

An Insidious Rare Cause of Abdominal Pain Presentation in The Emergency Department: Splenic Artery Aneurysms Complicated by Rupture – A Systematic Review of The Literature

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Abstract

The splenic artery aneurysms (SAA) are vascular malformations with spontaneous localized dilations of the splenic artery, which may cause life-threatening conditions. It is the most common visceral artery aneurysm – but SAA are rare, present in approximately 0.2% to 10% of general population. Meaning of SAA is underlined by the propensity for its rupture, which occurs with a frequency of 10% to 20% and results in chance of hemorrhage with catastrophic potential, with consequent high mortality rates; Such hemorrhage is usually presented with non-focalizing abdominal pain, thereby making diagnosis a challenge in the emergency department because of the absence of pathognomonic features and variable clinical presentation.

Keywords: Splenic Artery Aneurysm, Hemoperitoneum, Non-Traumatic Hemorrhage Shock CT Angiography, Endovascular Treatment.

Introduction

Worldwide, the aberrant dilation of the splenic artery (i.e., splenic artery aneurysm (SAA)) is the most prevalent visceral artery aneurysm. Splenic artery aneurysms are responsible for 70%-90% of all visceral aneurysms. Incidence rates vary from 0.2% to over 10%, but their precise incidence is difficult to assess based on imaging pathology studies [1]. Because they are usually asymptomatic, splenic artery aneurysms are often found in-

cidentally during surgery for other conditions or during imaging studies such as abdominal ultrasound, CT, or MRI performed for other reasons. If symptoms develop, they consist of abdominal or cardiac pain, and symptoms are due to ruptured aneurysm in 5%-20% of cases. Aneurysms may also present with other symptoms related to the sequelae of rupture or compression of adjacent structures, such as leg edema, hypovolemic shock due to intra-abdominal hemorrhage, and left-sided pleural effusion

due to bleeding into the space of Morrison.

A review of the literature regarding ruptured SAAs is provided. Since its discovery, more than 900 cases of SAA ruptures have been reported. Centrally located SAAs (i.e., SAA with lesser curvature and located 5 cm from f- BGEH) were significantly more likely to rupture at presentation. Additionally, lesions >2cm are much more likely to rupture at presentation [2]. Out of 101 cases, 59 patients survived the rupture of a splenic artery impeller, indicating that the prognosis of those presenting with this type of rupture is grim.

Anatomy of the Splenic Artery

The splenic artery arises from the celiac trunk as its largest branch. It usually runs horizontally, from right to left, to reach the hilum of the spleen. The splenic artery deserves special attention because it gives off branches to areas, including the pancreas, stomach, omentum, diaphragm, and colon 2. The splenic artery in adults has an average length of 8.7 to 19.3 cm, an average inner diameter of 3.5 mm, and an average branching angle from the celiac trunk of 55 degrees 1. Rudimentary branches of the celiac trunk frequently give rise to an indeterminate number of splenic artery branches [3].

The splenic artery follows a serpentine course behind the body

and tail of the pancreas. It runs in the spleno-pancreatic ligament and then arches back. Near the hilum of the spleen, it divides into several terminal branches that enter the spleen. The branches are termed the splenic artery branches and continue into the substance of the spleen, known as trabecular arteries. The smaller branches of the blood vessels end in the pulp of the spleen without forming anastomosis and terminate as fine capillaries. Thus, infarcts occur in this region. The splenic artery gives five to six pancreatic arteries to the body and tail of the pancreas. The other branches include an omental branch, which contains the artery to the great omentum and a colic branch. The warning signs should be reported to avoid complications. These aneurysms rarely present rupture or symptoms due to their location deep in the abdominal cavity. Understanding splenic anatomy and variations has clinical and surgical significance.

Overview of Splenic Artery Aneurysms

Splenic artery aneurysms (SAAs) are the most prevalent visceral artery aneurysms, accounting for approximately 60% of all visceral artery aneurysms. Initially, SAAs are most discovered during imaging for unrelated reasons and typically have a bilateral distribution. They are often asymptomatic, with a limited number of reported spontaneous ruptures. Key risk factors include being female, having certain.

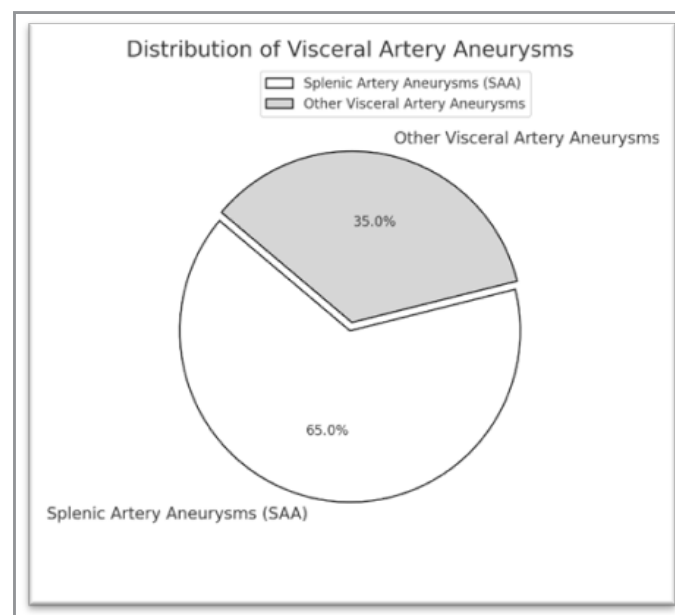


Figure 1: Distribution of Visceral Artery Aneurysms Rupture.

connective tissue diseases, and undergoing pregnancy [4]. SAAs may cause a variety of signs and symptoms. A ruptured SAA will result in signs of hemodynamic instability, such as abdominal pain, peritoneal irritation due to intraperitoneal bleeding, or flank pain due to retroperitoneal. Diagnosis can be made via CT or, in case of increased hemorrhagic suspicion, via either CTA or US. CT is the test of choice due to its high availability, speed, and reliability. The diameter of the splenic artery will be measured, and a specific cutoff will be used as a threshold for the diagnosis of SAA. Treatment can be surgical (open/vascular surgery, laparoscopic) or endovascular (covered stent-graft, coiling). Surgery is the best treatment option for those patients

with ruptured SAAs. However, patients with SAA who are asymptomatic can be treated by waiting and careful follow-up, provided they are hemodynamically stable [5]. Endovascular techniques are now widely applied due to the development of new devices. Embolization of SAAs can also achieve a variety of approaches. Intra-arterial coiling is the go-to procedure and gold standard for large aneurysms. Covered stent-graft exclusion is the gold standard treatment for SAA in large SAA involving proximal artery. Selective embolization, in both the parent artery and feeding arteries, can also eliminate some small SAAs. Intra-operative laparotomy and ligation can protect against rupture propagation in larger SAAs.

Epidemiology

Splenic artery aneurysm rupture is an uncommon yet severe medical emergency condition that can result in serious hemorrhagic complications. SAAs can be found in pregnant women and individuals with connective tissue disorders. They may also be symptomatic and found incidentally on imaging done for other

reasons. Isolated cases of ruptured SAAs have been previously reported, leading to discussions on this rare occurrence. Women between 3rd and 4th decades commonly suffer from these aneurysms, and SAA ruptures are more common during pregnancy [6].

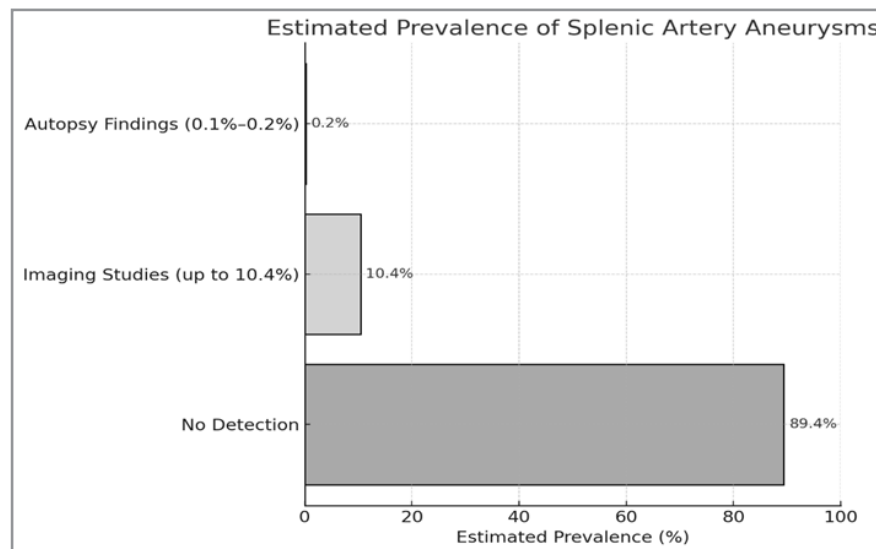


Figure 2: Prevalence of SAA

Ruptured SAAs are commonly found in women with pregnancies between the 14th and 22nd week. There are three possible reasons as to why this happens. First, there are hemodynamic changes during pregnancy. Second, hormonal changes affecting the medium and small-sized vessels. Lastly, splenic vessels can be affected by compression due to growth. Some systemic diseases can cause hepatic, splenic, or renal artery aneurysms. This sequence is well established in polycystic kidney disease, a disorder with transmission from parents to offspring, where other aneurysms may be found, such as in the liver and poten-

tially in the spleen [7]. Several connective tissue disorders may affect the splenic artery: Ehlers-Danlos syndrome, fibromuscular dysplasia and Marfan's Syndrome. These disorders may be inherited either as a dominant or recessive defect SAAs can be symptomatic, and abdominal pain, or epigastric pain, may follow the rupture of the aneurism. These may either appear as dull pain or as acute severe pain that might also be accompanied by vomiting and syncope. SAAs may also be completely asymptomatic and found incidentally when imaging is done for other unrelated reasons.

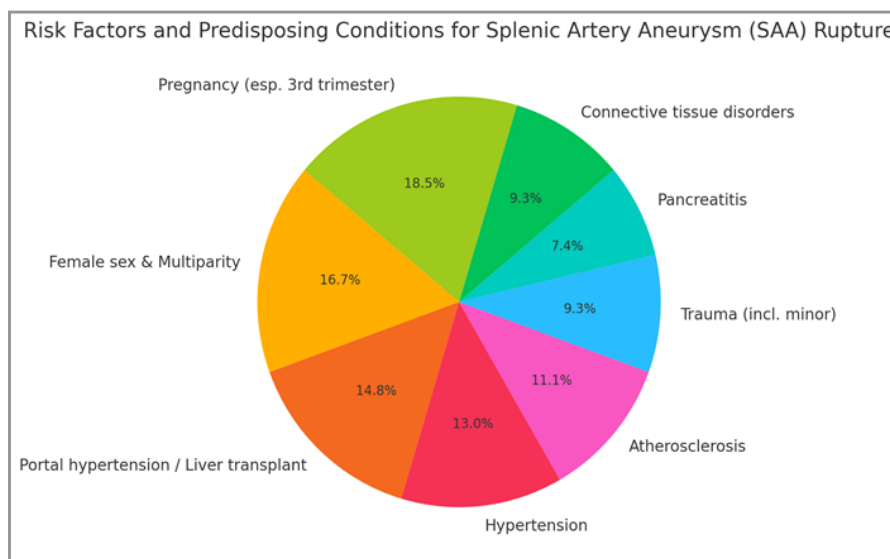


Figure 3: Risk factors and conditions for SAA-rupture

Pathophysiology of Aneurysm Formation

The initiation of the normal arterial wall involves the endothelial barrier, which triggers inflammation, resulting in chronic macrophage accumulation and thickened intima or an atheroma. In response to biomechanical stress, smooth muscle loss results in localized degenerative changes in the vessel wall. Progressive expansion may lead to rupture and thromboembolic occlusion of the distal vessel or center cavity growth and distal vessel hypertrophy. Aneurysms may be present as mass lesions that are asymptomatic, with symptomatic cases most commonly occurring after rupture. The splenic artery is a branch of the celiac trunk and winds along the superior border of the pancreas. It is prone to the development of atherotic plaques or aneurysms. Aneurysms of the splenic artery were relatively rare in the early 20th century; however, their incidence has increased considerably, probably owing to the advancement of evaluation techniques. Most splenic artery aneurysms are asymptomatic [8].

However, their rupture may give rise to potentially fatal intra-abdominal hemorrhage. Aneurysm formation has been attributed to various causes or associated factors such as hemodynamic stress, atherosclerosis, pregnancy, fibromuscular dysplasia, and bacterial or fungal infections. Nevertheless, the precise pathogenesis of SAA formation remains unclear in most patients. Athero-atheroma formation is the most widely accepted theory, and several models of hemodynamic-induced athero-atheroma formation in small male swine exposed to intra-arterial pressure-induced arterial injury have been developed. However, splenic artery aneurysms (and arteries in general) do not develop atheroma in an appropriate swine model, making it difficult to further examine the relationship between aneurysms and arterial wall degeneration.

Clinical Presentation

Rupture of splenic artery aneurysm (SAA) is a rare cause of intraabdominal bleeding that can cause equal mortality and morbidity in acute abdomen. Until the 1970s, SAA was considered an incidental finding that was treated only upon rupture. With increasing awareness and improvement of imaging techniques, diagnosis is being made prior to rupture in more patients. Pri-

mary abdominal complaints vary from vague abdominal pain, chronic or postprandial discomfort to acute localized pain. If hemorrhage is massive, it may result in hypotension, syncope, malaise and coma. Patients w/o preceding symptoms or with rare number of symptoms may still be diagnosed with ruptured SAA. Abdominal ultrasound is generally favored as the initial imaging technique in the evaluation of acute abdomen due to lower costs, portability and better acceptance of patients. Generally, DPL or CT scan is more accurate to evaluate the abdominal cavity and recognize the leak points. Standard DPL used to establish the diagnosis of hemoperitoneum in patients with suspected ruptured SAA. Surgical intervention by laparotomy, laparoscopy or interventional radiology is considered as the standard treatment of choice. The choice of surgical intervention is defined by age, clinical status of patients, spleen and proximity to main trunks of vessels [9].

Symptoms of Splenic Artery Aneurysm

Initial diagnosis may be difficult, as there are no clinical features specific to SAAs. They may be asymptomatic or diagnosed incidentally. The sudden onset of pain in the upper left quadrant, back or left flank due to rupture is the most common presentation of symptomatic SAA, which can radiate to the shoulder due to diaphragm irritation. Other atypical symptoms include nausea and vomiting, gastrointestinal bleed, and obstructive jaundice when it ruptures to the stomach or duodenum [10]. Sequestration is possible with persistent left upper quadrant pain, anemia, and simulating splenic malignancy. Intra-variation SAAs may present with bilateral abdominal masses, pain, and drainage. Both ruptured and unruptured SAA patients presented with abdominal pain, followed by hypotension, fever, and anemia. However, as SAAs are rare and not routinely monitored, patients with sudden-onset pain probably are misdiagnosed with benign conditions such as renal colic, intercostal myalgia, peptic ulcer disease, pancreatitis, and others, leading to delayed consultation and rupture. Occasionally, rupture evacuation often occurs three to five hours after the initial pain episode. These cases pointed out the rapid candidacy for ruptured evacuation if an SAA is clinically suspected in urgent surgical centers.

Figure 4: Different clinical Aspects Between SAA and SAP

Symptom / Sign	Condition Type	Clinical Notes
Asymptomatic (incidental finding)	SAA	Often discovered during imaging or surgery
Left upper quadrant (LUQ) pain	SAA	Most common symptoms of ruptured SAA
Back or left flank pain	SAA	Can radiate from LUQ or mimic renal colic
Referred shoulder pain (Kehr's sign)	SAA	Due to diaphragmatic irritation
Nausea and vomiting	SAA	Atypical presentation
Gastrointestinal bleeding (hematemesis/melena)	SAA	May indicate rupture into GI tract
Obstructive jaundice	SAA	Due to compression of biliary structures
Anemia	SAA	Possible chronic bleeding or sequestration
Hypotension / Hemodynamic instability	SAA	May follow rupture; urgent sign

Fever	SAA	Reported in both rup-tured/unruptured cases
Bilateral abdominal mass-es	SAA	Uncommon; in cases of intra-variation
Drainage from intra-variation SAA	SAA	Associated with anatomi-cal anomalies
Splenic mass simulation (mimicking malignancy)	SAA	May mimic neoplastic processes
Misdiagnosed as renal col-ic, peptic ulcer, myalgia, pancreatitis	SAA	Contributes to delayed di-agnosis
Rupture 3-5h after initial pain	SAA	Observed delay in surgical consultation
Chest pain (SAP)	SAP	May mimic cardiac or pleural conditions
Steady and worsening ab-dominal pain (SAP)	SAP	Typical SAP pain feature; non-pleuritic
Hemoperitoneum with acute abdomen (SAP)	SAP	Mimics gynecological emergencies
SAP presentation with GI bleeding (77%)	SAP	Most frequent SAP presen-tation
SAP presentation with ab-dominal pain (30%)	SAP	Occurs in one-third of SAP cases

Differential Diagnosis of Abdominal Pain

Splenic artery pseudoaneurysm (SAP) is a challenging and uncommon etiology of acute abdominal pain or hemorrhagic shock. Unlike splenic artery aneurysms (SAA), splenic artery pseudoaneurysms are usually secondary to trauma, pancreatitis, or infection. In the absence of these conditions, a splenic artery pseudoaneurysm should be deemed a Mayo Clinic Classification Type I. Spontaneous SAP rupture occurs most commonly due to atherosclerosis, followed by pancreatitis and vasculitis. Due to its rarity, as well as the vast differential diagnoses when presented with this acute presentation, a high degree of clinical suspicion is necessary to make the diagnosis in a timely manner [11]. Prompt detection and intervention are critical, as untreated SAP rupture has a 90% mortality. To avoid missed diagnoses, it is critical to maintain a broad differential diagnosis for abdominal pain. As in the presented case, the initial symptoms of SAP rupture may include non-specific signs and symptoms such as abdominal pain, flank pain, chest pain, nausea, vomiting, or gastrointestinal bleeding. The abdominal pain is typically steady and worsening with no alleviating factor, as the splenic artery is a retroperitoneal vessel and thus not as likely to cause pleuritic pain. Most patients present to the emergency department with hemodynamic instability due to SAP rupture; however, they may

also present without hypotension and other signs of hemorrhagic shock. When SAP rupture occurs, the resultant hemoperitoneum causes acute abdominal pain that can mimic more common conditions such as ruptured ectopic pregnancy or ruptured ovarian cyst. The most common presentation of this condition is gastrointestinal bleeding (77%); however, acute abdominal pain occurs in approximately 30% of patients, requiring a high degree of clinical suspicion for an early and accurate diagnosis [12].

Diagnosis

The diagnosis of a splenic arterial aneurism (SAA) may be confirmed using doppler ultrasonography, CT, MRI, or aortography after a clinical suspicion. The splenic arteries' selective catheterization or a survey of the abdominal aortogram may reveal the lesion as an out-pouching of the lumen of the artery. The lesion's precise location and size may be determined by doppler duplex ultrasonography. CT is a more accurate technique, revealing the aneurysm's morphology as a saccular expansion of the arterial lumen [13]. Although angiographic diagnosis has become the gold standard, doppler ultrasonography is often the preferred initial screening method.

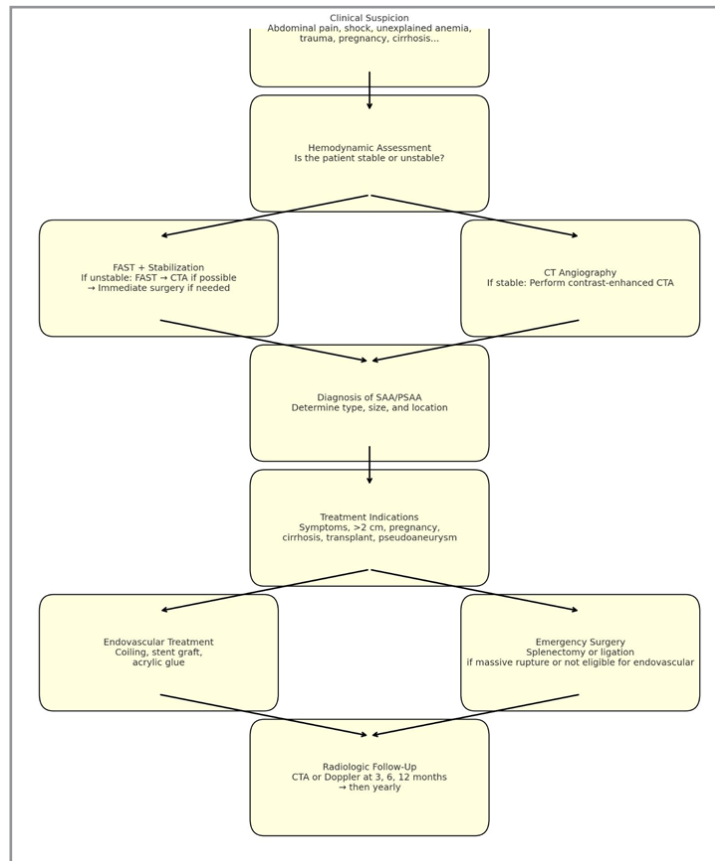


Figure 5: Diagnostic Algorithm for Suspected Splenic Artery Aneurysm (SAA) or Pseudoaneurysm (SAP)

The reported prevalence of splenic artery aneurysms is approximately 0.5%. Splenic artery aneurysms account for only 4% of all visceral arterial aneurysms, but they are the most frequent splenic artery vascular malformations. The implication of the diagnosis is profound. There is a substantial risk of rupture when the patient is a woman who has been or is going to become pregnant. In general, the result may be fatal. End-stage splenic artery aneurysms have been described in asymptomatic or involving cases; however, they are less well-documented. Historically, the Roberts-Peabody operation, which involved autotransplantation or excision, was employed. In more recent literature, mesh- and non-mesh-based methods have been detailed as simple surgical options to treat the disease primarily.

Imaging Techniques

A SAA is an abnormal dilation of the splenic artery, which can lead to aneurysm rupture and is a rare cause of splenic hemorrhage. SAA are usually asymptomatic and non-traumatic. Therefore, SAA are usually diagnosed incidentally in adults. The detection of an SAA in patients who are symptomatic or have an aneurysm >2 cm warrants further examination and surgical intervention. It is important for imaging procedures to quickly confirm or refute the diagnosis of SAA in ruptured patients [1]. In recent decades, ultrasound (US) ("Korean Flag" sign", see below), computed tomography (CT), and magnetic resonance angiography (MRA) have become the imaging techniques of

choice for SAA [14]. Doppler, routine or contrasted, is the recommended first approach in patients suspected to have an SAA.

There is a consensus that Doppler US has at least 80% sensitivity for detecting the presence of a visceral aneurysm, but a limited specificity due to other conditions. Some limitations of this method are the frequent need for additional studies to define vascular anatomy, and the greater experience required to minimally qualify radiologists who perform this imaging. However, as the first selection alternative in the case of a ruptured SAA, it should be utilized before and especially in lieu of CT or MRA, which often are not immediately available. When the Doppler study results are unclear and there is a diagnostic doubt, a contrast study is warranted to confirm or discard the existence of an SAA. US is a well-established radiation-free imaging methodology that is usually used as the first imaging test for detecting SAAs. Pseudoaneurysms produced by splenic artery pancreatitis erosions can also be represented in the US as echogenic masses near the splenic hilum. US is usually less sensitive than CT, especially with larger SAAs. Very sensitive Doppler US techniques can be employed, nevertheless, for the screening of SAAs even in very large patients. However, gas distension, abdominal walls and respiratory motion artifacts may frequently limit an adequately performed US exam. Additionally, a true aneurysmal growth >1 cm may not be detected with traditional US techniques, especially with larger patients.

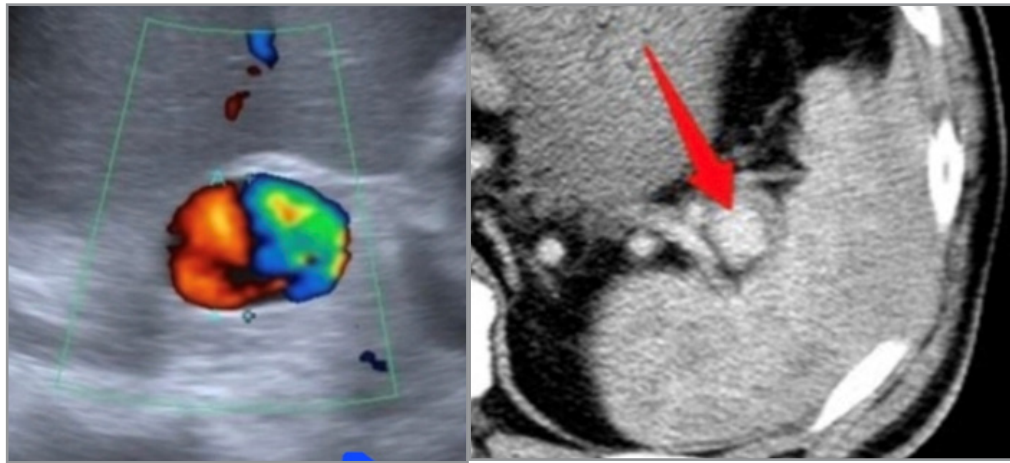


Figure 6: “Korean Flag pattern” (left), CT-angio (right). (From web)

Laboratory Findings

Laboratory evaluation is crucial for diagnosing ruptured splenic artery aneurysms, as many cases aren't discovered until a rupture occurs. Fluid resuscitation and blood transfusions are vital for unstable patients. White blood cell count may surpass 10,000/mm³. Blood typing and crossmatch are also important as many patients may need immediate transfusion. Other electrocardiography and imaging studies aren't generally needed to establish a diagnosis. Hb levels may decrease significantly, and platelets should be normal. The quantity of peritoneal and retroperitoneal bleeding is often trivial in SAAs. Peritoneal bleeding is usually acute and less than 150 cc, with blood becoming brownish as peritoneal irritation occurs [15]. During diagnosis, the hematological tests may reflect specific findings and are useful prior to surgical intervention. White blood cell count may be elevated in about 50% of patients. A variety of tests could indicate progressive anemia and may be used to assess the hemorrhagic rate. Patients may be hypovolemic and mildly hypertensive even after repeated cross-matching. Physical examination shows signs of organ failure as a result of prolonged hypoperfusion for unstable patients. Computerized tomography is the best method to ascertain the anatomical circumstances of bleeding and to plan surgical procedures. Aneurisms can be recognized as a focal structure that widens the artery. The disparity between CT-infusion and CT-angiography performed widely prior to surgical operation makes the vascular phase vital for diagnosis and assessment of rupture. Angiography may be seen in the vascular phase or the late parenchymal phase. In the late phase the contrast agent can't retain harmonic and as follows no sign of lesions is detectable. To maintain the harmonic for a longer period of time, ISO-osmolar contrast medium or newer agents with the parallel solution-detector capability may be used. Contrast agent and a time interval close to the large expectation may retain anatomical characteristics. There're various ways to diagnose splenic artery aneurysms with each having distinct pros and cons. Transfusion is essential in critically unstable patients, as patients must be stabilized before an imaging investigation. Although laboratory findings may vary, arterial bleeding can be supported by hemoglobin measurement, which significantly lowers to may make other sources more probable.

Management of Splenic Artery Aneurysm

Surgical repair of a SAA is a simple thing, yet it may become a

challenging or almost impossible task for the surgeon who suddenly receives the ruptured one. Management of SAA rupture is classically performed through emergency open procedures, considering the method a routine laparotomy and splenectomy. The situation may become hard, depending on the acuity of hemorrhage, anatomical situation, but also the operator experience. Many questions arise when dealing with a ruptured SAA, for example, whether or not technical failure should provoke splenectomy, how to deal with a huge spleen, how to augment the surgical field, which repair option is safer, or possible failure of repair. These questions have led to new thoughts and established different techniques to help the surgeon deal with such challenges. Several procedures have emerged, almost rendering the classical open procedure with regular laparotomy and splenic dissection. These include the laparoscopic, hybrid, and embolization techniques. Another idea is to deal with the ruptured aneurysm with the patient in transverse decubitus position. This might take even as long as 90 min before conversion to an “open procedure” thereafter. Still others have stressed performing emergency laparotomy from outside the peritoneal cavity, to allow quick control of hemorrhage, or to use nasal packing prior to anesthesia. These examples illustrate that management of a ruptured SAA may indeed warrant innovation and improvisation depending on the situation. Management of a ruptured SAA can be done through various means, from classical to pioneering, or judged easy to very difficult. Each operator's collection of cases and situations will determine his/her capabilities to deal with such challenges. Experience is the best teacher. Nonetheless, awareness of innovative instruments is crucial; inspiration from usage and discoveries by others has often helped nursing, engineering and medical professionals make major advances. Such perspective view may lead to future modification of instruments and techniques to improve care.

Conservative Management

The bleeding at the base of the splenic artery after ruptured SAA might be, if detected before the appearance of ATM, successfully treated with conservative management. To identify cases where an attempt at conservative management might be clinically appropriate, timely under a CT biopsy rhythm should be done, targeting the SAA ruptured point, possibly allowing the percutaneous placement of a pseudoaneurysm catheter occlusion coil avoiding splenectomy. Although a splenic artery

coil occlusion might be a temporary measure; repeated selective catheterization of the splenic artery could be done with the resolution of multiple pseudoaneurysms and patent splenic artery in CT follow-up. Very few clinically reported cases are available with conservative treatment. [7] A misdiagnosed SAA after trauma ruptured patient with acute hypotension and splenic focal bleeding should warrant a continuation path in newly stepped hospitals, where an older CT or trans-abdominal US scanning may later be used to safely figure out a further step: conservative management for SAA cases or splenectomy otherwise.

If the focal bleeding is suspected to happen at the vicinity of the splenic artery outside the hilum of the splenectomy, a close indication of delays at bleedings should be targeted at smaller aneurysms and attempts made to maintain the patency of the splenic artery and avoid bigger events. Debridement of such ruptured focal bleeds done right away or up to 5 hours later has been successful reported with close indications. The key information gained, and clinical suggestion depicted from the case report are that; The failed traditional squeeze-2 pinch maneuver directed hastily to the splenic hilum pons on CT follow-up might have hastened the post-hand surgery ATM event. The ATAP prescreening passive blood pressure loss testing should have been rendered afterwards to gain more confirmatory information, rather than directly proceed to exploration.

Surgical Interventions

Endovascular treatment has emerged as a less invasive option for SAAs repair, and its use is increasing. Numerous endovascular techniques and devices are now available to treat suspected ruptured and no ruptured SAAs. Embolization using coils and Onyx is the most widely used technique for treating SAAs worldwide 7. Stenting is a promising alternative to prevent re-occlusion after coil embolization. Several case series with a limited number of patients have reported the successful use of other endovascular devices. However, most of these devices were custom-made or not specifically designed for use in the splenic artery. Open surgery remains the gold-standard treatment, with high clinical success rates. Similarly, to transcatheter techniques, several open surgery techniques have been developed and modified to treat SAAs. As SAA and splenic artery anatomy varies from case to case, a thorough understanding of the anatomy is crucial when selecting the appropriate technique [16]. There is a lack of consensus on the first-line treatment approach for ruptured SAAs 8. Practically, treatment methods are commonly determined and modified based on institutional experience, operator proficiency, and patient condition. However, surgery can be a difficult and challenging option for most

rupture cases, particularly because many patients present with hypotension after rupture.

Timing is critical, and preparations, such as ensuring adequate resuscitation and angiography availability, must be made. Rushed transfer to the institution may prevent optimized preparation and endanger patient lives. Therefore, a reliable treatment option with minimum institutional requirements is an important consideration in determining how to manage ruptured SAAs. Surgery is the treatment of choice for ruptured SAAs, especially in patients presenting with hypotensive shock or during pregnancy [17]. Most surgeons prefer open surgery, since it more reliably prevents rebleeding and is better suited for managing various variables, such as anatomic variations, multiple arteries supplying the spleen, and dense adhesion after previous procedures. Consequently, surgery was chosen in 65% of patients with ruptured SAAs. In patients with suspected ruptured SAAs, especially in those presenting in shock, endovascular treatment is the usual first-line treatment option to quickly gain access to the vascular system and to achieve provisional hemostasis. However, surgery must be performed to prevent rebleeding.

Complications of Aneurysm Rupture

Splenic artery aneurysms are the most common visceral artery aneurysms encountered in clinical practice and comprise 60% of all visceral artery aneurysms. There is a 60% prevalence of multiple aneurysms in patients and a 25% incidence of distal splenic artery aneurysms. This is an enormous vascular malformation causing a difficult treatment-related dilemma. Dissection and rupture are rare but dangerous complications due to the proximal location, extensive vessels, and vascular territories, leading to external and internal bleeding. The reported incidence of rupture is 20% to 30%. Extra-digestive hemorrhages, peritonitis, abdominal or dorsal pain, hypovolemic shock, febrile syndrome, pancreatic necrosis, and loose ileus episode may occur as acute onset symptoms. Diagnosis is difficult, complex, and costly to perform [18].

At the time of diagnosis, 60% of patients with splenic artery aneurysms are symptomatic. In case of sudden rupture, significant internal bleeding or avulsions may occur, or the spleen and pancreas may become necrotized because of ischemia. Death rates may be high due to difficult diagnosis and treatment. Moreover, even in the case of diagnosis, diagnosis and treatment are difficult. A systematic review of the published literature regarding spontaneous ruptured splenic artery aneurysms is performed to discuss uncommon encounter aspects of treatment and outcome. Specific keywords are used for identifying related case series.

Figure 7: Surgical Pathology and Indicated Treatment

Artery	Indications for Repair
Hepatic Artery	• Symptomatic aneurysms • Diameter > 2 cm • Growth rate > 0.5 cm/year
Splenic Artery	• All pseudoaneurysms • Diameter > 3 cm • All sizes in women of childbearing age

Renal Artery	• Symptomatic aneurysms • Diameter > 3 cm • All sizes in: - Women of childbearing age - Patients with refractory hypertension and renal artery stenosis
Renal Artery	• All pseudoaneurysms • Diameter > 2 cm
Celiac Artery	• Repair all aneurysms regardless of size
Pancreaticoduodenal and Gastroduodenal Arteries	• Repair all aneurysms regardless of size
Superior Mesenteric Artery	• Repair all aneurysms regardless of size
Jejunal and Ileal Arteries	• Symptomatic aneurysms • Diameter > 2 cm
Colic Artery	• Repair all aneurysms regardless of size

Case Studies

SAAs are rare vascular lesions, with an estimated prevalence of less than 2% in the general population. Spontaneous rupture is an uncommon and possibly life-threatening complication. The case of a 38-year-old female presenting with an acute abdomen caused by splenic artery aneurysm rupture is described. Transabdominal ultrasonography revealed an intra-abdominal hematoma and splenic artery aneurysm. Angiography showed large size of the aneurysm, which was treated with a spleen-preserving procedure—splenic artery resection and distal arteriostasis. Successful anatomical and clinical endpoints with a good angiogram and preserved spleen on the 14th day were achieved. SAAs are rare but should be considered as a possible cause of abdominal pain and a hemodynamically unstable patient. The diagnosis is primarily made by abdominal ultrasonography and confirmed by angiography. Surgical intervention should be performed in symptomatic cases as soon as possible. Spleen-preserving techniques should be thought of and evaluated in cases with no other co-morbidities.

A 19-year-old male presented to the Emergency Department with acute onset abdominal pain concurrent of vomit, fever, and chills with a past medical history suggestive of splenomegaly. CT angiography revealed a 20 mm splenic artery saccular aneurysm with extravasation of the contrast at the dome of the aneurysm with subcapsular hematoma of the spleen 2. He underwent exploratory laparotomy, which revealed a ruptured SAA with a large hematoma and devascularized spleen. The peritoneal drain was secured after the ligation of the splenic artery at its distal portion. The patient was stable in the post-operative period and discharged after six days. Literature review reveals spontaneous SAA rupture usually occurs among women between the ages of 30 and 50. Factors causing increased blood flow to the splenic artery are believed to trigger aneurysm formation. Most SAAs are asymptomatic.

With rupture, 20% to 30% of patients are hemodynamically stable. When patients are present with hypotension, the mortality is 50% or greater. Several recent case reports published the spontaneous rupture of a splenic artery aneurysm in a 38-year-old female treated with a spleen-preserving procedure. A review of the literature on clinical presentation, diagnosis, and treatment modality was conducted with special attention to the spleen-preserving procedures. Patients usually complain of abdominal pain

due to bleeding into the peritoneal cavity, and imaging is mostly by computed tomography, which is a good non-invasive method for diagnosis in hemodynamically stable patients.

Most patients require surgical intervention, and procedures include splenectomy, excision and ligation, or fenestration. The latter techniques are spleen-preserving techniques, which are more recent. Altogether, a case of spontaneous rupture of a splenic artery aneurysm in a young female with a successful spleen-preserving procedure is presented, including a review of the related literature. SAAs are the most common type of splanchnic artery aneurysms. There are only a few case reports on the spontaneous rupture of SAAs. Although ruptured SAAs are not common, they can lead to serious complications with a high mortality rate of up to 25% on admission. A slit-like defect in the back wall of the aneurysm and a small amount of hematoma in the splenic hilum vessels led to delayed emergence of a clinical picture of acute abdomen 1.5 h after admission. The patient was taken to the operation theatre for an urgent exploratory laparotomy. Due to ongoing freshness of the bleeding, oozing was primarily stopped from the splenic artery with a Cope ligature and reach window-type capsular resection and approximating of the spleen capsule. Excision of the preoperative parent artery and aneurysm was not possible. The ruptured aneurysm in the splenic hilum was approached by an intraglandular route. The hematoma was evacuated using a tourniquet on the splenic artery. Reconstruction of the splenic artery was done using the parent artery beyond the ruptured site and reconstructing the splenic artery with a carotid interposition graft. The second case involved a patient with abdominal pain, vomiting, and fever lasting from the morning complaining of frequent vomiting, abdominal pain, chillness, and headache [19].

Analysis of Outcomes

The aim of the present study is to review the literature on demographic characteristics, management, morbidity and mortality of spontaneous rupture of splenic artery aneurysms. Though r-SAAs are not frequent, a case is presented with the intent of emphasizing their challenging management. This systematic review provides literature data concerning the outcomes of r-SAAs. Rupture of a splenic artery aneurysm is a rare but dangerous condition, with a reported incidence of 0.01 to 0.2 % in the general population. It is often associated with high mortality and morbidity. Though usually asymptomatic, approximately

20–30 % of patients may present with rupture.

The first clinical symptom is usually sudden severe pain in the left upper abdominal quadrant, which may radiate to the left shoulder due to diaphragmatic neuropathy. The severity of the pain and the presence of some other symptoms depend on whether the aneurysm ruptures or not. The rupture of an SAA can be either intraparenchymatous, which is less frequent, or subcapsular. SAFs, following complete rupture, do not remain without extra splenic fluid collection. They are usually large, expanding in time, and may also contain necrotic fragments, in-

ducing a state of sepsis in the patient. Though splenectomy was previously considered the hallmark of treatment for r-SAAs, a spleen-preserving procedure using only interventional radiology is now also available. Minimal invasive techniques have better short-term outcomes. To conclude, r-SAA has high mortality and morbidity. The optimal management is still controversial; the best available evidence is presented in this review. Endovascular treatment compares favorably to surgery with regard to reintervention; however, no evidence of better outcomes in terms of death and complications is available. This is especially true for patients with hemodynamic instability during pregnancy [20].

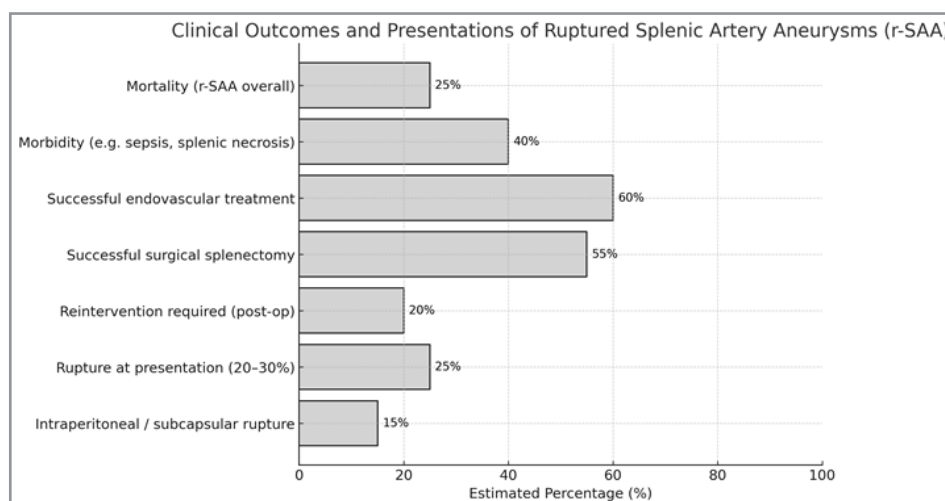


Figure 8: Clinical outcomes of Ruptured SAA

Recent Advances in Research

SAAs are rare vascular malformations. SAAs are most often found incidentally. Spontaneous SAA is a common emergency in general surgery. Distal vascular interventions can be achieved with an intact spleen through very few saccular SAAs. The mode of spontaneous rupture of SAA remains obscure. The SAA did not grow for many years, and the mechanism of spontaneous rupture was observed to be the exposure of closer-encysted ruptured points. The last ruptured point was a projection into the vascular cavity, ignoring the non-luminal vascular cavity. Spontaneous rupture of SAA is a rare life-threatening condition. Because SAA can be discovered incidentally, there is rising awareness of identifying this entity in the absence of a prior diagnosis. Considering the scarcity of published review papers, there is suspicion that the literature on SAA has not kept pace with its clinical importance. A literature review of SAA focusing on pathogenesis, morphology, diagnosis and management is provided. Regardless of modality or expertise, the high detection rate of SAA in most radiological practices indicates that awareness of this entity is growing, prompting the belief it must be studied further.

Emergency Department Protocols

In the Emergency Department setting, strict protocols should be adhered to when assessing potential ruptured splenic artery aneurysms to ensure no critical steps are missed [2]. A detailed history and examination should be the first line of investigation for potential pre-existing splenic artery aneurysms. This should include careful checking for features of hepatosplenomegaly, liver dysfunction, signs of fluid overload, peritoneal irritation, such as palpable splenomegaly and a tender mass in the left upper

quadrant. Patients should be investigated with a supine abdominal X-ray or ultrasonography and a CT scan of the abdomen and pelvis with an intravenous contrast [21].

CXR may show blunting of the fissures or pleural effusion, while the US may show a hemoperitoneum. If available, a focused assessment with sonography for trauma may also assist in the diagnosis. In experienced hands, CT angiogram is the investigation of choice. This provides anatomical definition of aneurysm, vascular branches and feeding vessels, and organ involvement. In addition, it may provide the opportunity for interventional radiology to embolize the aneurysm or feeding vessel.

Management of ruptured splenic artery aneurysms is dictated by degree of hemodynamic instability and surgeon preference. In patients with class I hemodynamic stability (systolic BP > 90) angiography and selective embolization is recommended. Alternatively, in those with uncontrolled bleeding, laparotomy with splenic artery replumbing, splenectomy and/or packing is recommended. In patients with class II-III (systolic BP 60-90) and absolute hypo-perfusion, emergent negative exploration, splenectomy, and/or aortic cross clamping is recommended. Finally, in patients with class IV-early class III (systolic BP < 60) L3, laparotomy with splenectomy is indicated in cases without increased INR and low fibrinogen.

Initial Assessment and Triage

Patients with SAA rupture are most commonly present with an acute onset of left upper quadrant pain, which may refer to the left shoulder as Kehr's sign. Rarer presentations include corroborating complaints of syncope, epigastric pain, or abdominal

distension. In addition, as patients may present to an obstetrics literature with abdominal pain and a positive pregnancy test, care must be taken in females of reproductive age not to miss this diagnosis. Young patients presenting with acute abdominal pain and mild tenderness, as opposed to peritonitis, should be screened for SAA, as high-risk patients may be missed by careful clinical assessment alone. Those with known high-risk factors or signs of ruptured SAA should have tissues for type and crossmatch sent immediately, and a working hemoglobin should be obtained [22].

If not already available, portable chest X-ray and flat abdominal X-ray should be ordered. Plasma volume expanders, such as lactated Ringer's infusion, should commence immediately with the intention of maintaining the systolic blood pressure between 90 and 100 mmHg if refractory shock is present, as this must be preserved for successful angiographic embolization. An immediate surgical consult should be called, and the patient should go to the UR. If the patient is hemodynamically stable, there should be an expeditious CT scan of the abdomen and pelvis obtained post up-right portable chest X-ray to safely rule out an intraperitoneal hemorrhage prior to going for any increasing level of sedation or anesthesia. Stable patients with solid organ hemorrhagic injury seen on pre-operative CT scan at investigation of active trauma behave differently from those on whom angiographic embolization is obtained prior to going to surgery. Patient gender should be factored into presentation guidance, as in cases where splenic artery aneurysm (SAA) rupture causes quick and massive volume loss there may be pre-existing etiological factors unseen at presentation.

Management Guidelines

Interpretation of splenic artery aneurysms and fissions varies widely among different studies, which cover several different types of lesions. When only true splenic artery aneurysms were

included, the rupture rates were considerable heterogeneous, ranging from 6.67% to 75%, with the median rupture rate being around 29.73%. The rate of ruptured splenic artery aneurysms climbed significantly following pregnancy, based on spontaneous rupture, possibly due to hemodynamic changes [23]. From the diagnosis and few treatment methods of the splenic artery aneurysm rupture, some general guidelines can be construed. Before splenic artery aneurysm rupture clinics or routine procedures, the physicians must discriminate their cases carefully through sufficient screening.

Because most symptomatic patients need surgeries and there is no ideal approach to overcome splenic artery aneurysm rupture's associated risk, the detection and surveillance of patients with splenic artery aneurysms is paramount. If a patient is suspected of splenic artery aneurysm rupture and shows serious symptoms, imaging studies, especially US, should be performed as soon as possible, and the surgical or interventional treatment should be well arranged by experienced surgeons before imaging studies finish. If a patient has a stable condition or a possible diagnosis of splenic artery aneurysm without rupture, the imaging studies could be performed before treatment [24].

The imaging studies could begin with an electronically expanded US examination, and if the US suspicion is high, CT or MRI can be considered in turn. If the imaging studies show some abnormalities such as a representative renal artery lesion or a known renal artery dissection and suspicious splenic artery aneurysm, those lesions should be evaluated carefully and treated potentially if considered unmanageable by the vascular specialists. If no vascular lesions are suspected but there were significant symptoms, symptomatic etiologies such as malignancy or stone diseases could be considered, and treatment should be addressed as needed. Otherwise, conservative treatment with analgesics should be likely sufficient.

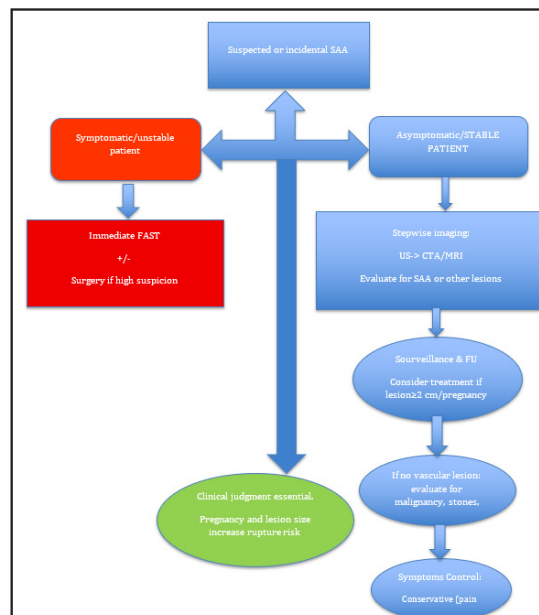


Figure 9: Management Algorithm for Splenic Artery Aneurysm (SAA).

This Decision-Making Algorithm Summarizes the Evidence-Based Management of Suspected or Confirmed Splenic Artery Aneurysms, Emphasizing Clinical Stability, Imaging

Pathways, and Treatment Modalities (from current PubMed and Cochrane literature)

Role of Multidisciplinary Teams

Management of ruptured SAAs requires a coordinated approach involving multiple specialties. Because of the critical workup, preparation, and treatment needed for SAAs, a multidisciplinary team (MDT) is often needed for major trauma patients. Although it is less common for patients to present with SAA ruptures, the complexity of care needed to stabilize and treat these patients warrants collaboration by many specialties. Typically, patients with ruptured SAAs present to the emergency department (ED), often in extreme pain. In the ED, a traumatic rupture can mimic a splenic rupture; in fact, patients typically share many common symptoms [25].

A stable patient may undergo surgical CT scans to determine their ailment, while a hemodynamically unstable patient may undergo FAST. On arrival to CT, the technologist will need to check the order carefully to obtain an appropriate study. Attending radiologists and the on-call trauma surgeon typically obscure the analysis of the scan and surgical planning in the discussion broadcasted over the trauma channel. If a rupture is consistent with a traumatic rupture, the trauma surgeon may mobilize the trauma team to the OR pre-emptively while the scan is still underway and the formal report is being produced. If SAAs are discovered on CT intervention radiology (IR) is contacted for repair via embolization. Otherwise, the case will be put on the trauma list for a splenectomy. If there is too much blood in the abdomen or if the spleen has already ruptured, IV fluids will be aggressive, and the trauma team will call a blue surgical or trauma, mobilizing the entire surgical and anesthesia team as well as all anesthesia attendants [26].

As the patient is being moved to the OR, consultation may be requested for urology and vascular. The patient will then be prepped and draped in the OR. This is a level one procedure where circulating nurses will have monitoring, resuscitation flowsheets, I.V. medications, and anesthesia setup also expanded [27].

Future Directions in Research

New perspectives on the aggressiveness of hemoperitoneum, primarily due to rupture of splenic artery aneurysms, are gaining traction among researchers. By conducting more basic research, it is possible to more accurately classify the SAA rupture (definitive or suspected), segment (partial dorsal, complete dorsal, or ventral), or type (cystic, broad neck, etc.), which can provide a more accurate prediction of the aggressiveness of hemoperitoneum and possibly establish treatment standards and the possibility of a “conservative” strategy. Though various courses have different aggressiveness, the treatment of SAAs is primarily invasive. Confirming suspicion of SAA rupture involves CT scan or MRI. Classic angiography is useful for the definitive diagnosis of ruptured SAAs. Though these imaging techniques are proficient, complications can frequently occur. Soon, more basic research is anticipated to be able to more accurately diagnose SAA rupture (total or partial) with no or less invasive techniques [28].

Health care costs related to acute ruptures of SAAs are massive, yet there are no established standards. Standardized procedures across healthcare centers are anticipated to lead to improved outcomes. The timely diagnosis of a ruptured SAA remains a challenge for emergency physicians and radiologists. Though

there have been advances in imaging techniques, these are not broadly available in their entirety. More successful detection protocols, based on basic research through artificial intelligence and machine learning, are anticipated in the future [29].

Final Thoughts and Conclusion

SAAs, which are the third most common form of splanchnic artery aneurysm, pose a significant threat to life. These are often asymptomatic, with fewer than 5% being symptomatic, mainly with pain or discomfort in the upper left quadrant, and splenic rupture, which can be complicated by shock or abdominal hemorrhage. Ruptured SAAs are present most commonly to the emergency department, where requests for CT and ultrasound imaging might be triggered by abdominal pain. CT offers the advantage of depicting the CT triad classically associated with these entities, which consists of splenic hematoma, foci of high attenuation along the splenorenal ligament, and peritoneal hematoma. Other cross-sectional imaging modalities include MRI and DSA, particularly when embolization or surgical intervention is planned. Apart from the classical triad of findings on CT imaging, a few post-contrast imaging features are highly suggestive of splenic artery aneurysm suppuration. These include an irregular, lobulated/enhancing mass adjacent to the periphery of the anterior segment of the spleen, marked stranding and enhancement around the mass correlating with thickening of the vascular supply on CT MRA, and the presence of high-density foci within the mass correlating with calcific debris. Immunocompromised patients depending on whether they are on steroids or have undergone irradiation.

The ruptured aneurysms in symptomatic cases are much larger than asymptomatic ones, appropriately suggesting screening and follow-up of those with larger more perilous ones. Referring to conventional angiography is preferred for better delineation of vascular anatomy, simultaneously allowing treatment, but the CT-derived perfusion method can gain similar plethysmographic data regarding SAA. With their low level of invasiveness, stent-graft exclusion is becoming the method of choice.

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