

CASE REPORT

Open Access



# Spontaneous rupture of a large splenic artery aneurysm in a 59-year-old male patient with pemphigus vulgaris: a case report

Ahmad Hosseinzadeh<sup>1</sup>, Reza Shahriarad<sup>1,2\*</sup> , Vahid Asgharzadeh Majdazar<sup>3</sup>, Mohammad Moeini Farsani<sup>3</sup> and Seyed Mohammad Kazem Tadayon<sup>4</sup>

## Abstract

**Background:** There is currently no information on the anatomical risk factors for splenic artery aneurysm rupture, specifically the location or size of the lesion; therefore, reporting this entity to obtain data and ultimately reduce morbidity and mortality is essential. Here we report a case of a male patient with spontaneous rupture of a large splenic artery aneurysm presenting with abdominal pain.

**Case presentation:** A 59-year middle-eastern male, with known pemphigus vulgaris presented with a chief complaint of headache and syncope, followed by abdominal pain along with severe metabolic acidosis. A contrast-enhanced computed tomography scan of the abdomen and pelvic showed a splenic artery aneurysm of 33 × 30 mm with a 150 × 90 mm hematoma formation around the aneurysm site. The patient underwent an operation and splenectomy, with confirmation of the diagnosis of ruptured splenic artery aneurysm.

**Conclusion:** It is essential to consider splenic aneurysm rupture as a second-line differential diagnosis, especially among patients with comorbid diseases, as this can lead to timely and appropriate lifesaving intervention.

**Keywords:** Splenectomy, Aneurysm, Ruptured, Splenic artery, Vascular, Artery, Pemphigus vulgaris

## Introduction

Splenic artery aneurysms (SAA) are the third most common type of arterial aneurysm, with diameters ranging from 0.6 to 30 cm. The majority of patients show no signs or symptoms [1]. The exact cause of a splenic artery aneurysm is uncertain, while its combination with the possibility of rupture, can result in a clinical picture ranging from nonspecific abdominal symptoms (making prerupture diagnosis difficult) to a more dramatic intraperitoneal hemorrhage with hypovolemic shock and high morbidity and mortality, making it a retrospective

diagnosis [2]. There is currently no information on the anatomical risk factors for SAA rupture, especially the location or size of the lesion; therefore, reporting this entity to obtain data for decreasing mortality and morbidity is essential. Here we report a male patient with the cutaneous disease of pemphigus vulgaris, who developed a spontaneous rupture of a large SAA, presenting with abdominal pain.

## Case report

A 59-year middle-eastern male, with known pemphigus vulgaris treated with a daily dose of 5 mg prednisolone, presented to the emergency department with a chief complaint of headache and syncope 3 days before presentation, with subsequent onset of severe abdominal pain. On admission, the patient's Glasgow Coma Scale level was 13, he was tachycardic (rate: 130) and had severe

\*Correspondence: [R.shahriari1995@gmail.com](mailto:R.shahriari1995@gmail.com)

<sup>1</sup>Thoracic and Vascular Surgery Research Center, Shiraz University of Medical Science, Shiraz, Iran  
Full list of author information is available at the end of the article



© The Author(s) 2022. **Open Access** This article is licensed under a Creative Commons Attribution 4.0 International License, which permits use, sharing, adaptation, distribution and reproduction in any medium or format, as long as you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons licence, and indicate if changes were made. The images or other third party material in this article are included in the article's Creative Commons licence, unless indicated otherwise in a credit line to the material. If material is not included in the article's Creative Commons licence and your intended use is not permitted by statutory regulation or exceeds the permitted use, you will need to obtain permission directly from the copyright holder. To view a copy of this licence, visit <http://creativecommons.org/licenses/by/4.0/>. The Creative Commons Public Domain Dedication waiver (<http://creativecommons.org/publicdomain/zero/1.0/>) applies to the data made available in this article, unless otherwise stated in a credit line to the data.

epigastric pain, along with left upper quadrant pain and tenderness, without radiation or association with nausea and vomiting. Arterial blood gas evaluation demonstrated severe metabolic acidosis. Electrocardiography only showed premature ventricular complexes. Abdominal pelvic sonography demonstrated free fluid in the upper pole of the left kidney and around the spleen, without stasis.

Initial laboratory data demonstrated white blood cell levels of 18.8 (neutrophil 81.4% and lymphocyte of 9.2%), hemoglobin of 7.7 g/dL, platelet count of  $179 \times 10^3/\mu\text{L}$ , creatinine of 1.9  $\mu\text{mol/L}$ , and blood urease nitrate (BUN) of 30 mmol/L. Initial arterial blood gas had a pH of 7.19 with PaCO<sub>2</sub> of 44.4 mmHg, PaO<sub>2</sub> of 22.6 mmHg, HCO<sub>3</sub> of 16.6 mmol/L, and base excess of -11.1, which then improved to PpH 7.36, PaCO<sub>2</sub> 31.7 mmHg, PaO<sub>2</sub> 57 mmHg, HCO<sub>3</sub> 17.7 mmol/L, and base excess -6.4 after fluid resuscitation.

Brain computed tomography (CT) and echocardiography were requested for the patient, which were unremarkable. A contrast-enhanced computed tomography scan of the abdomen and pelvis showed a splenic artery aneurysm of 33 × 30 mm with 150 × 90 mm hematoma formation around the aneurysm site (Fig. 1). Based on the patient's condition, he was immediately transferred to the operating room, where a diagnosis of ruptured splenic artery aneurysm was made.



**Fig. 1** Computed tomography scan of a 59-year-old man with a ruptured splenic aneurysm

An exploratory laparotomy was performed, and a 1200 cc blood clot in the abdominal cavity was observed, along with a massive retroperitoneal hematoma that extended to the pelvic cavity, and a hematoma at the meso of the transverse colon. Initially, the lesser sac was explored to gain proximal and distal control, but owing to lack of access to the aneurysm, which was at the middle part of the splenic artery, left medial visceral rotation was performed. The spleen was mobilized, an aneurysm sac was detected and ligated, and a splenectomy was performed (Fig. 2). A Nelaton drain was also inserted.

The patient had no postoperative complications: the drain was removed on the 7th day, and thrombocytosis was treated with aspirin. Pneumovax was injected in the following days. During his 1-week follow-up, he had achieved full recovery and had no sign of any complications and normal laboratory data. He was under routine follow-up and steroids were tapered and discontinued. He also had no complications or cutaneous manifestations.

**Discussion**

Ruptured SAAs are an uncommon cause of hemorrhagic shock, but the splenic artery accounts for 60% of visceral aneurysms [3]. SAA has been most frequently reported in females, with a 4:1 female-to-male ratio [3–5], although rupture may occur more frequently in men [6]. The majority of SAAs (> 90%) are asymptomatic and are discovered incidentally during imaging studies [7]. SAAs



**Fig. 2** Ruptured aneurysm with hematoma

are also more common after the sixth decade of life, with 80% of cases occurring in patients over 50 years of age [1], similar to our case. In contrast to pseudoaneurysms, typically associated with pancreatitis and iatrogenic instrumentation, true SAA retains all three layers of the blood vessel but is thinned out.

Patients are often asymptomatic, and when symptoms are present, they are often vague and varied (17%) [4], making a prerule diagnosis challenging, especially when the aneurysm is tiny. When an aneurysm ruptures, it can cause a dramatic hypotensive shock with a high mortality rate, typically resulting in death outside of the hospital or a rare double-rupture occurrence. The interval between these two episodes can be as brief as a few hours to many days [8–10], providing a window for life-saving action. If SAA rupture is still not recognized in the diagnosis, this can cause confusion in the clinical picture. The rupture occurs in nearby organs such as the stomach, pancreas, or colon in 30% of cases [11, 12]. SAAs generate considerable blood loss after rupture, with hemodynamic instability appearing in 6–96 hours, affording time for repair if detected. Non-pregnant patients have a mortality rate of 10–36%. [13, 14]; however, for pregnant women and those with pre-existing portal hypertension, this number doubles [14]. Wiener *et al.* [15] proposed an approach regarding the management of SAA in pregnancy, for which, in asymptomatic cases with an SAA of less than 2 cm, conservative surveillance can be applied. Planned cesarean delivery at 34–36 weeks gestation followed by elective SAA embolization will evade extreme premature delivery as well as splenectomy and radiation hazard during pregnancy. However, since SAA has been associated with high mortality and rapid progression in pregnancy [16], it is vital to achieve diagnosis and treatment as soon as possible.

The condition is largely diagnosed on contrast CT performed for abdominal pain, and is seen as free fluid around the spleen or other viscera and as leaked contrast media or contrast-enhanced hematoma. As part of the workup for abdominal pain, a ruptured SAA with intra-abdominal hemorrhage may be discovered. Free fluid around the spleen and other viscera is usually detected by CT, and contrast media leakage or contrast-enhanced hematoma may also be present. Because the majority of aneurysms develop late in pregnancy, placental abruption is the most common misdiagnosis, and it is usually only corrected during exploration, and complex diagnostic facilities are not readily available. Due to the double rupture phenomenon, aneurysm rupture can be difficult to diagnose at first. Rupture into the lesser omental can temporarily tamponade bleeding and cause acute abdominal pain and hypotension. As the bleeding continues, the lesser sac ruptures, releasing the tamponade and

resulting in massive abdominal bleeding and cardiovascular collapse [17].

The treatment of a ruptured SAA necessitates knowledge and an aggressive surgical approach. There are no well-established guidelines because this is a rare condition. Regardless of symptoms or size, elective repair is preferred during pregnancy, while planning for pregnancy, and in liver transplant candidates. Regardless of the size, intervention is recommended in symptomatic aneurysms. Other SAAs without symptoms can be effectively monitored with serial imaging [18], and all aneurysms larger than 2 cm should be treated, regardless of symptoms. The procedures described are aneurysmectomy with splenectomy or left splenopancreatectomy, ligation of the proximal and distal splenic arteries, and aneurysmectomy for the distal, mid, and proximal third SAAs, respectively [19, 20]. Although splenic preservation is desirable, it is difficult to achieve in an emergency setting with a ruptured SAA [20]. For pseudoaneurysms and unruptured true aneurysms, angiography and embolization have been described [19].

In general, the use of endovascular interventions in the form of coil embolization or stent grafting has its place in SAA repair, and in many centers, this is considered the primary approach [21, 22]. However, in the case of our patient, due to the instability of the patient and the lack of an endovascular setting in the center, open surgery was preferred. The choice of surgical method depends on the general approaches of the center, access to various blocking agents equipment, access to staff at all hours of the day and night and in emergencies, and the anatomical characteristics of the splenic artery, including the degree of tortuosity of the artery, and also access to the artery through devices available are all important factors in decision making.

The current case is even more intriguing because the patient had pemphigus vulgaris and was on steroid therapy. Although vessels are rarely involved in pemphigus vulgaris, rheumatologic disorders have been linked to vasculitis, and several cases have been reported in which aneurysms in the splenic artery have developed as a result of vasculitis involvement [23, 24]. Aydın *et al.* also reported the rupture of a splenic aneurysm in a male patient with sarcoidosis [25]. The most common cause of death in Behcet's disease is an arterial aneurysmal rupture [26]. Akiyama *et al.* [27] also proposed that the long-term use of corticosteroids can weaken the arterial walls, and subsequently increase the risk of rupture. However, there is not enough evidence to support the increased risk of an aneurysm in rheumatological diseases. A spontaneous splenic artery dissection rupture may be diagnosed only in the presence of relevant symptoms, such as a limited retroperitoneal hematoma visible, for example,

with a contrast-enhanced CT scan followed by a more specific technique, such as selected angiography, given the extreme rarity of this disease, and the one-of-a-kind case with successful treatment. Further reporting of such entities is required, along with a detailed pathophysiological evaluation to obtain the necessary information to draw a clear conclusion.

## Conclusion

Aneurysms of the splenic artery are a rare type of aneurysm that are often asymptomatic. It is essential to consider splenic aneurysm rupture as a second-line differential diagnosis, as this can lead to timely and appropriate lifesaving intervention.

## Abbreviations

BUN: Blood urease nitrate; CT: Computed tomography; SAA: Splenic artery aneurysm.

## Acknowledgements

None to declare.

## Author contributions

AH made the diagnosis while VA, MM, and SMKT participated in the patients treatment course. RS drafted the manuscript. All authors read and approved the final manuscript.

## Funding

No financial support was received for this report.

## Availability of data and materials

All data regarding this study has been reported in the manuscript. Please contact the corresponding author if you are interested in any further information.

## Declarations

### Ethics approval and consent to participate

Written informed consent was obtained from the patient in our study. The purpose of this research was completely explained to the patients and they were assured that their information will be kept confidential by the researcher.

### Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

### Competing interests

The authors declare that they have no competing interests.

### Author details

<sup>1</sup>Thoracic and Vascular Surgery Research Center, Shiraz University of Medical Science, Shiraz, Iran. <sup>2</sup>School of medicine, Shiraz University of Medical Science, Shiraz, Iran. <sup>3</sup>Department of Surgery, Shiraz University of Medical Sciences, Shiraz, Iran. <sup>4</sup>Colorectal Research Center, Shiraz University of Medical Sciences, Shiraz, Iran.

Received: 14 July 2022 Accepted: 21 September 2022

Published online: 21 October 2022

## References

1. Abbas MA, Stone WM, Fowl RJ, Glociczki P, Oldenburg WA, Pairolero PC, et al. Splenic artery aneurysms: two decades experience at Mayo clinic. *Ann Vasc Surg.* 2002;16(4):442–9.
2. Beaussier M. Sur un aneurisme de l'artere splinique don't les parois se sont ossifies. *J Med Toulouse.* 1770;32:157–62.
3. Berceci SA. Hepatic and splenic artery aneurysms. In: *Seminars in vascular surgery.* Philadelphia: Elsevier; 2005.
4. Trastek VF, Pairolero PC, Joyce JW, Hollier LH, Bernatz PE. Splenic artery aneurysms. *Surgery.* 1982;91(6):694–9.
5. Pulli R, Dorigo W, Troisi N, Pratesi G, Innocenti AA, Pratesi C. Surgical treatment of visceral artery aneurysms: a 25-year experience. *J Vasc Surg.* 2008;48(2):334–42.
6. Ierardi AM, Petrillo M, Bacuzzi A, Floridi C, Dionigi G, Piffaretti G, et al. Endovascular retreatment of a splenic artery aneurysm refilled by colateral branches of the left gastric artery: a case report. *J Med Case Rep.* 2014;8(1):436.
7. Tani R, Hori T, Yamamoto H, Harada H, Yamamoto M, Yamada M, et al. Severely calcified true aneurysm: a thought-provoking case of solitary origin and postoperative management. *Am J Case Rep.* 2019;20:620–7.
8. Deshpande AA, Kulkarni VM, Rege S, Dalvi AN, Hardikar JV. Ruptured true aneurysm of the splenic artery: an unusual cause of haemoperitoneum. *J Postgrad Med.* 2000;46(3):191–2.
9. Davis T, Minardi J, Knight J, Larrabee H, Schaefer G. Ruptured splenic artery aneurysm: rare cause of shock diagnosed with bedside ultrasound. *West J Emerg Med.* 2015;16(5):762–5.
10. Oakley E, Ho JD, Johnson V, Vancamp J, Melson T, Hick JL. Splenic artery aneurysm: an important cause of hemoperitoneum and shock. *J Emerg Med.* 2014;46(3):e65–7.
11. Kaszyński M, Hasiura R. Splenic artery aneurysm—case report. *Med Sci Monit.* 1999;5(6):CS1213–5.
12. Ek ET, Moulton CA, Mackay S. Catastrophic rectal bleeding from a ruptured splenic artery aneurysm. *ANZ J Surg.* 2003;73(5):361–4.
13. Abdulrahman A, Shabkah A, Hassanain M, Aljiffry M. Ruptured spontaneous splenic artery aneurysm: a case report and review of the literature. *Int J Surg Case Rep.* 2014;5(10):754–7.
14. Jackson HT, Diaconu SC, Maluso PJ, Abell B, Lee J. Ruptured splenic artery aneurysms and the use of an adapted fast protocol in reproductive age women with hemodynamic collapse: case series. *Case Rep Emerg Med.* 2014. <https://doi.org/10.1155/2014/454923>.
15. Wiener Y, Tomashev R, Neeman O, Itzhakov Z, Heldenberg E, Melcer Y, et al. Splenic artery aneurysms during pregnancy: an obstetric nightmare. *Eur J Obstet Gynecol Reprod Biol.* 2019;237:121–5.
16. Aung YY, Berry C, Jayaram PR, Woon EV. Splenic artery aneurysm in pregnancy: a systematic review. *Int J Gynaecol Obstet.* 2022. <https://doi.org/10.1002/ijgo.14278>.
17. Popham P, Buettner A. Arterial aneurysms of the lienorenal axis during pregnancy. *Int J Obstet Anesth.* 2003;12(2):117–9.
18. Tessier DJ, Stone WM, Fowl RJ, Abbas MA, Andrews JC, Bower TC, et al. Clinical features and management of splenic artery pseudoaneurysm: case series and cumulative review of literature. *J Vasc Surg.* 2003;38(5):969–74.
19. Mattar SG, Lumsden AB. The management of splenic artery aneurysms: experience with 23 cases. *Am J Surg.* 1995;169(6):580–4.
20. de Perrot M, Buhler L, Deleaval J, Borisch B, Mentha G, Morel P. Management of true aneurysms of the splenic artery. *Am J Surg.* 1998;175(6):466–8.
21. Ouchi T, Kato N, Nakajima K, Higashigawa T, Hashimoto T, Chino S, et al. Splenic artery aneurysm treated with endovascular stent grafting: a case report and review of literature. *Vasc Endovasc Surg.* 2018;52(8):663–8.
22. Akbulut S, Otan E. Management of giant splenic artery aneurysm: comprehensive literature review. *Medicine.* 2015. <https://doi.org/10.1097/MD.0000000000001016>.
23. Dolar E, Uslusoy H, Kiyici M, Gurel S, Nak SG, Gulten M, et al. Rupture of the splenic arterial aneurysm due to Behcet's disease. *Rheumatology.* 2005;44(10):1327–8.
24. Desinan L, Scott CA, Piai I, Mazzolo GM. Sudden death due to spontaneous rupture in splenic artery atypical dissection with features of vasculitis: case report and review of the literature. *Forensic Sci Int.* 2010;200(1–3):e1–5.

25. Aydin MT, Fersahoglu MM, Tezer S, Okuducu M, Agca B, Memisoglu K. Spontaneous rupture of the splenic artery aneurysm: a rare clinical presentation of acute abdomen. *Ulus Travma Acil Cerrahi Derg.* 2016;22(1):106–8.
26. Park JH, Chung JW, Joh JH, Song SY, Shin SJ, Chung KS, *et al.* Aortic and arterial aneurysms in Behçet disease: management with stent-grafts—initial experience. *Radiology.* 2001;220(3):745–50.
27. Akiyama M, Kaneko Y, Takeuchi T. Characteristics and prognosis of IgG4-related periaortitis/periarteritis: a systematic literature review. *Autoimmun Rev.* 2019;18(9): 102354.

### **Publisher's Note**

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

**Ready to submit your research? Choose BMC and benefit from:**

- fast, convenient online submission
- thorough peer review by experienced researchers in your field
- rapid publication on acceptance
- support for research data, including large and complex data types
- gold Open Access which fosters wider collaboration and increased citations
- maximum visibility for your research: over 100M website views per year

**At BMC, research is always in progress.**

Learn more [biomedcentral.com/submissions](https://biomedcentral.com/submissions)

