**REVIEW**

**Visceral Venous Aneurysms: Clinical Presentation, Natural History and Their Management: A Systematic Review**

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Submitted 14 April 2009; accepted 26 May 2009

Available online 27 June 2009

**KEYWORDS**
Visceral vein; Portal vein; Splenic vein; Mesenteric vein; Renal vein; Aneurysm

**Abstract**  
**Aim:** Aneurysms of the visceral veins are considered rare clinical entities. The aim is to assess their clinical presentation, natural history and management.  
**Methods:** An electronic search of the pertinent English and French literature was undertaken. All studies reporting on aneurysms of visceral veins were considered. Cases describing patients with arterial–venous fistulae and extrahepatic or intra-hepatic portosystemic venous shunts were excluded.  
**Results:** Ninety-three reports were identified, including 176 patients with 198 visceral venous aneurysms. Patients’ age ranges from 0 to 87 years, and there is no apparent male/female preponderance. The commonest location of visceral venous aneurysms is the portal venous system (87 of 93 reports, 170 of 176 patients, 191 of 198 aneurysms). Aneurysms of the renal veins and inferior mesenteric vein are also described. Portal system venous aneurysms were present with abdominal pain in 44.7% of the patients, gastrointestinal bleeding in 7.3%, and are asymptomatic in 38.2%. Portal hypertension is reported in 30.8% and liver cirrhosis in 28.3%. Thrombosis occurred in 13.6% and rupture in 2.2% of the patients. Adjacent organ compression is reported in 2.2% (organs compressed: common bile duct, duodenum, inferior vena cava). The management ranged from watchful waiting to intervention. In 94% of the cases, aneurysm diameter remained stable and no complications occurred during follow-up. In most of the cases, indications for operation were symptoms and complications. Six cases of renal vein aneurysm are reported; three of them were asymptomatic. Three of these patients were treated surgically.  
**Conclusion:** The most frequent location of visceral venous aneurysms is the portal venous system. They are often associated with cirrhosis and portal hypertension. They may be
Primary venous aneurysms are not as common as arterial aneurysms. Venous aneurysms are described in the popliteal, jugular and saphenous veins, but rarely described in other veins. Although visceral venous aneurysms are rare lesions, they are increasingly described in recent years, probably because of the increasing availability of advanced radiological imaging in clinical practice.

Their prevalence, clinical presentation and complications have not been adequately reviewed. Most of the visceral aneurysms are in the form of case reports, and there are few published case series that specifically address indications for surgery and optimal surgical techniques. This study is conducted to systematically review the published data regarding visceral venous aneurysms aiming to assess their clinical presentation, natural history and management.

Methods

A systematic Medline search was undertaken to identify all reported cases of visceral venous aneurysms. The keywords used were ‘visceral vein’, ‘splanchnic vein’, ‘portal vein’, ‘intra-hepatic portal vein’, ‘extrahepatic portal vein’, ‘splenic vein’, ‘superior mesenteric vein’, ‘umbilical vein’, ‘inferior mesenteric vein’, ‘renal vein’ and ‘aneurysm’. The retrieved articles were also searched for any relevant references.

Only reports written in English and French and describing aneurysms of the visceral veins without arterial—venous fistulae, extrahepatic or intra-hepatic portosystemic venous shunts were included in the analysis.

Results

Ninety-three reports were identified, including 176 patients with 198 visceral venous aneurysms. Patients’ age ranges from 0 to 87 years, and there is no apparent male/female preponderance. The most frequent location of visceral venous aneurysms is the portal venous system (87 of 93 reports, 170 of 176 patients 191 of 198 aneurysms).1–87 Renal vein aneurysms (six reports, six patients and six aneurysms)88–92 and inferior mesenteric vein (one aneurysm)93 also are described.

Portal venous system

Anatomic location

In the portal venous system, the aneurysm is located in the main extrahepatic portal vein in 52 cases, splenic vein in 28 cases, superior mesenteric vein in 17 cases, splenic—superior mesenteric vein confluence in 37 cases, right portal vein in three cases, left portal vein in one case, intrahepatic portal vein in 34 cases and umbilical portion of the left portal vein in 19 cases (Fig. 1). Extrahepatic portal venous aneurysms range from 2.0 to 8.0 cm in diameter and the intrahepatic from 1.0 to 7.0 cm.

Clinical presentation

Data regarding clinical presentation are available for 123 patients. Abdominal pain is reported in 44.7% (55 of 123 patients) and gastrointestinal bleeding in 7.3% (nine of 123) of the patients. The aneurysm is asymptomatic and discovered incidentally during abdominal scanning, usually abdominal computed tomography (CT) scan and abdominal ultrasound, in 38.2% of the patients (47 of 123). Other symptoms such as fever, abdominal distension, nausea, loss of appetite, weight loss, vomit, malaise and jaundice are infrequently reported. Proposed aetiological factors include liver disease (mainly cirrhosis) and portal hypertension. Data regarding these possible aetiological factors are available for 162 patients. Portal hypertension is reported in 30.8% (50 of 162) and liver cirrhosis in 28.3% (46 of 162) of the patients.

Complication

Reported complications of the untreated visceral venous aneurysms are thrombosis, rupture and compression of adjacent structures. Complete thrombosis occurred in 24 patients (13.6%) and non-occlusive thrombus existed in six.
Rupture is described in four patients (2.2%); one of them during the postpartum period. Of the four ruptured, two are splenic vein aneurysms, one intrahepatic and one aneurysm of the right portal vein. The diameter of the ruptured aneurysms was 2 cm in three of the four cases. In two cases, the aneurysm compressed the common bile duct, the duodenum in two and the inferior vena cava in one.

Natural history and management
The management ranged from watchful waiting to intervention. There are available data for 87 patients. Fifty-three of them were followed up for a time period ranging from 1 to 72 months (mean: 21.15 months). Follow-up is most commonly performed using abdominal ultrasound. In 50 of them (94%), the diameter of the aneurysm remained stable and no complications occurred. In two patients, the aneurysm diameter increased and one underwent cavernous transformation. Thirty-four patients in total were operated (Table 1). In most of the cases, indication for operation was the occurrence of a complication (thrombosis and rupture) or presence of symptoms.

Operations performed include splenectomy (seven patients), aneurysmorrhaphy (eight), aneurysmectomy (three), aneurysmectomy and splenectomy (one), aneurysmectomy with splenectomy and shunt (one), aneurysmorrhaphy and splenectomy (one), distal pancreatectomy and splenectomy (two), aneurysmorrhaphy and portocaval shunt (one), splenectomy, distal pancreatectomy and liver transplantation (two) and transhepatic thrombectomy and thrombolysis (three) (Fig. 2).

Nine patients with portal venous aneurysm died during follow-up, resulting to 10.3% mortality. Three of them presented with complications; rupture or thrombosis. Five of these nine patients were operated (Table 1); two had been submitted to liver transplantation because of liver cirrhosis and two had a co-existing malignancy.

Renal vein
Six patients (five males) with renal vein aneurysm are reported. Patients’ age ranges from 33 to 73 years. In four cases, the aneurysm was located in the left renal vein. Aneurysm diameter ranged from 4 to 5.5 cm. Three cases presented with abdominal pain. Three patients were operated; aneurysm resection and reconstruction of the renal vein (two) and nephrectomy (one).

Inferior mesenteric vein
The only case of inferior mesenteric vein aneurysm is described in a 31-year-old woman, who also had a superior mesenteric vein aneurysm and presented with thrombosis. She presented aneurysmal dilatation of the inferior vena cava, the hemi-azygos vein, the right ovarian and the right iliac internal vein. She underwent arterial thrombolysis and transhepatic thrombus aspiration, which resulted in re-canalisation of the superior mesenteric vein aneurysm. Inferior mesenteric vein aneurysm remained occluded.

Discussion
The most frequent location of visceral venous aneurysms is the portal venous system and represents 3% of all venous aneurysms. Koc et al. reported a 0.43% prevalence of portal venous aneurysm among 4186 patients. As there are variations in the diameters of both normal and cirrhotic portal veins, an aneurysm of the portal venous system is considered to be present if the vessel diameter is larger than 20 mm, especially if the morphology is saccular or fusiform. Aneurysms are usually located at the main portal vein, the junction of the superior mesenteric vein and the splenic vein or at the hepatic hilus. Intrahepatic venous aneurysms are rare.

There are two main theories regarding the aetiology of portal vein aneurysms: congenital and acquired. During the embryonic development, three anastomoses form between right and left vitelline veins around the future duodenum. A complex process of involution and interconnection of these vitelline veins results in the portal vein. Abnormal development of the portal venous system during this critical period may give rise to an extrahepatic portal vein aneurysm. Incomplete regression of the distal right primitive vitelline vein may later form a portal vein aneurysm. Portal vein anomalies, including the right anterior segmental portal vein or the right anterior and posterior segmental portal veins originating from the umbilical portion of the portal vein, and a rightward deviation of the umbilical portion of the portal vein, are associated with aneurysms of the umbilical portion of the left portal vein. An inherent weakness of the vessel wall is another theory proposed to support a congenital origin. The congenital theory implies a developmental defect of the vein wall as the main cause of aneurysm development. Congenitally or developmentally defective segments may give rise to an aneurysm. The congenital theory is based on the presence of aneurysms in children and young adults without portal hypertension. Supporting evidence of a congenital theory lies in the in utero diagnosis of a portal vein aneurysm. The acquired lesions are secondary to chronic liver disease, mainly cirrhosis, portal hypertension, trauma and pancreatitis. Portal hypertension is reported in 30.8% and liver cirrhosis in 28.3% of the patients. Thrombophilia is recently suggested as an aetiologic factor of portal aneurysms. Recurrent thrombosis can cause portal vein occlusion that result in acute or chronic symptoms of portal hypertension and aneurysm formation. Congenital portal venous aneurysms are generally considered stable lesions and regular follow-up is usually sufficient. Acquired portal venous aneurysms, mainly when they are combined with liver cirrhosis and portal hypertension can have a more unpredictable evolution through time, requiring closer follow-up and intervention when complications occur.

The clinical importance of a portal vein aneurysm is related to its size. A small aneurysm usually does not show symptoms, while large ones are described as the cause...
<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Patients</th>
<th>Location of the aneurysm</th>
<th>Indication for surgery</th>
<th>Treatment</th>
<th>Follow-up (months)</th>
<th>Outcome</th>
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<tr>
<td>Barzilai and Kleckner⁴¹</td>
<td>1956</td>
<td>1</td>
<td>Right PV</td>
<td>GI bleed</td>
<td>Splenectomy</td>
<td>11</td>
<td>Death</td>
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<tr>
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<td>1960</td>
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<td>EPV</td>
<td>GI bleed</td>
<td>Splenectomy</td>
<td>1</td>
<td>Death</td>
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<td>Sedgwick³</td>
<td>1960</td>
<td>1</td>
<td>SV-SMV C</td>
<td>Compression of CBD</td>
<td>Cholecystojejunostomy, spleno-renal shunt</td>
<td>10</td>
<td>Good</td>
</tr>
<tr>
<td>Hermann and Shafer⁴</td>
<td>1965</td>
<td>1</td>
<td>SV-SMV C</td>
<td>Portal hypertension</td>
<td>Portocaval shunt</td>
<td>3</td>
<td>Good</td>
</tr>
<tr>
<td>Thomas⁵</td>
<td>1967</td>
<td>1</td>
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<td>GI bleed</td>
<td>Portocaval shunt</td>
<td>48</td>
<td>Good</td>
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<tr>
<td>Liebowitz and Rousselot⁶</td>
<td>1967</td>
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<td>Agnogenic myeloid metaplasia</td>
<td>Splenectomy</td>
<td>5</td>
<td>Death</td>
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<td>1987</td>
<td>1</td>
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<td>Thrombosis</td>
<td>Aneurysmectomy</td>
<td>nr</td>
<td>nr</td>
</tr>
<tr>
<td>Matthias et al.²⁰</td>
<td>1987</td>
<td>1</td>
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<td>Symptom</td>
<td>Aneurysmorrhaphy</td>
<td>nr</td>
<td>nr</td>
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<tr>
<td>Andoh et al.²¹</td>
<td>1988</td>
<td>1</td>
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<td>Symptom</td>
<td>Aneurysmorrhaphy, splenectomy</td>
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<td>Baker and Nepute²⁴</td>
<td>1990</td>
<td>1</td>
<td>SV-SMV C</td>
<td>Acute thrombosis, no collateral vessels present</td>
<td>Aneurysmorrhaphy, splenectomy, shunt</td>
<td>0</td>
<td>Good</td>
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<td>Glazer et al.³¹</td>
<td>1992</td>
<td>1</td>
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<td>Thrombectomy, aneurysmorrhaphy</td>
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<td>1996</td>
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<td>1997</td>
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<td>1</td>
<td>SV</td>
<td>Symptoms</td>
<td>Aneurysmectomy, splenectomy</td>
<td>nr</td>
<td>ni</td>
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<td>2002</td>
<td>1</td>
<td>EPV</td>
<td>Acute thrombosis, collateral vessels present</td>
<td>Thrombectomy, aneurysmorrhaphy</td>
<td>6</td>
<td>Good</td>
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<tr>
<td>Mucenic⁵⁷</td>
<td>2002</td>
<td>1</td>
<td>IPV</td>
<td>Portal hypertension</td>
<td>Splenectomy</td>
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<td>Good</td>
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<tr>
<td>Flis et al.⁵⁸</td>
<td>2003</td>
<td>1</td>
<td>EPV</td>
<td>Symptom</td>
<td>Aneurysmorrhaphy</td>
<td>6</td>
<td>Good</td>
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<td>Shimoda et al.⁵⁹</td>
<td>2003</td>
<td>1</td>
<td>SV</td>
<td>Rupture</td>
<td>Distal pancreatectomy, splenectomy</td>
<td>nr</td>
<td>Good</td>
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<td>SMV, IMV</td>
<td>Thrombosis</td>
<td>Thrombolysis Transhepatic thrombectomy</td>
<td>nr</td>
<td>nr</td>
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<td>Jin et al.⁶⁹</td>
<td>2005</td>
<td>2</td>
<td>EPV, SV-SMV C</td>
<td>Symptom, Prophylactic surgery</td>
<td>Aneurysmorrhaphy, splenectomy</td>
<td>6,6</td>
<td>Good, good</td>
</tr>
<tr>
<td>Luo et al.⁷⁶</td>
<td>2006</td>
<td>1</td>
<td>EPV</td>
<td>Symptom</td>
<td>Splenectomy, spleno-renal shunt</td>
<td>6</td>
<td>Good</td>
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<tr>
<td>Wolff et al.⁷⁷</td>
<td>2006</td>
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<td>EPV</td>
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<td>Thrombectomy, aneurysmorrhaphy, portocaval shunt</td>
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<td>Shah and Robbani⁷⁹</td>
<td>2006</td>
<td>1</td>
<td>SV</td>
<td>No</td>
<td>Splenectomy, distal pancreatectomy, lienorenal shunt</td>
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<td>nr</td>
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<td>Cho et al.⁸⁰</td>
<td>2007</td>
<td>4</td>
<td>EPV, SV-SMV C (2), left portal vein</td>
<td>Abdominal pain, peri- pancreatic mass, Mass effect on the duodenum, Gallstone pancreatitis, Liver cirrhosis</td>
<td>Aneurysmorrhaphy (1), Aneurysmorrhaphy, cholecystectomy (1), cholecystectomy Aneurysmectomy, iliac venous interposition allograft, Roux-en-Y hepaticojejunostomy (1), liver transplantation (1)</td>
<td>2–73</td>
<td>Good (3), Death (1)</td>
</tr>
</tbody>
</table>

(continued on next page)
of duodenal compression, inferior vena cava compression, biliary tract obstruction and portal vein thrombosis. Rupture of a portal venous aneurysm is reported in four patients, one of whom died. A case of splenic vein aneurysm rupture is described in a young woman a few hours after delivery.

Portal venous system aneurysms require no treatment in most cases. Serial follow-up of the patients with abdominal ultrasound is usually sufficient. In 88% of the patients who were followed up, aneurysm diameter remained stable and no complications occurred. Portal vein thrombosis may necessitate anticoagulation therapy or percutaneous intervention with thrombectomy or thrombolysis.

Symptomatic aneurysms and presence of complications, such as thrombosis, rupture and adjacent structure compression, are generally considered indications for operation. Thirty-four patients with portal vein aneurysm were treated surgically or interventionally, and some of them had more than one procedures performed (Table 1, Fig. 2). The type of the procedure is based on the location and size of the aneurysm, the presence of complications and the co-morbidities (portal hypertension and liver cirrhosis). Aneurysmorrhaphy is the easiest procedure to excise the aneurysm, mainly when it is saccular, and restore normal luminal diameter of the portal vein. In cases of fusiform aneurysms, if an aneurysmectomy is performed, the conduit used to replace the portal vein can be an allograft from cadaveric donor, or a synthetic graft.

The location of the aneurysm is significant for the choice of the procedure. The location is determined in 30 of the 34 operated patients: in 29 it was located in the extrahepatic portal system. In four of them, the aneurysm was located in the splenic vein. All patients underwent splenectomy, combined with distal pancreatectomy in three cases. Two of the four patients with superior mesenteric vein aneurysm underwent aneurysmolysis. Patients operated with aneurysm of the extrahepatic main portal vein or superior mesenteric–splenic vein confluence underwent various procedures, most frequently aneurysmolysis. A single patient with intrahepatic portal aneurysm was operated: he had documented portal hypertension and underwent splenectomy (Fig. 2).

Several patients with documented portal hypertension underwent surgical shunt procedures, alone or combined with various other procedures. These shunt procedures are performed to decompress portal hypertension and do not specifically treat the venous aneurysm. Patients with liver cirrhosis present increased perioperative risk, and two out of the five operated died during follow-up. Because of the low reported rate of rupture and the risk of surgery in the presence of portal hypertension and liver cirrhosis, there is no strong evidence that prophylactic resection of the portal vein aneurysm is beneficial in these patients.

Renal vein aneurysms are very rare, with only six cases reported in the English literature. Five were male and three cases presented with abdominal pain. The remaining three cases were discovered incidentally or...
during laparotomy. The aneurysm was located on the left renal vein in four cases. Left renal vein is considered to be involved more often in aneurysm formation because of its more complicated embryologic development. Renal vein aneurysms must be differentiated from distended left renal vein that is recognised as a normal variant. The nutcracker phenomenon is attributed to compression of the left renal vein as it courses between the superior mesenteric artery anteriorly and the aorta posteriorly. Renal vein aneurysms should also be differentiated from idiopathic renal vein varices, especially solitary ones. Renal vein varices are usually smaller than aneurysms and typically accompanied by a dilated venous network adjacent to the renal pelvis and upper ureter. Because of the small number of renal vein aneurysms reported, there are insufficient data regarding optimal treatment. Three of the six patients were treated surgically, two underwent renal vein reconstruction and one nephrectomy.

Conclusion

Visceral vein aneurysms may not be as uncommon as previously thought, and their most frequent location is the portal system. They are often associated with cirrhosis and portal hypertension, and their presentation includes abdominal pain and other non-specific symptoms, or discovered incidentally. Watchful waiting is an appropriate treatment, except when complications occur. Most common complications include thrombosis and rupture.

Conflict of interest/funding

None.

References


Visceral Venous Aneurysms: A Systematic Review