Hypoplasia of the Spleen: Review of Pathogenesis, Diagnosis, and Potential Clinical Implications

Iswanto Sucandy, Harsha V. Polavarapu, Christopher M. Pezzi

Department of Surgery, Abington Memorial Hospital, Abington, Pennsylvania 19001, USA

Abstract

Context: Splenic aplasia is seen when the spleen is congenitally absent, has been surgically removed, or becomes atrophic secondary to episodes of arterial/venous occlusion, which result in splenic infarction. This rare condition is caused by a heterogeneous group of diseases, which may present a wide spectrum of clinical manifestations. Splenic hypoplasia is defined as reduction in splenic mass and functions caused by incomplete splenic development or secondary parenchymal involution. Splenic infarction may be clinically silent and only discovered incidentally during abdominal exploration for other conditions. Case Report: We present an unusual case of hypoplastic spleen with calcifications, which was preoperatively found during radiologic workup for gastric carcinoma. An 88-year-old woman presented with coffee-ground emesis. Her past medical history was only significant for atrial fibrillation. Esophagogastroduodenoscopy demonstrated gastric carcinoma, for which a subtotal gastrectomy was planned. Preoperative computed tomography scan showed a hypoplastic spleen with calcifications in the left upper quadrant. Symptoms of immunologic deficiency were not present. During laparotomy, an atrophied and calcified spleen was identified and left in situ. The patient made an uneventful postoperative recovery. Splenic hypoplasia is an unique entity, which may be seen in the setting of atrial fibrillation and abdominal malignancy. Conclusion: Splenic hypoplasia may be detected incidentally during radiologic workup or abdominal exploration. Abdominal symptoms or immunologic deficiency are not always present.

Keywords: Diagnosis, hypoplasia, implications, pathogenesis, splenic aplasia

Address for correspondence: Dr. Iswanto Sucandy, Department of Surgery, Abington Memorial Hospital, 1200 Old York Road, Abington, Pennsylvania 19001, USA. E-mail: iswanto_sucandy@yahoo.com

Introduction

Splenic function is compromised when the spleen is congenitally absent, has been surgically removed, or becomes atrophic from repeat episodes of arterial/venous occlusion causing a splenic infarction. Splenic infarction occurs when the splenic artery or one of its major branches becomes occluded. In the modern era of percutaneous endovascular intervention, arterial embolization of the splenic artery has become the preferred treatment modality for many conditions such as traumatic splenic injury, massive variceal bleeding from splenic vein thrombosis, sinistral portal hypertension, or splenic sequestration.\(^1\) In a large autopsy series by O’Keefe et al. in 1986, only 10% of the splenic infarctions were diagnosed antemortem.\(^4\) This condition is caused by a heterogeneous group of diseases and can present a wide spectrum of clinical manifestations. Splenic infarction which may leads to splenic hypoplasia could be spontaneous, clinically silent, or iatrogenic (such as seen in the more widely accepted and performed splenic artery embolization during splenic hemorrhage).\(^1\) Splenic hypoplasia may only be discovered incidentally during an abdominal exploration for other conditions. In other cases, splenic infarction may produce subacute or acute abdominal symptoms. We present an unusual case of incidentally found hypoplastic spleen during workup and treatment of gastric carcinoma.

Case Presentation

An 88-year-old Caucasian woman presented with coffee-ground emesis. Her past medical history was significant for atrial fibrillation. Esophagogastroduodenoscopy
demonstrated the presence of a gastric adenocarcinoma, for which open subtotal gastrectomy was planned. Preoperative computed tomography (CT) showed an aplastic spleen with calcifications in the left upper quadrant [Figure 1]. Symptoms of immunologic deficiency were not present. During laparotomy, an atrophied and calcified spleen was identified [Figure 2]. The aplastic spleen was left in place. The patient made a full postoperative recovery without any complications.

Discussion

In a study of 49 patients with splenic infarction, Antopolsky et al. reported that, predisposing factors to infarction were present in 71% of the patients.[5] The most common predisposing factor was atrial fibrillation, which occurred in 23% of the reported cases. Less common factors included antiphospholipid syndrome, previous history of thromboembolism, use of oral contraceptives, previous heart valve surgery, congenital or acquired hematologic disorders, nonhematologic cancer, and severe liver disease.[5] The mechanism of splenic infarction is identical for both benign and malignant etiologies. While hereditary deficiency of protein C is one of the congenital causes of hypercoagulability resulting in splenic infarction, myeloproliferative disorders, lupus anticoagulant, polycythemia vera, and malignancy are more common. In clinical settings, splenic thromboembolism is associated with diverse cardiovascular conditions including atherosclerosis, acute myocardial infarction with left ventricular mural thrombus, atrial fibrillation, or bacterial endocarditis. Splenic artery embolism could also present as a complication of cardiac catheterization.

Görg et al. reported a series of 10 patients with splenic infarction secondary to cancer, which included two patients with pancreatic adenocarcinoma and one patient with neuroendocrine tumor. All of the 10 patients were asymptomatic. The splenic infarction was incidentally diagnosed during routine ultrasonography (US) for tumor staging. In patients with pancreatic cancer, torsion of the splenic vessels, direct vessel invasion by the tumor, and extrinsic compression have been reported to cause a splenic infarction. When cancer is the underlying etiology, complete, instead of partial parenchymal infarction is more commonly seen. This malignant pathology often results in a small underperfused hypodense spleen.[6]

Only 69% of patients with this condition are symptomatic. The most common presenting symptom is left upper quadrant abdominal pain or discomfort. Other less frequent symptoms include fever, pleuritic chest pain, back pain, left shoulder pain, and increased abdominal girth due to splenic congestion (seen in pediatric populations). In 16.6% of patients, the diagnosis of splenic infarction was the first sign, which led to the diagnosis of underlying conditions, such as hypercoagulable state, atrial fibrillation, and polycythemia vera.[7] A large number of experimental and clinical studies have contributed to current understanding of the physiology of the spleen and its role in the immunological defense against bacterial infection.[8-10] Overwhelming postplenectomy sepsis (OPSI) has been the most feared potential complication after splenectomy. Vilde reported 0.6% incidence of death caused by OPSI in a series of 288 patients with splenic trauma.[11] In another adult series by Vichard and Dreyfus-Schmidt, 2.2% of patients experienced fatal OPSI following trauma splenectomy.[12] In cases of splenic aplasia or hypoplasia, however, symptoms of immunodeficiency are not always observed, such as in this case.

Computed tomography scan with intravenous contrast is currently the most effective noninvasive test available to diagnose splenic infarction. CT scan...
has the advantage of showing the infarcted spleen and other target organs, the extent of thrombosis, in addition to the potential underlying source of the problem. The most common finding is an area of decreased attenuation within the splenic parenchyma, which is not always wedge-shaped. US has lower sensitivity, but it may be used to differentiate hypogenic acute infarction from a hypercoechoic healed lesion with or without calcifications. US can also be used to quickly evaluate the evolution of a splenic infarction into an abscess. CT scan and ultrasound have supplanted the use of traditional radionuclides studies for detection of splenic infarction.\(^7\)

Splenic infarction by itself is not an indication for a splenectomy unless persistent symptoms or complications such as splenic congestion, rupture, large cystic degeneration, or parenchymal abscess develop.\(^{[13,14]}\) Laskou et al. in their retrospective study of 167 patients have categorized indications for splenectomy into seven groups: Splenic trauma, hematological diseases, malignancies, congestive and vascular diseases, symptomatic splenic cysts, iatrogenic rupture, and inflammatory diseases.\(^{[14]}\) In the current case, we hypothesize that the underlying atrial fibrillation and hypercoagulability related to gastric carcinoma may have caused the subclinical splenic infarction. The nonperfused spleen gradually became ischemic and atrophied. Cellular ischemia subsequently caused loss of membrane integrity, which is followed by calcium influx into the cell. The regenerative process as part of wound healing then continued with deposition of collagen by fibroblasts and other extracellular matrix components, causing the formation of fibrotic tissue. The end product of these processes was an involuted, fibrotic, and hypoplastic spleen with calcific changes.

**Conclusion**

Splenic hypoplasia is an interesting entity, which may be seen in the setting of atrial fibrillation and abdominal malignancy. Splenic hypoplasia may be detected incidentally during radiologic workup or abdominal exploration, because abdominal symptoms or immunologic deficiency are not always present.

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