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Spontaneous non-traumatic splenic artery aneurysm rupture: a case report and review of the literature

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Abstract. – The current case report is about spontaneous non-traumatic rupture of a splenic artery aneurysm (SAA) in a 53-year-old woman with no particular medical history. An emergent laparotomy with splenectomy was required, unfortunately without success as the patient died.

SAA is the most common visceral artery aneurysm. Most of SAA remain asymptomatic and are discovered incidentally on imaging. The overall risk of rupture increases with the size of SAA, especially when above 2 cm. Initial presentation of SAA has been associated with acute rupture and hemodynamic instability leading to substantial perioperative morbidity and mortality.

Key Words:
Splenic artery, Aneurysm, Rupture, Laparotomy.

Introduction

Splenic artery aneurysm (SAA) was first described in 1770 by Beaussier (Sur un anévrisme de l'artère splénique: dont les parois se sont ossifiées. J Med Clin Pharmacol 1770; 32: 157), but surgical repair was not reported until 1940[1,2]. The splenic artery is considered aneurysmal when its size reaches > 1 cm in diameter[1]. SAA is the most common visceral artery aneurysm[3]. Historical studies described an occurrence of 10% in routine post-mortem examination of elderly asymptomatic patients[4]. Another cadaveric radiological study with barium injections observed SAA in 7.5% of cases[5]. In a retrospective study including 100 patients with SAA, the mean aneurysm diameter was 2.1 cm (with cases up to 30 cm) and a majority (78%) were located in the distal third of the splenic artery and were saccular[3]. Furthermore, the female to male ratio is 4:1[3,6,7]. SAA has an indolent course with non-specific abdominal symptoms (17%)[3]. Over 2cm, the risk of rupture increases significantly and is embedded with a high mortality reaching 66%[1,8]. Although patients suffering from SAA should be monitored, no consensus was established neither on the modalities (computed tomography, magnetic resonance angiography, ultrasonography, and digital subtraction angiography) nor on the timing[9,10].

Nevertheless, SAA larger than 2 cm should be considered for repair, especially in women in childbearing age or pregnant, in whom occurrence and mortality seem to be higher[3,11,12].

The current report describes a spontaneous non-traumatic rupture of SAA in a 53-year-old woman with no particular medical history.

Clinical Presentation

A 53-year-old woman without any medical history or medications presented sudden acute diffuse abdominal pain and vomiting after getting up in the morning. A few minutes later, cardiorespiratory arrest occurred at home. The resuscitation was promptly initiated by her husband and continued by the pre-hospital team according to the advanced cardiac life support (ACLS) guidelines[13]. The pre-hospital return of spontaneous circulation (ROSC) was estimated at 10 min.

In the Emergency Room, the patient presented a severe hemodynamic instability requiring high doses of vasopressors combined with high volume expansion and administration of erythrocytic concentrates (O-negative). In parallel, the coagulation pathways were optimized using protamine, coagulation factors and anti-fibrinolytic drugs. The initial arterial blood gas revealed a severe metabolic acidosis (pH = 6.9 and lactates 17 mmol/l) as well as lowered hemoglobin level (70 g/L). The focused assessment with sonography for trauma (FAST) revealed a significant amount of perihepatic, perisplenic and pelvic intra-abdo-

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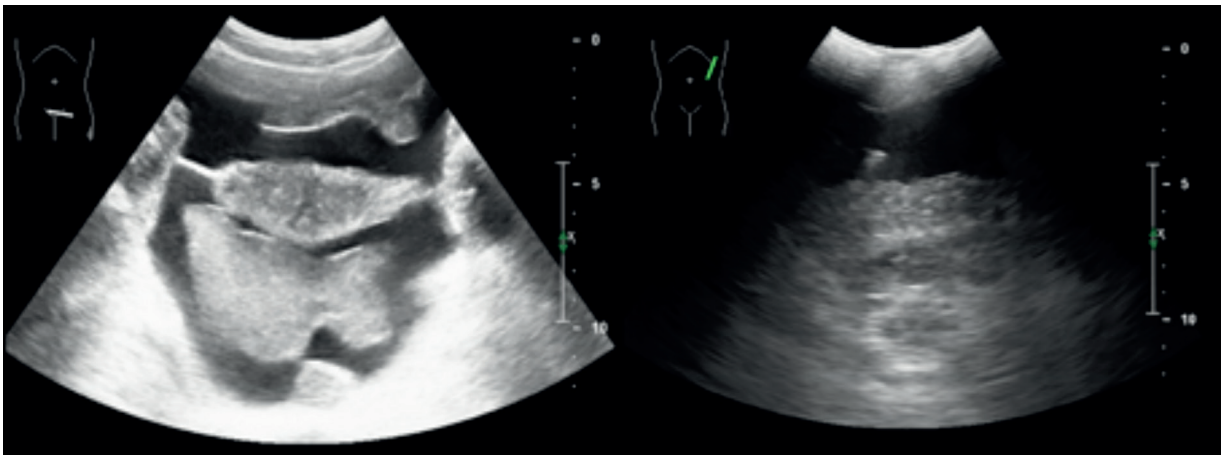

Figure 1. Focused assessment with sonography for trauma (FAST) revealed significant amount of pelvic intra-abdominal ***(a)*** and perisplenic ***(b)*** free fluid.

minal free fluid (Figure 1). An ECG identified a right branch block image with ST segment elevation in V1, V2, V3, leading to the differential diagnosis of an anteroseptal STEMI or a syndrome of Brugada. Transesophageal echocardiography performed by a cardiologist was readily available and could exclude massive pulmonary embolism or aortic dissection.

Due to hemodynamic instability and the presence of intra-abdominal fluid, an emergency exploratory laparotomy was decided without prior CT scan evaluation. During the transport to the OR, the patient presented a new cardiorespiratory arrest and the external cardiac massage was resumed.

During the entire operation, the patient received external cardiac massage without regaining her own rhythm. The exploratory laparotomy revealed a large quantity of blood (3 liters). As a rupture of an aortic aneurysm was suspected, the retroperitoneum was dissected and the aorta exposed with the help of a senior vascular surgeon. The aorta was intact without aneurysm. No clamping maneuver was performed. The abdominal cavity was packed and the exploration performed sequentially. The bleeding appeared to originate from the left hypochondrium without clear origin. A splenectomy was performed but did not control the hemorrhage. The omental bursa and the rest of the abdominal cavity were revised without finding the source of bleeding. The hemorrhage could not be controlled. Thus, the resuscitation was stopped after 1 h without return to a spontaneous cardiac rhythm. The patient died in the operating room. The family accepted medical autopsy and post-mortem examination revealed a spleen of 85 g without parenchymatous lesion. At the level of the splenic hilum, a ruptured cystic formation measuring 6 x 4 x 3 cm was observed, corresponding to a ruptured aneurysm (Figure 2). Histological analysis confirmed the presence of an arterial wall with aneurysmal dilatation containing severe degenerative changes and architectural disorders. The intima and the media were hardly distinguishable and the internal elastic membrane was become fragmented to finally disappear (Figures 3 and 4). There was no significant inflammatory component. This clinical picture opened the differential diagnosis between alterations secondary to a possible atherosclerosis and a fibromuscular dysplasia. However, forensic autopsy revealed nothing special, particularly in terms of vascular status.

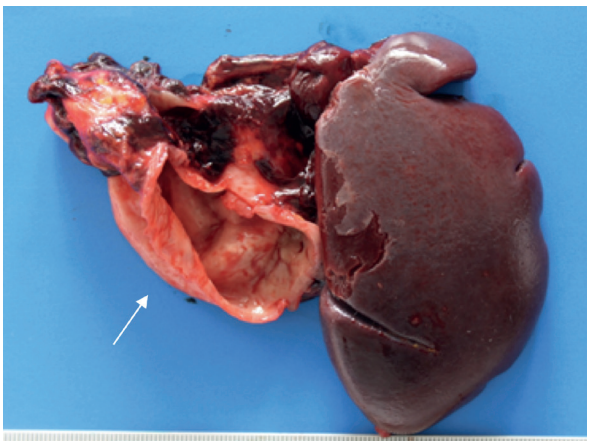

Figure 2. Splenic pathology: ruptured aneurysm of the splenic artery measuring 6 x 4 x 3 cm (white arrow).

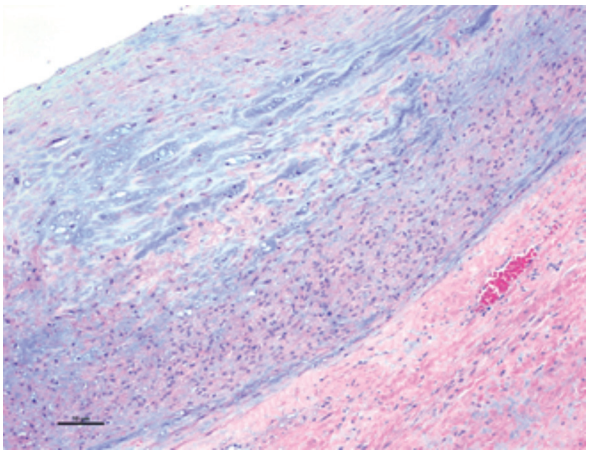

Figure 3. Aneurysm wall of the splenic artery with major degenerative changes. The intima and the media are hardly distinguishable and the muscular layers are partly replaced by fibro-mucoid plaques. Hematoxylin and eosin (HE) staining (100x).

Discussion

This case brings an old deadly foe back into light. The abrupt onset of non-traumatic severe hemodynamic shock in a patient without medical history is a rare but life threatening event[7]. Whereas the indication for surgery may seem obvious, the localization of the bleeding can be challenging once an aortic aneurysm has been excluded. Moreover, the systematic examination of the abdominal cavity can be difficult to perform due to complications induced by the severe shock such as coagulopathy or cardiac arrest. Once a ruptured aortic aneurysm is excluded along with a bleeding from a pathological spleen, SAA or other rare visceral aneurysms should be considered. The literature about management of acute rupture of SAA is scarce and disseminated over several small studies during a broad period of time[1]. Thus, no clear overviews, guidelines, or management recommendations are available. In the present case report, the ruptured distal SAA was located in the splenic hilum region, requiring emergent laparotomy with splenectomy, even if during the procedure the diagnosis was not established. Before the era of modern imaging, SAA was often discovered during an acute rupture[7]. The widespread use of computed tomography has changed the clinical presentation of SAA due to the increased detection of asymptomatic visceral artery aneurysms[14]. A surgical or endovascular management should be considered for symptomatic aneurysms, for aneurysms ≥ 2 cm in size, or for any SAA in female patients who are pregnant or in childbearing age[8]. A conservative approach, such as beta-blocking treatment, should be proposed to selected patients only, as a recent study showed an increased late mortality[1]. Whereas numerous therapeutic strategies are known to treat SAA, no definitive consensus has been reached. Endovascular therapy, open surgery or laparoscopic approaches are still valid therapies[3,15]. Open ligation, transcatheter embolization or even laparoscopic aneurysmectomy with end-to-end anastomosis have been described for elective cases[8,15]. A recent systematic review showed that endovascular repair of SAA has better short-term outcomes compared to open approaches, including significantly lower perioperative mortality[1]. Generally, aneurysmectomy and reconstruction is an option for proximal SAA, while distal SAA requires aneurysmectomy with splenectomy, and sometimes even distal pancreatectomy if the aneurysm is too closely adherent to the tail of pancreas[1,16]. As far as possible, splenectomy should be avoided to prevent post-splenectomy thrombocytosis and potential immunodeficiency[17].

The pathophysiology of SAA is not well known. A combination of medial hyperplasia and fragmentation of the elastic lamina, especially in patients with portal hypertension has been postulated[7,18,19]. Other possible mechanisms are fibromuscular dysplasia, polyarteritis nodosa, α-1 antitrypsin deficiency, systemic hypertension and infective factors[7,8,20,21]. The reason why SAA predominates in women is not clear, but hormonal contribution was

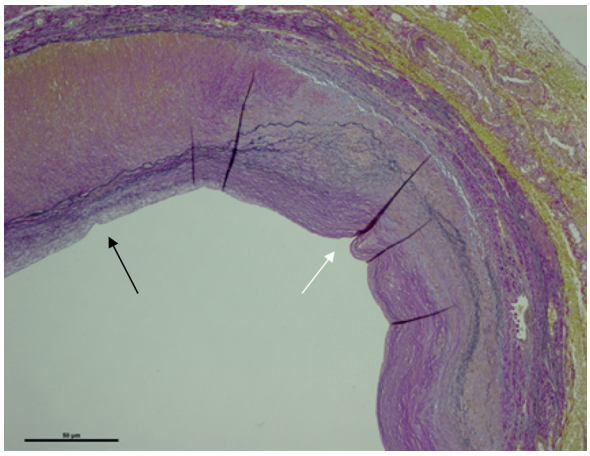

Figure 4. Transition between the normal arterial wall (black arrow) and the aneurysmal wall (white arrow) of the splenic artery. Progressively in the direction of the aneurysmal wall, the internal elastic membrane fragments and finally disappears. The muscular layer of the media disrupts and is replaced by fibrosis intermixed with mucoid substance. Van Gieson-elastin staining (40x).

suggested[20]. While a significant number of splenic artery aneurysms show calcifications and other pathologic features as observed in atherosclerosis, these changes appear secondary to the arterial degeneration rather than atherosclerosis being the primary underlying etiology. In the present case, the pathological examination could not differentiate between alterations secondary to atherosclerosis with fibromuscular dysplasia.

Learning Points

A spontaneous SAA rupture belongs to the differential diagnosis in patients with history of abdominal pain and hemodynamic shock.

A salvage laparotomy should be performed, with aneurysm resection and splenectomy depending on SAA location.

The mortality of SAA rupture is high, and the precise etiology remains unclear.

Conflict of Interest

The Authors declare that they have no conflict of interest.

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