Arteriovenous Fistula of the Splenic Vessels Producing Ascites

By William G. Cassel, John A. Spittel, Jr., F. Henry Ellis, Jr., and André J. Bruwer

Arteriovenous fistula involving the portal vein or its tributaries is a rare condition that in some cases can be treated successfully by surgery if it is recognized. The clinical manifestations and diagnostic criteria are presented.

The opportunity to cure ascites is a gratifying clinical experience but is seldom encountered. Recently we have observed and successfully treated a patient with ascites and an arteriovenous fistula of the splenic artery and vein.

Arteriovenous fistulas may occur wherever arteries and veins lie in proximity. The usual clinical manifestations, such as thrill, machinery murmur, increased venous pressure with its sequelae in the involved region, and at times cardiac enlargement and failure, are well known. An arteriovenous communication between a systemic artery and the portal vein or one of its tributaries quite logically might produce portal hypertension and its sequelae. Sigwart1 reviewed the literature prior to 1953 and found 3 cases2-4 of splenic arteriovenous fistula reported in detail and another5 mentioned, and he added 1. In 1955 Stener6 reported a case. Of these, only 1, the patient of Goodhart,3 had ascites, while 4 patients presented with repeated gastrointestinal bleeding from esophageal varices.1, 3, 4, 6 Two instances of hepatoportal arteriovenous fistula have been reported7, 8 and 1 of these patients had ascites.7

Case Report

A 34-year-old housewife came to the Mayo Clinic on October 29, 1956 because of swelling of the abdomen of 4 to 5 weeks' duration. Three months prior to admission her third pregnancy was terminated normally. The pregnancy was complicated by hypertension and edema of the ankles during the eighth and ninth months. For 2 months post partum she felt well. Four to 5 weeks prior to admission she noted rapid increase in the size of her abdomen unaccompanied by dyspnea, peripheral edema, fever, or pain. Two to 3 weeks prior to admission anorexia nausea, and vomiting developed and she noted a full sensation in her abdomen. X-ray studies revealed the presence of calcified cyst-like structures in the left upper quadrant of the abdomen. Laparotomy was performed on October 26, 1956, with a preoperative diagnosis of pancreatic cyst. After 4,500 ml. of straw-colored ascitic fluid were removed, the left kidney was seen to be depressed by a cystic mass that was said to involve the left kidney and to be fixed to the spleen. Aspiration of the mass disclosed blood. Further exploration was not performed and the abdomen was closed. Three days postoperatively the patient was referred to the Mayo Clinic for further diagnosis and treatment.

The history revealed that 11 years prior to admission the patient had sustained a bullet wound of the left lower part of the thorax. The bullet entered anteriorly at the sixth intercostal space in the midclavicular line, and came out at the costal margin in the left posterior axillary line. The patient was hospitalized and treated conservatively, and recovered in 2 weeks.

Physical examination revealed a slender, chronically ill young woman whose weight was 135 pounds. Entrance and exit scars of the old bullet wound were present on the left lower part of the thorax. There was a recent left abdominal incision. The abdomen was distended, with evidence of free fluid. The edge of the liver, felt 1 to 2 fingerbreadths below the right costal margin, was smooth and nontender. The tip of the spleen was palpable. A continuous multiple-pitched murmur was audible over the left posterolateral part of the thorax about 2 cm. lateral to the bullet scar. The results of physical examination were otherwise negative. There was no peripheral edema, distention of the veins of the neck, or spider angiomas.

Laboratory studies revealed the following: moderate albuminuria with microhematuria; hemoglobin, 12 Gm. per 100 ml. of blood; leukocyte count, 12,100 per mm.3; erythrocyte sedimentation rate, 65 mm. in the first hour (Westergren); flocculation reaction for syphilis, negative; fasting blood sugar, 128 mg. per 100 ml.; blood urea, 38 mg. per 100 ml.; serum bilirubin, negative reaction direct, and 0.9 mg. per 100 ml. indirect; sulfobromophthalein (Bromsulphalein) retention after 1 hour, grade 3 (34 per cent); thymol turbidity, 0 unit; cephalin-cholesterol flocculation, negative; alkaline phospha-
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Fig. 1. a. Roentgenogram of the abdomen showing crescentic areas of calcification in the left upper quadrant. b. Aortogram showing enlarged tortuous splenic artery and opacification within the calcified areas. c. Aortogram following second injection of dye. There is opacification of the venous aneurysm adjacent and medial to the calcium-containing aneurysms. d. Roentgenogram taken 10 seconds after the one represented in c, showing enlarged splenic and portal veins.

tase, 58 King and Armstrong units per 100 ml. of serum; serum proteins, 7 Gm. per 100 ml., with 5 Gm. of albumin and 2 Gm. of globulin.

Roentgenographic examination did not disclose any abnormality of the thorax, but did show linear calcification in the shape of an incomplete oval, measuring 4 by 3 cm., in the left upper quadrant of the abdomen.

Excretory urography showed 2 egg-shaped areas of calcification 4 cm. in diameter in the left suprarenal area (fig. 1a). These were thought to be extrarenal, and the upper pole of the left kidney appeared to be compressed as if by a suprarenal mass. Left retrograde pyelographic examination gave no additional information.

Abdominal paracentesis yielded 3,000 ml. of serosanguineous fluid. Culture of the ascitic fluid for bacteria and examination of it for malignant cells gave negative results.

The ascites, the classic murmur of an arteriovenous fistula over the left lower part of the thorax, the calcifications in the left upper quadrant of the abdomen as revealed roentgenographically, and the history of a bullet wound of the left lower part of the thorax 11 years previously suggested a splenic arteriovenous fistula.

Aortography was performed to confirm the clinical impression. Because of the possible danger of passing a needle into an aneurysm, it was decided that aortography performed via a catheter in the
aorta would be safer than translumbar aortography. However, an attempt to pass a catheter down the descending thoracic aorta via the left brachial artery was unsuccessful. Routine translumbar aortography was therefore performed, the needle being passed into the aorta at the level of the first lumbar vertebra. When the tip of the needle was about half way between the skin and the aorta, blood was aspirated from a low pressure site; it seemed probable that the venous sac of the arteriovenous aneurysm was traversed.

After the needle had been placed in the aorta, two injections of 70 per cent acetrizoate (Urokon) sodium were made. After the first injection, consisting of about 10 ml., the roentgenogram (fig. 1b) revealed an enlarged, tortuous splenic artery and dense opacification within the calcified areas in the left upper quadrant. At the end of the second injection, consisting of about 35 ml. of opaque material, the roentgenogram showed that a large uncalcified aneurysm had become opacified adjacent and medial to the calcium-containing aneurysms (fig. 1c). Another roentgenogram (fig. 1d) taken about 10 seconds after the end of the second injection demonstrated that the opaque material that had pooled in the uncalcified area emptied into the splenic vein, and that the splenic and portal veins were considerably enlarged. It was evident from these studies that

**Fig. 2.** Biopsy specimen from the liver. The parenchyma is normal (hematoxylin and eosin; X 200).

**Fig. 3.** Drawing of gross specimen.
the calcified shadows in the left upper quadrant were related to the arterial aneurysm and that the even larger venous aneurysm contained no calcium.

All other branches of the abdominal aorta were of normal caliber.

Operation was undertaken on the ninth day after admission. A left thoraco-abdominal incision was made, the old scar being excised and the incision being continued laterally over the costal arch and into the thorax through the seventh intercostal space. The diaphragm was opened partially in the line of the incision.

The liver was tremendously enlarged and the left lobe extended laterally well under the left leaf of the diaphragm. The liver was smooth in contour. Biopsy showed normal hepatic parenchyma (fig. 2). The gastrohepatic omentum was opened and an enlarged, tortuous splenic artery was visualized; this was doubly ligated near its origin. Immediately the continuous murmur audible to the anesthesiologist over the left posterior part of the thorax disappeared. The veins in the mesentry of the small intestine were not dilated. The remainder of the abdomen at exploration appeared normal except for the region of the spleen.

In the hilar region of the spleen a lobulated aneurysm of the splenic artery was visualized, and associated with it was a much larger aneurysm of the splenic vein; the latter aneurysm lay anteriorly and somewhat inferiorly (fig. 3). The spleen was densely adherent to the diaphragm and was dissected away with some difficulty. It was eventually retracted upward and medially, whereupon the aneurysms of the splenic vein and artery were thoroughly exposed. As the dissection continued, a portion of the splenic vein was reached that, although enlarged, was small enough to permit ligation and division. The splenic artery was likewise ligated and divided. It was necessary to remove a portion of the tail of the pancreas as well.

The excised specimen therefore consisted of the spleen, the aneurysms of the splenic artery and the splenic vein, and a portion of the tail of the pancreas. When the specimen was opened, a communication 4 mm. in diameter was found between a bilocular aneurysm of the splenic artery and an aneurysm of the splenic vein (fig. 4). Each locule of the arterial aneurysm measured 4 cm. in diameter, and the venous aneurysm measured 8 by 5 by 5 cm.

The cut end of the pancreas was oversewn, the diaphragm was closed, the costal arch was reapproximated, and the thorax and abdomen were closed.

The patient's postoperative course was uneventful. The mild diabetes, as evidenced by the increased concentration of blood sugar, was managed without incident by a diabetic diet. The platelet count rose from 240,000 per mm.³ on November 10 to 572,000 on November 13 and to 653,000 on November 16. A routine urinalysis on November 9 gave negative results. The serum bilirubin reaction remained normal postoperatively, and the concentration of alkaline phosphatase decreased to 38.7 King and Armstrong units per 100 ml. of serum by the tenth postoperative day. Sulfobromophthalein retention at the end of 1 hour on the ninth postoperative day was grade 1 (6 per cent). Three months postoperatively the patient said that she was well and had noted no further ascites.

DISCUSSION

The mechanism of formation of ascites is not fully understood. Volwiler, Grindlay, and Bollman observed in dogs with normal plasma
Table 1.—Features of Reported Cases of Splenic Arteriovenous Fistula

<table>
<thead>
<tr>
<th>Author</th>
<th>Age and sex</th>
<th>Gastrointestinal bleeding</th>
<th>Ascites</th>
<th>Bruit</th>
<th>Site of fistula</th>
<th>Miscellaneous</th>
</tr>
</thead>
<tbody>
<tr>
<td>Weigert² (1886)</td>
<td>49 F</td>
<td>Repeated hematemesis and melena, 30 mo.</td>
<td>Present</td>
<td>Not stated</td>
<td>Hilus of spleen</td>
<td>Cause of death not given. Multiple arteriovenous communications at necropsy.</td>
</tr>
<tr>
<td>Goodhart³ (1889)</td>
<td>49 F</td>
<td>Repeated hematemesis, 20 yr.</td>
<td>Absent</td>
<td>Not stated</td>
<td>2 in. from hilus of spleen</td>
<td>Died from hematemesis. Single arteriovenous communication at necropsy.</td>
</tr>
<tr>
<td>Blakemore⁴ (1948)</td>
<td>23 F</td>
<td>Absent*</td>
<td>Present</td>
<td>Not reported adequately</td>
<td>In spleen</td>
<td>Died postoperatively. Multiple arteriovenous communications. Portal pressure 350 mm. of water.</td>
</tr>
<tr>
<td>Sigwart¹ (1953)</td>
<td>25 F</td>
<td>Repeated hematemesis, 9 yr.</td>
<td>None</td>
<td>Present</td>
<td>Hilus of spleen</td>
<td>No gastrointestinal bleeding after removal of arteriovenous fistula and spleen. Portal pressure at operation 430 mm. of water. Single communication.</td>
</tr>
<tr>
<td>Stener⁶ (1955)</td>
<td>38 F</td>
<td>Repeated hematemesis and melena, 20 yr.</td>
<td>Absent*</td>
<td>Present</td>
<td>Splenic hilar area, 2 cm. from spleen</td>
<td>No gastrointestinal bleeding after removal of arteriovenous communication and spleen. Multiple arteriovenous communications.</td>
</tr>
<tr>
<td>Authors’ case</td>
<td>34 F</td>
<td>Not stated</td>
<td>Present</td>
<td>Present</td>
<td>Spontaneous hemorrhage, 2 cm. from spleen</td>
<td>Ascites disappeared after removal of arteriovenous communication and spleen. Single arteriovenous communication.</td>
</tr>
</tbody>
</table>

* Ascites occurred postoperatively but was temporary.

protein that there had to be some element of hepatic congestion to produce marked spontaneous, progressive ascites; such congestion was produced by constriction of the thoracic inferior vena cava. In another group of dogs constriction of the inferior vena cava and portal vein produced slight to moderate ascites when the dogs were made hypoproteinemic by plasmapheresis. Baggenstoss and Wollaege⁵ found ascites in 5 of 15 necropsy cases in which portal hypertension was due to occlusion of the extrahepatic portion of the portal vein and in which there was no complicating disease that might in itself have been a cause of ascites. They concluded that portal hypertension per se was an important factor in the development of ascites in these cases. They found evidence that ascites was more likely to occur when the hepatopetal collateral circulation was inadequately developed.

Schilling and McKee¹¹ produced hepatopetal arteriovenous fistulas in dogs and found that ascites developed in only 1 of them, this occurring 30 months postoperatively; this animal had evidence of disturbed hepatic function, and microscopic examination of its liver revealed marked scarring and fatty infiltration in many places. Servello and Rossi¹² did not observe ascites in dogs in which they produced splenic arteriovenous fistulas; this fact may be related to the brief period of observation (45 days). They did, however, note hepatic congestion and microscopic evidence of dilatation of the hepatic vascular bed.

Besides our patient, only 1 other patient with arteriovenous communication of the splenic vessels has been reported as having ascites (Table 1). Bleeding from esophageal varices, often repeated over many years, was a more common clinical manifestation. Blakemore⁴ and Sigwart¹ found an increase of portal pressure in their patients. The typical bruit of an arterio-
Arteriovenous fistula has been noted in only 3 cases, including the present case.

The cause of the arteriovenous fistulas in the previously reported cases reviewed in table 1 is obscure. Pre-existing trauma is not mentioned in any of them. Three patients had multiple arteriovenous communications and 3, including ours, had a single communication. The existence of multiple communications suggests a congenital origin. In Sigwart’s case the communication was single but within the spleen and, in his opinion, was probably congenital. In the 2 cases with multiple communications and in which the history was reported, and in Sigwart’s case, evidence of portal hypertension (gastrointestinal hemorrhage) appeared in childhood or early adult life; this suggests a congenital anomaly. Goodhart’s patient, the only one other than ours who had ascites, was 49 years old and had hematemesis and melena for only 5 months prior to death; the single arteriovenous communication was likely acquired and possibly the result of rupture of an aneurysm of the splenic artery into the adjacent vein.

The close relationship of the development of ascites to parturition observed in our case suggests the possibility that an aneurysm of the splenic artery ruptured into the splenic vein as a result of changes in intra-abdominal pressure incident to parturition. Owens and Coffey have shown a relationship between pregnancy and rupture of a pre-existing aneurysm of the splenic artery. It is equally possible that the splenic arteriovenous fistula developed in our case at the time the bullet wound was inflicted 11 years prior to the onset of ascites.

Ascites in arteriovenous fistula involving the portal system could be produced by either or both of 2 factors—portal hypertension and hepatic congestion. As previously mentioned, portal hypertension was shown to exist in such cases by Blakemore and Sigwart at the time of operation. According to Baggenstoss and Wollaeger, portal hypertension in itself can cause ascites. In addition, transmission of arterial pressure to the portal system may cause hepatic congestion and ascites in a manner analogous to that in the experiments of Volwiler and co-workers in which occlusion of the thoracic inferior vena cava caused hepatic congestion and massive ascites. Significantly, those patients whose fistulas were presumably congenital did not have ascites. It is possible that in patients with multiple arteriovenous fistulas involving the portal system there is sufficient hepatofugal circulation during the developmental stage to prevent ascites.

Strickler and Madding, with their co-workers, have each recently reported a case of single communication between the hepatic artery and the portal vein, which would provide an analogous alteration in the hemodynamics of the portal vein, namely transmission of arterial pressure to the portal vein. The patient of Strickler and co-workers had ascites and hematemesis. The arteriovenous fistula was thought to have been acquired as a result of rupture of a congenital aneurysm into the portal vein, as may have occurred in Goodhart’s case. The patient of Madding and co-workers had gastrointestinal bleeding without ascites. These authors expressed the belief that the arteriovenous fistula was of congenital origin.

In 3 of the reported cases (table 1) and in our case surgical approaches were made. The fistula was successfully removed in 3. In the 2 reported cases of hepatoportal arteriovenous fistula operation was unsuccessful.

When calcium is present in an aneurysm of the splenic artery the diagnosis can be made with considerable confidence by routine roentgenologic methods. In 1932 Lindboe made this diagnosis roentgenologically, and since then the roentgenologic features of such aneurysms have been described repeatedly. There is no reason to believe that the arterial component of a splenic arteriovenous aneurysm, once calcified, should have a different appearance.

Calcium deposition in an aneurysm of the splenic artery usually is manifested as a linear opacity in the shape of a complete or incomplete oval representing shadows caused by the tangential trajectory of the beam. These crescentic densities enclose mottled, ill-defined, calcific densities, representing the regions where the roentgen rays have passed through the aneurysm en face.

Culver and Pirson have indicated that it may take 10 to 20 years for calcification to develop in the walls of an aneurysm of the splenic artery. Particularly in cases prior to calcification, abdominal aortography should be performed on the basis of clinical suspicion.
Furthermore, aortography should always be performed when the question of an intra-abdominal arteriovenous fistula arises, because by this means only can the extent of the uncalcified portion of the lesion be demonstrated preoperatively. In our case the venous (uncalciﬁed) aneurysm was larger than the arterial (calcified) component.

**Summary and Conclusions**

Arteriovenous fistula involving the portal vein or its tributaries is a rare condition and may be accompanied by ascites. In this paper the reports of 5 cases are reviewed and a new case is reported; in addition, reports of 2 cases of hepatoportal fistula are reviewed.

Ascites, which was reported in 2 of the 6 cases of splenic arteriovenous fistula, may be explained on the basis of portal hypertension or hepatic congestion or a combination of both these factors. It occurred only in patients with a single arteriovenous fistula of the splenic vessels.

In patients with ascites or signs of portal hypertension, arteriovenous fistula involving the portal system should be suspected, particularly if a machinery murmur can be heard or if there is x-ray evidence of calcification in the region of the portal vein or its tributaries. The diagnosis can be established by aortography. Surgical cure is possible in some cases.

**Summario in Interlingua**

Fistula arteriovenose afficien te le vena portal o su tributaris es un condition rar e poti esser accompaniata de ascites. Le presente articulo passa in revista le reportos de 5 casos e reporta un caso additional. In plus, reportos de 2 casos de fistula hepaticoportal es passate in revista.

Ascites, que esseva reportate in 2 del 6 casos de splenic fistula arteriovenose, poti esser explicate supe le basi de hypertension portal o de congestion hepatic o de un combination de iste factores. Illo occurreva solmente in patientes con un unice fistula arteriovenose del vasos splenic.

In patientes con ascites o signos de hypertension portal, fistula arteriovenose afficien te le systema portal debe esser suspicious, specialmente si un murmure de locomotiva es audi-bile o si il ha evidentia roentgenologico de calcification in le region del vena portal o su tributaris. Le diagnose poti esser estabite per aortographia. Curation chirurgic es possibile in certe cases.

**References**

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