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Case report

Fatal bleeding from duodenal varices as a late complication of neonatal thrombosis of the inferior vena cava

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Summary. A 21-year-old man was hospitalized with repeated bleeding from the gastrointestinal tract, the cause of which could not be detected by gastroscopy, colonoscopy or arteriography. After the third surgical intervention he died of uncontrollable bleeding. At autopsy, marked and partly ulcerated duodenal varices were found to be the cause of the bleeding. They were part of a collateral circulation that had developed from chronic obstruction of the inferior vena cava, where thrombosis had arisen in association with neonatal renal vein thrombosis. There was consecutive renal infarction which had required a left nephrectomy on the 2nd day after birth.

Key words: Thrombosis of inferior vena cava – Collateral circulation – Duodenal varices – Neonatal renal vein thrombosis – Autopsy

Introduction

Duodenal varices are exceptionally rare. Since the first report by Alberti (1931) they have been described in liver cirrhosis (Swart 1968; Amin et al. 1985; Khouqueer et al. 1987) portal vein thrombosis (Itzchak and Glickman 1977; Rösch 1981) and mesenteric thrombosis (Rösch 1981; Ullrich et al. 1981; Reis et al. 1982). In a few cases other diseases such as a congenital vascular anomaly (Gushurst and Lesesne 1984) and arteriovenous aneurysms (Wheeler and Warren 1957) may have caused the development of varices. We present here an unusual case of a young patient who died of diffusely bleeding duodenal varices, which had formed following a perinatal thrombosis of renal vein and inferior vena cava.

Case report

After episodes of intermittent stomach trouble for 2 years a 21-year-old student was admitted to hospital 2 months before his death because of upper abdominal pain, nausea and hourly diarrhoea of dark faeces. Emergency colonoscopy and subsequent endoscopy of the upper digestive tract showed no source of bleeding. An exploratory laparotomy with gastrotomy and duodenotomy was performed and an assumed source in the gastric wall was sutured. On the 4th day of hospitalization the patient was transferred to the surgical department of the University of Bonn because of continuous uncontrollable upper gastrointestinal bleeding. Despite emergency coeliac and mesenteric arteriography, the source of bleeding could not be found. A significant finding was marked dilatation of the inferior mesenteric vein whereas the hepatic portal vein appeared to be unaltered.

Repeat laparotomy with partial gastrectomy and excision of the enlarged spleen was performed. Intraoperatively, congested veins in the portal system and bleeding from congested short gastric veins was observed.

Seventeen days after the gastrectomy, the patient suffered shock due to a suture dehiscence and further continuous bleeding. At a third laparotomy the residual stomach was resected and an oesophagojejunostomy was performed. A suture dehiscence of the duodenal stump with resultant bile peritonitis occurred post-operatively. The patient developed renal failure and shock lung which proved fatal.

In the perinatal period the patient had shown swelling of the lower extremities. Two days post-partum he had undergone a left nephrectomy because of haemorrhagic renal infarction resulting from a fulminant renal vein thrombosis. Because of acute vertebral pain 1 year prior to his death computed tomography had been performed which had shown a prominence of the azygos and hemiazygos veins.

Pathological findings

Histologically, the samples of the gastric fundus showed multiple areas of complete haemorrhagic necrosis of the mucosa and extensive haemorrhage in the submucosa. Slightly dilated veins were seen in the submucosa while oesophageal varices could not be found. The spleen (weighing 270 g) demonstrated the characteristic changes typical of portal hypertension. The specimen of the residual stomach revealed large ischaemic transmural necrosis and suppurative peritonitis. No evidence of varicose veins was found.

At autopsy, no varices were found in the oesophagus.

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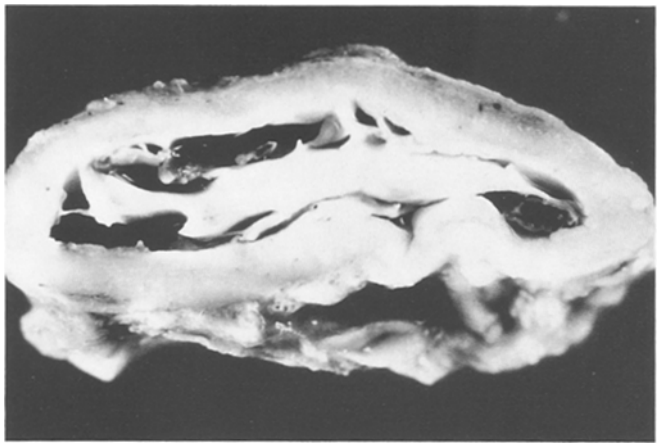

Fig. 1. Cross-section of the cordlike transformed vena cava inferior. Sieve-like openings visualized on cross-sections were just patent for stylets and partly communicated to the right renal vein. ×4.5

The proximal suture of the duodenal loop was dehiscent at a distance of 2 cm leading to a bile peritonitis. In the duodenal mucosa there were several withish, slightly prominent areas of about the size of a rice grain. Structural alterations were not seen an the surface or on gross sections in the enlarged liver (4200 g).

The inferior vena cava was transformed into a fibrous cord measuring about 2 × 0.7 cm between the renal vein openings and the common iliac veins. Sieve-like openings were visualized on cross-sections (Fig. 1) and were just patent to stylets and partly communicated with the right renal vein. The right kidney, weighed 250 g and showed a solitary cyst and a slightly scarred surface.

Histologically, numerous grossly ectatic submucous veins of the duodenum were found to communicate with similarly dilated subserosal vessels (Fig. 2) which occasionally showed marked intimal fibrosis. At several places the varices were closely adjacent to the mucosal ulcer and only partially separated from the duodenal lumen by blood clots. The cord-like vena cava showed multiple re-canalized luminal surrounded by mature connective tissue containing elastic fibres. Similar alterations could be followed up to the right renal vein, from which some collateral vessels branched off to the retroperitoneal soft tissue. No significant histological alterations were seen in the liver.

Discussion

Renal vein thrombosis often occurs at an early age in childhood and may be bilateral. At an advanced stage it is frequently combined with thrombosis of the inferior vena cava (Feriozi et al. 1951; Fielding et al. 1986; Pollak and Weiss 1989). In the present case, the clinical and pathological findings suggest that the left-sided renal vein thrombosis occurred simultaneously or immediately previous to the extended occlusion of the inferior vena cava. A complete infarction of the right kidney had obviously been prevented by the retroperitoneal col-

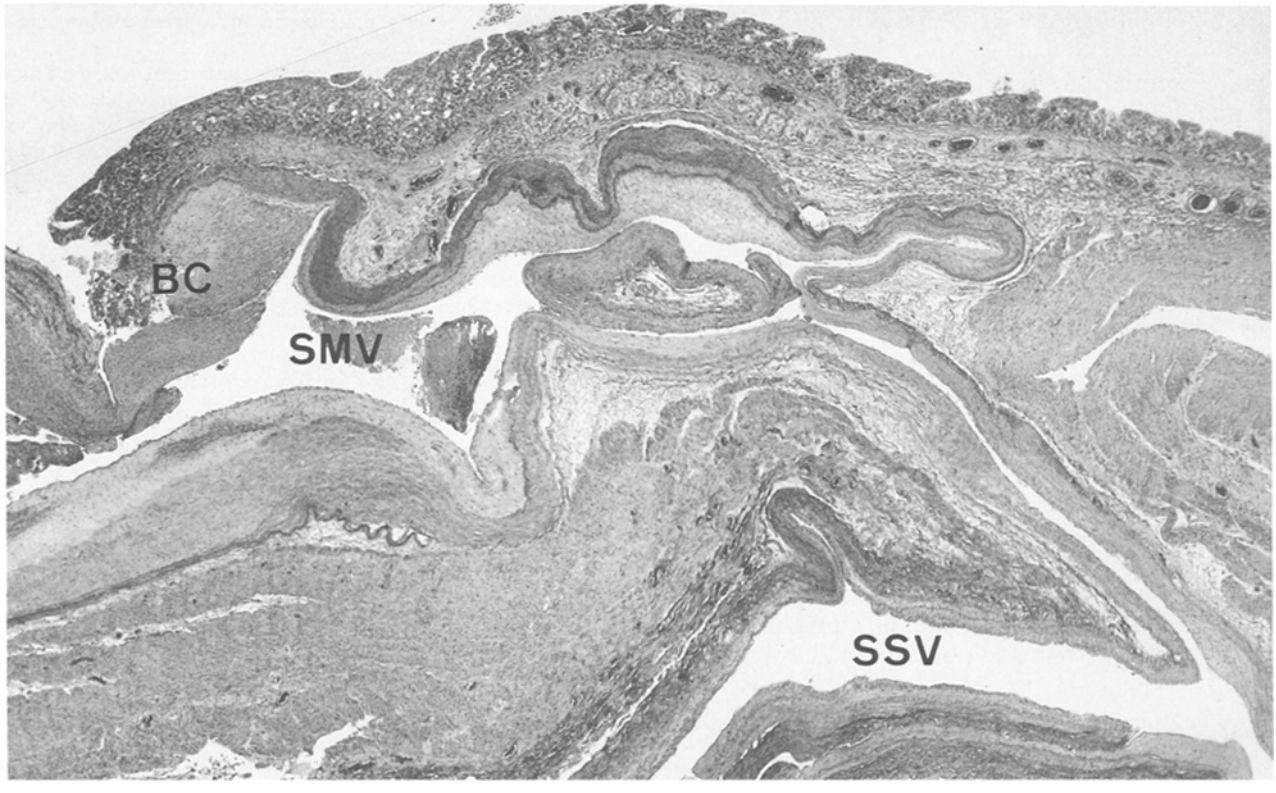

Fig. 2. Histologically, an extremely ectatic submucous vein (*SMV*) is shown to communicate with a similarly dilated subserous vessel (*SSV*). The submucous vein shows a marked intimal fibrosis. At the left edge the rupture of this vein can be seen, covered by a blood clot (*BC*). Elastica van Gieson, ×16

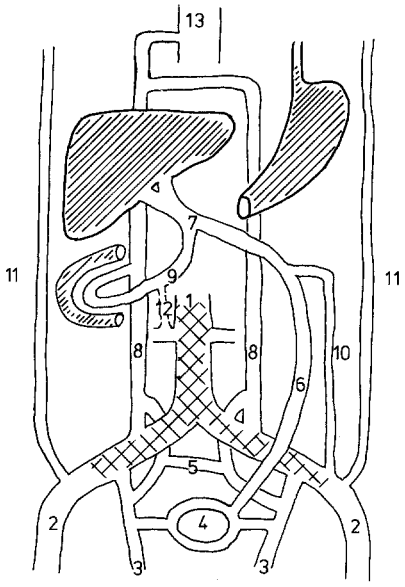

Fig. 3. Scheme of the vessels which can contribute to collateral circulation in inferior vena caval obstruction. 1, inferior vena cava; 2, external iliac veins; 3, internal iliac veins; 4, superior rectal plexus; 5, presacral plexus/lumbar plexus; 6, inferior mesenteric vein; 7, portal vein; 8, lumbal veins/azygos and hemi-azygos veins; 9, retroduodenal collateral veins; 10, umbilical veins/inferior epigastric veins; 11, subcutaneous veins of the thoraco-abdominal wall; 12, collateral veins from renal vein to retroperitoneum; 13, superior vena cava

laterals coming from the right renal vein which had allowed partial drainage of blood.

The venous return from the lower part of the body in inferior vena caval occlusion is generally possible through the connections between the superior and inferior vena cavae and the rarely encountered development of a cavo-portal collateral circulation.

Cavo-caval anastomoses develop predominantly via the lumbar veins which empty into the azygos and hemiazygos veins above the diaphragm (Ferris et al. 1967; Fletcher and Thomas 1968; Hach 1971; Sörensen and Taenzer 1973; Vogel et al. 1991). In the event of involvement of the iliac veins the lumbal veins are supplied by the paravertebral and presacral plexus (Fletcher and Thomas 1968; Preuß and Hänsgen 1981). Further collaterals may involve the subcutaneous veins of the thoraco-abdominal wall (Ferris et al. 1967; Fletcher and Thomas 1968; Hach 1971; see Fig. 3). With thrombosis of the inferior vena caval trunk below the renal vein, the gonadal and periureteric veins may also function as collaterals (Hach 1971; Friedman et al. 1978; Chatel et al. 1979) leading to destruction of the vertebral column (Singson et al. 1984) or to ureteric compression (Friedman et al. 1978) in a few cases.

The rarer cavo-portal collateral circulation may imitate the porto-caval anastomoses typical of portal hypertension in liver cirrhosis (Fig. 3). Thus the blood of the pelvis and the lower extremities can be shunted to the portal vein via the superior rectal plexus and the inferior mesenteric vein (Kendall 1965; Ferris et al. 1967; Fletcher and Thomas 1968; Sörensen and Taenzer 1973; Zückmantel and Endert 1988). Even the inferior epigastric vein in the anterior abdominal wall may occasionally drain into the portal vein via paraumbilical and umbilical veins (Ferris et al. 1967; Mignon and Flora 1974; Zückmantel and Endert 1988).

Only one case in the literature reports a radiologically documented development of duodenal varices caused by an inferior vena caval occlusion. In a 54-year-old patient Greenspan and Bryk (1973) demonstrated duodenal varices angiographically. These had developed after chronic occlusion of the inferior vena cava within the retroduodenal space and drained blood from the ascending lumbar veins into the portal vein.

In the present case, there had probably been drainage of blood from the lower part of the body into the portal vein via the haemorrhoidal plexus and the markedly dilated inferior mesenteric vein. The dilation of the lumbar veins identified by computed tomography, however, points to collateral drainage via the azygos-hemiazygos system. As in the case of Greenspan and Bryk (1973) venous connections between the lumbar veins and the portal vein may have formed in the retroduodenal space and caused the development of the duodenal varices. The retroperitoneal venous vessels branching off from the renal vein suggest that renal blood may have contributed to the development of duodenal varices (see Fig. 3).

The fatal course of the disease in this young patient clearly demonstrates that in the case of undetermined upper gastrointestinal bleeding not originating in the oesophagus or stomach, duodenal varices must be considered. In this connection not only portal hypertension but also the vena cava occlusion syndrome has to be taken into account.

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