Review

Splenic artery aneurysms in pregnancy – A systematic review

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\textbf{ABSTRACT}

Visceral artery aneurysm is an uncommon pathology, with a potential for rupture. Splenic artery aneurysms (SAA) are most commonly (60\%) associated with a high mortality rate of 25\% in case of aneurysm rupture. This increases disproportionately to 75\% among pregnant women with fetal mortality of 95\%. Although this is a rare event, because of the associated catastrophic consequences, prompt management of splenic artery aneurysms (SAA) is of prime importance. This systematic review provides up-to-date information about the management of splenic artery aneurysms in pregnancy.

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\section*{1. Introduction}

Visceral artery aneurysm is an uncommon pathology, with a potential for rupture. Splenic artery aneurysms (SAA) are the most common (60\%), followed by hepatic (20\%), superior mesenteric (5.9\%) and celiac (4\%) artery aneurysms.\textsuperscript{7} SAAs are either discovered incidentally\textsuperscript{2–5} or in symptomatic cases when they rupture.\textsuperscript{6–8} The latter is associated with a 25\% mortality\textsuperscript{9,10} which increases disproportionately to 75\% among pregnant women with fetal mortality of 95\%.\textsuperscript{9–11} The fetal distress and mortality usually results from hypovolemia, shock and their resulting consequences.

Various anatomical and physiological changes occur during pregnancy, which can alter the presentation of conditions that require surgery making the diagnosis, evaluation and treatment of a pregnant woman difficult. We present a comprehensive systematic literature review of the pathophysiology, clinical features, diagnosis and treatment options of this rare and potentially fatal condition.

\section*{2. Methods}

Two reviewers did a comprehensive systematic English language literature search using PUBMED and EMBASE search engines independently. The search terms used were “Splenic artery aneurysm” and “Pregnancy”. All related case reports, case series, review articles which were published between April 1976 and April 2007 were reviewed systematically.
3. Demographics

The true prevalence of SAA in pregnancy or women of childbearing age is unknown. This is because most of them are asymptomatic. The prevalence data in general population is also variable ranging from 0.1–0.2% in a large autopsy series to 10.4% in an autopsy series on patients 60 years or older with special attention given to the splenic artery. In a study of 3600 arteriograms the incidence was found to be 0.78%. Parity, however, seems to influence the incidence of SAA in women, being four times more common in multiparous women. A significant number of women are pregnant when the diagnosis is made probably due to augmented use of ultrasonography and use of high-resolution cross-sectional imaging techniques. Two-thirds of aneurysms rupture in third trimester followed by second trimester ruptures.

4. Pathophysiology

The precise etiology of splenic aneurysms remains unclear. Different etiological factors attributed to aneurysm formation include angiodysplasia, portal hypertension, pregnancy, atherosclerosis, diabetes, intracranial aneurysm, polyarteritis nodosa, alpha-1-antitrypsin deficiency and infective factors. Among them pregnancy has a strong association with splenic artery aneurysm formation.

Different mechanisms have been proposed to play a role in the pathogenesis. Trimble and Hill’s proposals remain even valid today, when they suggested that aneurysmal dilatation of an artery results from two contributing factors: weakness in the arterial wall and increase in the blood pressure.

In pregnancy, influence of hormones namely estrogen, progesterone and relaxin on the arterial wall plays a significant role. The first two hormones presumably result in histological alteration of the arterial wall mainly medial degeneration leading to aneurysmal dilatation. Histological changes that have been documented include sub-endothelial thickening, internal elastic lamina fragmentation, medial fibrolysplasia, and accumulation of acid glycosaminoglycans in both subintimal and medial layers. Microcystic degeneration and external elastic lamina disruption have also been reported, the latter in patients with portal hypertension. Relaxin may augment the effect of these hormones by further enhancing the elasticity of splenic artery as suggested by de Vries et al. In patients having portal hypertension, which can be concomitantly present in pregnancy, hormones like aldosterone and rennin have also been implicated to cause arterial wall thinning. Concomitant presence of alpha-1-antitrypsin deficiency can further increase the likelihood of aneurysm formation.

Physiological changes during pregnancy also enhance the stress on the arterial wall. These include enhanced cardiac output, increased blood volume and portal hypertension. The collative effect of these changes is increased chance of splenic aneurysm formation and/or rupture in pregnancy. Other most commonly reported arteries to have aneurysm rupture during pregnancy include aorta, cerebral arteries, renal, coronary and ovarian arteries.

4.1. Pathomechanics of rupture

The frequency of rupture increases in pregnancy and with associated portal hypertension, the former affecting 20–50% of women. Early reports of splenectomy aneurysm suggested the risk of rupture to be 10%, however, more recent data suggest rupture rates closer to 2–3%. The size of the aneurysm is usually more than 2.5 cm in most patients at the time of rupture, however, rupture of smaller aneurysms has also been reported. This rupture can be either sudden rupture or a two-stage rupture, which is present in 20–25% of cases. The latter is characterized by containment of initial rupture within the lesser sac by either omentum and/or blood clots that block foramen of Winslow. This is followed by free rupture into greater sac when the tension within the lesser sac increases. The clinical presentation is in the form of sudden initial abdominal pain with an intermediate stable time period followed by sudden collapse. There has been only one report in literature, which describes rupture of splenic artery into splenic vein in a patient with acute portal hypertension in the post-partum period.

5. Diagnostic features

The clinical features result from aneurysm rupture usually which is characterized consistently with sharp abdominal pain, either in the epigastrium or more often localized in the left hypochondrial area with associated pain in the tip of the left shoulder (Kehr’s sign). This is associated with nausea, vomiting and sudden collapse. Abdominal tenderness can be elicited in cases of rupture though signs of shock are the most suggestive features. Abnormal cardiotocogram (CTG) tracing is also witnessed in cases of advanced pregnancy. Fender and colleagues have reported a case in which clinical signs of rupture were masked by epidural anesthesia used during delivery. Richardson and colleagues reported a case of SAA rupture in pregnancy presenting in a manner similar to pulmonary embolism characterized by left sided chest pain, breathlessness, low oxygen saturation and ECG changes indicative of massive pulmonary embolus. Therefore, high degree of suspicion is required when making the diagnosis in pregnancy with confirmation usually made at the time of laparotomy. Placental abruption is one of the most commonly made misdiagnosis; however, uterine rupture and rupture of other arterial aneurysms may share similar clinical features.

The radiological investigations are quite helpful in making the diagnosis in both emergency and elective settings; however, their sensitivities and specificities remain unknown in pregnancy. Moreover, they should not delay the immediate resuscitation and control of the hemorrhage by emergency surgery. It should also be borne in mind that use of investigations that involve exposure of the fetus to radiations should be kept to a minimum because of the danger of teratogenicity.
5.1. Abdominal X-ray

Although it is not the first line investigative tool for SAA, abdominal X-ray carried out for some other abdominal pathology may reveal calcified SAA as characteristic calcified ring with a central lucent area to the left of the first lumbar vertebral body.45

5.2. Ultrasonography and Doppler ultrasonography

It is preferable in pregnancy23 because it is non-invasive and cost effective. In emergency cases it usually reveals presence of free fluid in abdomen and the diagnosis confirmed at laparotomy. There are no contraindications to ultrasound procedures during pregnancy and this modality has largely replaced X-ray as the primary method of fetal imaging during pregnancy.46 There have been no reports of documented adverse fetal effects for diagnostic ultrasound procedures, including duplex Doppler imaging. However, its utility is limited by operator dependency, obese patients, bowel gas shadow and arteriosclerosis.47,48 The likelihood of missing smaller lesions is also quite high because of limited spatial resolution.49,50

5.3. Computerized tomography (CT) and magnetic resonance (MR) imaging

Although contrast-enhanced CT and MRI are quite useful in improving three-dimensional evaluation of the splenic artery, readily distinguishing tortuous vessel from aneurysm, however, in pregnancy safety of the fetus has to be given due consideration while considering their use. There have been no documented adverse fetal effects reported, however, the National Radiological Protection Board arbitrarily advises against use of MRI in the first trimester.46 Similarly, teratogenesis is not a major concern after diagnostic CT studies in pregnancy, because the radiation dose is generally too low to cause such effects.51

However, intravascular contrast media should be avoided in pregnancy, in order to avoid any possible hazard to the fetus52 and should only be used if absolutely essential, and only after discussion of risks and benefits with the patient and referring clinician.

5.4. Digital subtraction angiography (DSA)

Though DSA is the gold standard for diagnosis of SAA, however, in pregnancy it is usually employed when concomitant radiological intervention like coil embolization or endoluminal stent deployment is to be performed.57,58 Nincheri and colleagues suggest transcatheter embolization or percutaneous embolization to urgently stop hemorrhage.53 To date no reports were found in literature where these treatment modalities were used in ruptured SAA in pregnancy. As far as safety of DSA is concerned, the above-mentioned rules for use of contrast media also hold true for DSA in pregnant women.

6. Management

6.1. Asymptomatic SAA

There is no consensus with regard to management of asymptomatic cases. But it is suggested that aneurysms that are larger than 2 cm should be treated.15,16 In pregnancy, minimally invasive techniques should be utilized which can be transcatheter embolization, percutaneous angiographic embolization or laparoscopic ligation or resection.53 However, appropriate treatment of SAA depends on location of the lesion, age of the patient, operative risks and clinical status.49

The preservation of spleen should be a consideration in the treatment of SAA.54 Aneurysms located in the proximal or middle third of the splenic artery may be treated with simple excision, with proximal and distal ligation of the artery and splenic preservation (through the short gastric vessels). For aneurysms located in the distal third, resection with splenectomy is most often performed, which is unfortunately the case in 70% of the patients with portal hypertension.17,54 The operative mortality is greater than 0.5% in elective cases.14 The use of embolization does significantly decrease the mortality and morbidity but data from long term follow up of these patients are not available and is believed to be less successful in 85% of the cases.8,53

When no pregnancy is planned and aneurysm is less than 2 cm, management options are inconclusive because the understanding of natural history of SAA is incomplete. However, in pregnancy proactive management of aneurysms less than 2 cm should be carried out.

6.2. Symptomatic and ruptured SAA

In symptomatic cases, immediate treatment is warranted which can be open, laparoscopic or as embolization of the aneurysm.53 However, in cases of rupture, the aim is immediate resuscitation and cessation of bleeding, which is usually through cesarian section laparotomy. Splenectomy or splenopancreatectomy is usually employed in these cases with ligation of the splenic artery.5

7. Conclusion

Although observant treatment can be adopted in aneurysms less than 2 cm in non-pregnant patients, however, keeping in view the catastrophic consequences of rupture in pregnancy, proactive approach should be adopted in women of childbearing age. More research is required in understanding the natural history of splenic aneurysms and their response to hormonal influence in controlled environment before a definite management strategy can be agreed upon in elective setting. In emergency settings, high degree of suspicion is required to make diagnosis of SAA rupture in pregnant females having sudden collapse with or without sharp abdominal pain. Immediate resuscitation and cessation of hemorrhage is essential for maternal and fetal survival. National health authorities should maintain their SAA databases for rupture and elective cases and report their results annually to further improve management strategies, and
obstetricians should involve vascular surgeons and interventional radiologists as soon as they suspect SAA.

Conflict of interest
The authors have no conflict of interest.

Funding
None.

REFERENCES


