criteria for safe non-surgical management of splenic injuries. We present an adult patient with the Proteus syndrome and associated coagulation abnormalities due to large subcutaneous hamartomas (Kasabach-Merritt syndrome) who sustained a laceration of the spleen following blunt injury.

Case report
A 23 year old male patient with the Proteus syndrome since childhood fell of a horse. He did not immediately seek medical attention. The morning after, however, he woke up with severe upper abdominal pain and fainting, and was admitted to the emergency department.
On admission, blood pressure was 115/65 mm Hg and pulse rate was 76 beats/min. The manifestations of the Proteus syndrome in this patient included exostoses of the facial bones, diffuse subcutaneous and intra-abdominal hamartomas, and macrodactyly of both feet. The patient looked anaemic. Clinical examination revealed hepatosplenomegaly; there was severe pain on palpation of the left and right upper abdomen. There was no rebound tenderness or guarding.

Laboratory analyses revealed a haemoglobin level of 14.2 g/dl, abnormally low fibrinogen (118 mg/dl) and prothrombin time (47%); and raised indirect bilirubin (2.45 mg/dl) and lactate dehydrogenase (510 U/litre). The patient was known to have a chronic intravascular haemolysis.

Ultrasound of the abdomen revealed a large, irregular liver; the spleen was also enlarged and contained a hypereffective zone of 4 cm diameter at the lower pole; intraperitoneal fluid was also noted. A computerised tomography (CT) scan of the abdomen showed an AAST grade III splenic injury at the lower pole and haemoperitoneum (fig 1). A nonoperative approach was preferred and the patient was admitted to the ICU. Initially, the packed cell volume remained stable around 30%. After 24 hours, however, it suddenly fell to 20% and did not increase despite a massive transfusion of packed red cells and fresh frozen plasma. Angiography of the splenic artery was then performed, and showed a contrast extravasation at the hilus and several cut-off images of smaller vessels inside the parenchyma (fig 2). Subsequently, embolisation of the splenic artery was performed using lipiodol and Histoacryl (TM); a more selective embolisation was not possible because of the diffuse vascular injury. Following this procedure, the transfusion requirement immediately decreased; the patient’s condition gradually improved and he was ultimately able to leave the hospital 10 days after admission.

Discussion

The Proteus syndrome is a congenital hamartomatous disorder characterised by partial gigantism, macrodactyly, subcutaneous tumours, and exostoses of the skull bones. The condition has a varying phenotype, and was named after the ancient Greek god Proteus, who could take various shapes to mislead enemies. First described by Wiedemann in 1983, approximately 60 cases have been described up to now.1,2

It is now generally accepted that Joseph Merrick, the famous Elephant Man, probably suffered from Proteus syndrome.3 Most cases are sporadic; in only two cases has transmission of the disease been suspected.3,4 A somatic mutation model has therefore been proposed, and there appears to be disturbed production of growth factors and growth factor binding proteins in the affected tissues.5

The presence of vascular tumours is one of the more constant features of Proteus syndrome; most of these tumours have been shown to be lymphatic hamartomas by immunohistological staining.3 The patient we describe suffered from extensive hamartomas of the abdominal wall; he was also known to have widespread hamartomata in the abdominal viscera. Moreover, the extent of vascular malformation in this patient gave rise to a chronic consumptive coagulopathy, known as the Kasabach-Merritt syndrome.

Non-operative management of splenic lacerations remains a matter of debate in trauma surgery. Some clinicians have advocated conservative treatment and have attempted to define criteria identifying patients in whom observation is the only necessary treatment. These criteria include haemodynamic stability, age of less than 55 years, absence of or minimal peritoneal signs, CT scan injury grade less than III (AAST), and absence of injury to other intra-abdominal organs.5,12

Other advise early laparotomy in order to maximise splenic salvage, rather than removing the spleen after failure of non-surgical treatment,
An unusual cause of rib fracture following a road traffic accident

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Abstract

A case is presented which is thought to be the first described example of rib fracture occurring as a result of airbag inflation. It would appear that the propellant cartridge came loose during deployment to form a missile, striking the patient on his chest and fracturing a rib.

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Keywords: airbag; airbag related injury; rib fracture.

Case report

The patient, a 73 year old asthmatic, was driving a 1994 Ford Mondeo 1.8 LX estate when he was involved in a low velocity frontal collision with another vehicle. The patient was wearing a seatbelt and the impact caused his airbag to inflate. Immediately after the impact, the patient felt a sharp blow on the left side of his chest. He received no other injuries. He was admitted to hospital complaining of chest pain, shortness of breath, and difficulty in breathing.

Examination of his chest revealed reduced expansion bilaterally, basal crepitations, and focal tenderness over his left anterolateral chest wall. Breath sounds were vesicular and no wheeze was heard. The jugular venous pulse was not raised and the trachea was central. Peak expiratory flow rate was 180 litres/min, compared with a normal value of 300 litres/min for this patient. An ECG showed no acute ischaemic changes and arterial blood gases on air were normal. A chest x-ray was taken of the patient which showed a fracture of the left ninth rib laterally (fig 1). The patient was treated with analgesia, and admitted overnight for observation. He made an uneventful recovery and was discharged the following day.

Close examination of the clothes that this patient was wearing at the time of the crash revealed a hole approximately 1.5 cm in diameter surrounded by a chemical residue in a position overlying the site of the fracture (figs 2 and 3). Unfortunately it was not possible for chemical analysis to be performed on this residue. The patient reported that the coat was undamaged before the accident.

Discussion

Many studies have shown the benefits of airbags as a passenger restraint system, particularly in conjunction with seatbelts, where their use has been estimated to reduce fatalities by


9 Smith JS, Wengrovitz MA, Delong BS. Prospective validation of criteria, including age, for safe nonsurgical management of the ruptured spleen. J Trauma 1992;33:363–8.


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