Spontaneous liver bleeding in a patient with congenital arterioportal fistulisation

Presentation of a casus princeps and review of the literature

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Abstract

We present the first case reported in the literature describing spontaneous liver haemorrhage due to diffuse arterioportal fistulae. A 48-year old Caucasian woman was admitted to the hospital complaining of acute epigastric pain eradiating to the right shoulder. Patient never had any penetrating or blunt abdominal trauma in the past nor any intervention on the liver. CT scan of the abdomen revealed a subcapsular haematoma originating from two bleeding sites in the right liver lobe. Arteriography of the common hepatic artery showed opacification of the portal branches, indicative of an arterioportal fistula. A hypertrophic feeding branch of the right hepatic artery was then embolized, resulting in disappearance of the fistula. After complete resolution of the haematoma, investigations to detect underlying liver lesions were repeatedly negative. Therefore we conclude that a diffuse congenital arterioportal fistula was the cause of spontaneous bleeding. This is to our knowledge the first case in whom a spontaneous liver bleeding secondary to diffuse arterioportal fistulisation is reported. A review of the literature regarding arterioportal fistulas and regarding the possible aetiology of spontaneous liver haematomas is provided. (Acta gastroenterol. belg., **2013**, 76, **62-65**).

Key words : liver haemorraghe, arterioportal fistulisation, etiology, congenital arterioportal fistula, liver lesion.

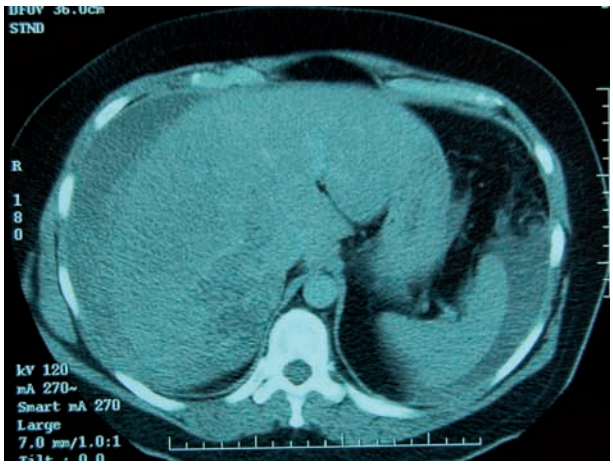

Fig. 1. — CT scan of the upper abdomen on day 1 of admission, showing subcapsular haematoma originating from two bleeding lesions in the right liver lobe.

List of abbreviations (in alphabetical order)

APF : arterioportal fistula
BRHA : branch of the right hepatic artery
HA : hepatic artery
LHA : left hepatic artery
RHA : right hepatic artery

Case report

A 48-year old Caucasian woman was admitted to the hospital complaining of acute epigastric pain and pain in the right shoulder. Patient was known to have diabetes mellitus type 2, arterial hypertension, mild psoriasis and astma. Her current treatment consisted of metformin, indapamide, simvastatin, atenolol, enalapril, amitriptylin and oral contraceptives. Acetylsalicylic acid was started two weeks before admission after coronarography. Patient never had any penetrating or blunt abdominal trauma in the past ; liver biopsy or any other intervention on the liver were never performed. Physical examination revealed no significant abnormalities. Laboratory results showed a normal haemoglobin of 13.5 g/dL, a normal C-reactive protein and normal troponins. Liver enzymes were slightly elevated (aspartate aminotransferase 65 U/L, alanine aminotransferase 72 U/L). CT scan of the abdomen was performed, showing two intrahepatic haematomas extending to the subcapsular space, originating from two bleeding sites in the right liver lobe (segment VII and VIII) (Fig. 1). Patient was admitted to the intensive care for haemodynamic monitoring. Haemoglobin decreased from 13.5 g/dL to 7.6 g/dL in three days time. Magnetic resonance imaging (MRI) of the liver four days after admission showed a heterogeneity of the right liver, the exact nature of which could not be identified. No solid liver tumour could be demonstrated (Fig. 2).

Patient remained haemodynamically stable and on day 7 of admission an arteriography was performed. There was a normal hepatic arterial vasculature in which no anatomic variation could be observed. Contrast injection into the common hepatic artery showed a slightly hypertrophic branch of the right hepatic artery (BRHA), irrigating the cranial part of the right lobe. No active

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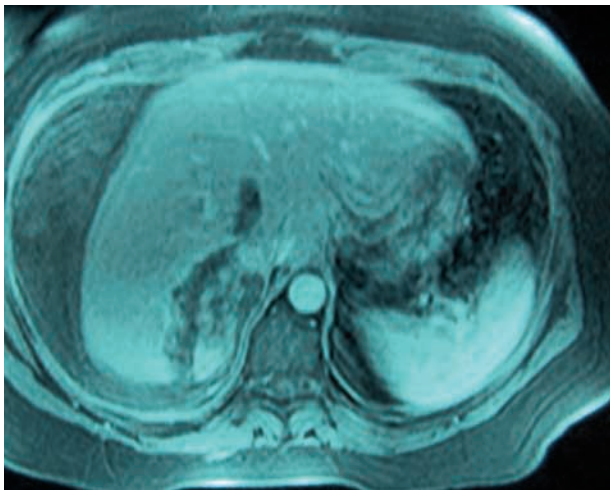

Fig. 2. — T2 weighted MRI of the upper abdomen on day 4 of admission, showing subcapsular haematoma and a lesion posteromedial in the right liver, expanding to anterior. Due to the blood present in the parenchyma the nature of the lesion cannot be identified.

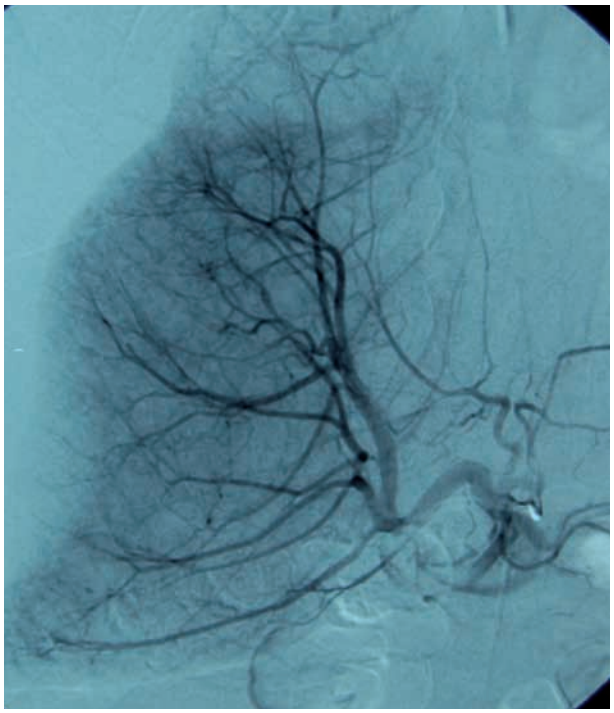

Fig. 3. — Arteriography of the liver on day 7 of admission. Image after injection of contrast in the arteria hepatica communis, showing a hypertrophic branch of the arteria hepatica dextra but no signs of acute bleeding.

bleeding was seen (Fig. 3). Just a few seconds after contrast injection the portal veins were already filling with contrast whereas in the remaining part of the liver no early contrast enhancement of portal vessels was observed (Fig. 4). The BRHA was subsequently embolized with contourparticles of 350 to 500 mm, resulting in disappearance of the fistulisation. Patient recovered well

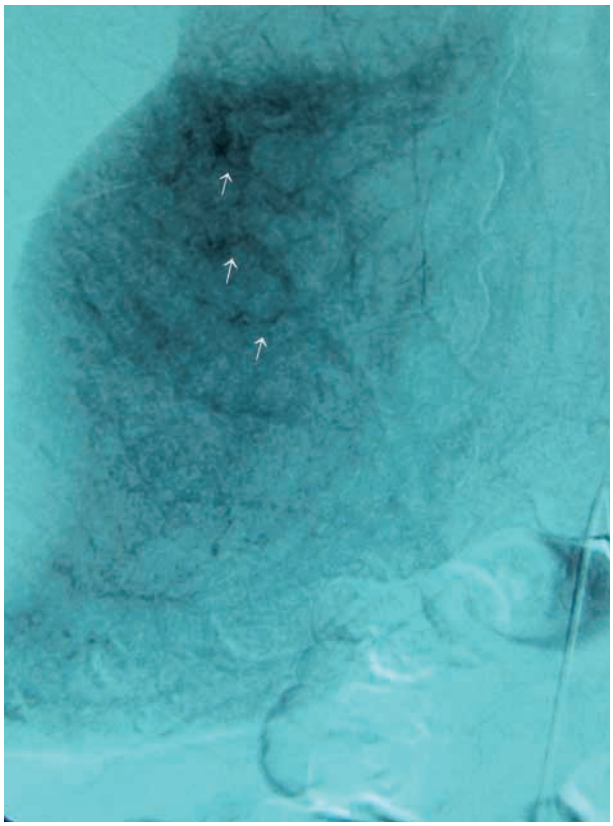

Fig. 4. — Arteriography, image 9 seconds after figure 3, showing diffuse opacification of the portal veins (which can be recognised by the horizontal lines as indicated).

and was discharged on day 21 of admission. Liver biopsy showed dilated sinusoids with focal accumulation of blood. However, there were no blood filled lakes suggestive of peliosis. Throughout the following 4 months the liquefied haematoma was repeatedly drained percutaneously up to complete resolution. After resolution, MRI of the liver 15 months later showed no underlying lesion, but the parenchyma appeared slightly altered with displacement of hepatic veins. Up to 4 years later, the patient remained without complaints.

Discussion

Spontaneous liver haemorrhage is a known complication of several disorders. The largest group consists of primary liver malignancies such as hepatocellular carcinoma (1,2). Other causes are benign lesions such as adenomas (oral contraceptive use is known to cause liver adenomata and can secondary cause bleeding (6)), haemangiomas (1), metastatic lesions (1) and peliosis hepatis (3). Cirrhosis can also cause spontaneous bleeding, either because of rupture of macronodular cirrhosis, or because of rupture of hepatic varicose veins and lymphatic vessels (1). Ehler Danlos type IV has also been described as a possible cause (4). Spontaneous liver haemorraghe can also occur in pregnant women with HELLP syndrome (5).

Table 1. — **Classification of arterioportal fistulas (APF) as proposed by Guzman *et al.* (7)**

	Definition	Clinical Findings	Natural history
Type 1	Small peripheral asymptomatic APF with minimal physiologic insult	Asymptomatic	Thrombose spontaneously within 1 month
Type 2	Larger central fistulas causing physiological insult	Portal hypertension	Portal hypertension
		History of penetrating abdominal trauma	Hepatoportal sclerosis
		Abdominal bruit	Possibly portal fibrosis
Type 3	Diffuse, intrahepatic congenital APF	Failure to thrive (in neonates) Portal hypertension Diarrhea	Possibly severe portal hypertension in infancy

Our patient did not have any of the above conditions. She had been taking oral contraceptives for decades but no liver bleeding secondary to oral contraceptives has been reported in the absence of adenoma (6). As mentioned earlier, no adenoma was found in our patient. Liver biopsy showed sinusoidal dilation. This dilation can be expected in patients with toxic liver damage, granulomatous liver disease, liver tumour or when outflow of the hepatic veins is impaired as in Budd-Chiari syndrome or cardiac failure (7). Our patient did not have any of the latter conditions. Oral contraceptives can cause sinusoidal dilation by a yet unknown mechanism (7). We assume that this is the reason of the sinusoidal dilation in our patient.

Arterioportal fistulas (APFs) are arteriovenous communications between splanchnic arteries and the portal vein. They can originate from any splanchnic artery, although the most common communications are hepatoportal (65%), splenoportal (11%) and between the superior mesenteric artery and the portal vein (10%) (8). Symptoms range from none to signs of severe portal hypertension with ascites and gastro-intestinal variceal bleeding. Congestive heart failure and diarrhea can also be observed (8). Whether a patient becomes symptomatic or not, depends mainly on the size and localisation of the fistula and the concomitant liver disease (9). An abdominal bruit in the right hypochondric area is an important clinical sign and can be heard in approximately 33% of patients. When fistulas are greater than 4 mm in diameter it is likely that this abdominal bruit can be heard (8,10). On ultrasound small diffuse APFs are generally not visualized, though a longer existing solitary fistula can be recognised as a tubular structure with arterial Doppler signal, corresponding to a dilated portal venous branch.

Recently a classification of APFs was proposed by Guzman *et al*. in which three classes are discerned. Type 1 consists of small peripheral asymptomatic APF. Type 2 consists of larger central fistula causing physiologic insult and signs of portal hypertension. Treatment consists of embolization if technically possible. If this is unfeasible or unsuccessful, then surgical approaches are warranted. Type 3 consists of diffuse fistulas. Treatment may consist of hepatic artery ligation, embolization, resection, or liver transplantation (8) (Table 1).

A classification of congenital intrahepatic APF was proposed by Norton e.a.. He classified congenital APFs according to the angiographic findings. A unilateral intrahepatic APF (type 1) is supplied by only the right, left or main hepatic artery. Bilateral lesions (type 2) include supply from branches of both the left and right hepatic artery. Finally, type 3 lesions are complex lesions, typically consisting of a plexiform vascular nidus with multiple feeding arteries, including supply from arteries other than the hepatic arteries (e.g. the left gastric artery) (11) (Table 2). Our patient would be classified as a APF type 3 according to Guzman *et al* and type 1 according to Norton *et al*.

As suggested by the classification, APFs can either be acquired or congenital. Acquired APFs can be seen after blunt or penetrating abdominal traumas, but are also seen after interventional hepatic procedures such as liver biopsy, percutaneous transhepatic cholangiography, transhepatic catheterization of bile ducts, liver surgery, radiofrequency ablation (8). Non iatrogenic acquired APFs have been described in association with entities such as haemangiomas, cirrhosis, regenerating liver nodules, hepatic abscess, Budd-Chiari syndrome, Rendu-Osler-Weber syndrome, type IV Ehlers-Danlos syndrome, aneurysms and Allagille syndrome (8). Our patient had none of the above mentioned conditions or history. The possibility of Rendu-Osler-Weber disease (hereditary haemorrhagic teleangiectasia, HHT) was even so considered. There was, however, no epistaxis. No mucocutaneous teleangiectasia nor any other visceral arteriovenous malformation could be documented and there was no family history of HHT (15). Liver involvement in HHT consists of multiple or diffuse teleangiectatic lesions (16) and is frequently associated with the presence of follicular nodular hyperplasia or other structural liver abnormalities (15). None of these features were present in our patient, who had a solitary fistula, rendering the diagnosis of HHT highly unlikely. Congenital APFs are usually diffuse. Diffuse intra-hepatic A-V fistulas are always congenital in origin whereas a solitary fistula is usually acquired (12). Congenital APFs are usually seen in

Table 2. — **Classification of congenital intrahepatic arterioportal fistulas as proposed by Norton-Jacobson *et al.* (11)**

	Afferent supply	Description of afferent vessel(s)
Type 1	Unilateral	RHA or LHA or main HA only
Type 2	Bilateral	Both RHA and LHA or equivalent supply from main HA
Type 3	Complex	RHA and/or LHA *and* nonhepatic artery supply

(RHA : right hepatic artery. LHA : left hepatic artery. HA : hepatic artery).

children ; the eldest patient reported was a 13-year old boy (13). Except for one solitary congenital APF in a 73-year old woman (14) no APFs have been described in patients older than 13 years. Our patient was 48 years old at presentation and is therefore the oldest case ever reported with diffuse APF. Moreover she is the first patient with congenital APF presenting with spontaneous bleeding.

Conclusion

Congenital arterioportal fistula is a condition consisting of arteriovenous communications between splanchnic arteries and the portal vein. Symptoms range from none to severe portal hypertension with ascites and gastro-intestinal variceal bleeding. This patient is the oldest patient reported with congenital diffuse APF. Moreover, to date there is no literature reporting spontaneous liver bleeding as a consequence of APF. The present case is therefore the first report of spontaneous liver bleeding as an initial presentation of APF. The possibility of an underlying arterioportal fistula should therefore be considered when a patient presents with signs of portal hypertension or liver haemorraghe e causa ignota.

Conflict of interest

All authors signed the copyright agreement. All authors have no conflict of interest to declare.

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