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Persistent ductus venosus without portal hypertension in a young alcoholic man


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SUMMARY The discovery of a large patent ductus venosus resulted from radiological investigations in a 34-year-old man, a chronic alcoholic of low mental status. Splenoportal and inferior caval venograms were performed because of recent exacerbation of the neurological symptoms and electroencephalographic criteria of portacaval encephalopathy. Portal pressure was 8 mm Hg. A liver scan, a laparoscopy, and a liver biopsy were performed. They showed that the gland was atrophic with a microscopic appearance of alcoholic fibrosis, but without any nodular regeneration. The relationship between the fistula, the mental state, and the atrophic liver is discussed. Such a malformation appears to be very uncommon.

Radiological investigations in the diagnosis of liver dysfunction, especially cirrhosis, lead commonly to the discovery of portacaval shunts. Most of these connexions are explained by portal hypertension itself. Yet, in some cases, the anastomotic veins have an unusual topography, either splenorenal or umbilical, and it is reasonable to assume the preexistence of a spontaneous portacaval derivation.

We recently observed a case of a large portacaval fistula with the radiological configuration of a persistent ductus venosus.

Case Report

A 34-year-old man, a construction worker of Italian origin, was admitted to the Montpellier University Hospital in March 1970 because of psychiatric symptoms of a few weeks' duration. He was a known alcoholic and these symptoms were at first related to his drinking. However, initial examination revealed a flapping tremor. The patient exhibited alternately indifference and agitation, and he became somnolent for a few days; normal consciousness reappeared rapidly and objective examination confirmed mental subnormality which probably preceded the recent psychiatric disturbance. Physical examination revealed no ascites or collateral circulation and the liver and spleen were not enlarged; there was neither jaundice nor oedema and no evidence of abdominal bruit or thrill could be elicited.

Biological results showed: a low bromsulphalein clearance (6%); prothrombin (53%); bilirubin 0.8 mg per 100 ml. The serum protein level was 6 g per 100 ml (serum albumin 48% and gamma globulins 29%); SGOT and SGPT were normal. The venous blood ammonia level was 63 µg per 100 ml. Blood urea nitrogen and blood sugar levels were normal, but an oral glucose tolerance test resulted in an excessive rise in blood sugar, followed by a prolonged and exaggerated plasma insulin response.

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Fig. 1 Reproduction of the liver scan.
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Fig. 2 Splenoportal venogram. This figure shows clearly the division of the portal vein: the right branch is atrophic, the left branch is greatly enlarged and probably has a high blood flow, as indicated by the dilution of the contrast substance. It continues into a large tortuous vessel which seems to be directed towards the spinal column. There are no visible derivative collaterals. The topography of the shunt provides consistent evidence of a persistent ductus venosus.

Fig. 3 Inferior caval venogram. After catheterization of the other extremity of the portacaval shunt, dye was injected against the flow. Partial injection suggests the existence of a high flow through the fistula, and confirms the features seen on the splenoportal venogram.

Fig. 4 Schematic representation of the radiographs. (1) Inferior vena cava; (2) left and (3) right branches of the portal vein; (4) opening of the left branch into the inferior vena cava; this appearance was seen on the caval venogram and also on the splenoportogram. (5) Representation of the right hepatic vein which did not communicate with the fistula.

Fig. 5 Inferior caval venogram. Direct injection of the right hepatic vein, which appeared to be normal in its shape and position.

Radiological investigation showed no oesophageal varices. A liver scan (Fig. 1) showed atrophy of the gland. A laparoscopy was performed which showed that the liver was atrophic, with a smooth surface and no regenerative nodules. A liver biopsy revealed severe dissecting fibrosis with Mallory bodies, periportal inflammation, and diffuse fat. A right hepatic vein was catheterized and appeared normal; the wedge hepatic venous pressure was 2.5 mm Hg above I.V.C. A first EEG recording, performed during the period of severe neurological disturbance, showed slow cerebral activity with diphasic and trapezoidal waves compatible with a portacaval encephalopathy. Recordings were later done during the patient's recovery and from time to time demonstrated the same abnormalities, although not so clearly.

Because of the abnormal liver and of the presence of the neurological syndrome, with no evidence of portal hypertension, a portal venogram was requested (Fig. 2). Intrasplenic pressure was 8 mm Hg; the
spleen, splenic and portal veins were found to be normal, but the right portal vein branch was very narrow, with an atrophic distribution; the left branch had increased dimensions but no collateral vessels to the left lobe could be seen. In fact, the left branch seemed to Anastomose directly with the inferior vena cava through a wide channel. A cavoagram (Figs. 3 and 4) showed the same formation and confirmed the portacaval fistula. The right hepatic vein (Fig. 5) seemed to be normal. A hepatic arteriogram did not reveal any other abnormality.

Ammonia blood concentration was measured in the right hepatic vein (30 µg/100 ml), in the inferior vena cava (96 µg/100 ml), and in the right atrium (80 µg/100 ml). Unfortunately, no ammonia level could be measured inside the fistula itself, as the blood flow was very high and prevented catheterization. Removal of ammonia from the blood was also studied following an intravenous infusion of 800 mg of NH₄Cl. The disappearance curve was within the normal range; however, the EEG recorded during NH₄Cl loading showed transient trapezoidal slow waves.

A blood volume determination was performed by the ⁸¹⁸ red cell labelling method. The result was 7 610 ml, a 37% increase according to weight and surface; the cardiac index (Cardioquant method) was 4.16 l/min/m². Plasma renin activity was 12 ng/l/min in basal conditions, a low value according to our method. Tetra-hydro-aldosterone urinary excretion was less than 4 µg/day.

The patient was discharged on a low protein diet and abstinence from alcohol was recommended. Unfortunately, no follow up is available.

Discussion

Although a full demonstration was not obtained in the absence of direct catheterization, there is strong radiological evidence that the portacaval shunt found in this patient represents a patent ductus venosus. However, the relationship between this abnormality and the patient’s other disorders seems complex.

Most of the unusual portacaval anastomoses have been found in patients with both raised portal pressure and liver cirrhosis. This apparent relationship could be explained by the fact that radiological vascular investigations are performed on these particular patients. However, this finding means also that the presence of a direct shunt of congenital origin between the portal and the caval systems is rarely, if at all, sufficient to prevent the development of portal hypertension. Also, it has become reasonable to assume that these anastomoses are small and non-functional at first (or, at least, that their blood flow is insignificant) and that their anatomical extension is an expression of portal hypertension itself. For this reason, during the simultaneous aggravation of the causal liver cirrhosis (usually in the alcoholic form), their subsequent role was never important enough to keep portal pressure levels down to normal.

In some cases, portal pressure was found to be closer to normal in the presence of a congenital shunt rather than is common in cases of portal hypertension (Détrie and Martini, 1960) but there was no clear relationship between the diameter of the shunt and the portal pressure. Anatomical peculiarities, such as tortuousness of the shunt, were postulated to explain this fact (Détrie and Martini, 1960; Paris, Gerard, Toison et al, 1969).

Splenorenal and umbilical derivations (Cruveilhier-Baumgarten syndrome) have often been observed. Other anatomical types have rarely been found; they include observations of splenocaval fistulae (Détrie and Martini, 1960) and venous formations having the characteristics of a persistent—or recanalized—ductus venosus (Détrie and Martini, 1960; Boquien, Morin, Leger, Roy-Camille and Graveleau, 1961; Coppo and Agnolucci, 1968; Neidballa, 1966).

The opposite concept has been suggested by the demonstration of the existence of similar congenital malformations in normal people, ie, without portal hypertension or liver cirrhosis. This has been shown by casual observations of patients with various diseases (Carol, Paraf, and Schwarzmman, 1951; Piccone, Lentino, and Le Veen, 1967) and by group necropsies (Carol et al, 1951). This finding indicates that such a shunt could develop in the absence of portal hypertension. Conflicting reports on what becomes of the ductus venosus point either to the constant early obliteration of this channel (Fontan, 1911; Ostroverkhov and Nikolsky, 1968, Asuncion and Silva, 1971) or to its persistence, with only interruption of its function (Doviner, 1962). The existence of a fully functioning ductus venosus has rarely been reported (Carol et al, 1951).

A stimulating problem is created by the recent observation of several cases of congenital portacaval fistulae with coexistent liver disease, but with strictly normal portal pressure (Darnis, Mosse, Gutmann, Khalifat, Amigue, and Gillot, 1966; Mahoudeau, Dubrisay, and Ruff, 1965; Price, Voorhees, and Blakemore, 1963; Raskin, Price, and Fishman, 1964). The usual initial symptom is a recurrent encephalopathy with hyper ammoniemia, without any clinical evidence of portal hypertension (Darnis et al, 1966; Raskin et al, 1964). We observed this fact in the present case, and it was also found in patients with other types of congenital anastomoses (Price et al, 1963; Raskin et al, 1964). Thus, there is a strong relationship between portacaval encephalopathy and concomitant liver dysfunction. Years ago, experi-
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mental studies (Eck's fistulae) and partial hepatectomies gave identical findings, mainly that neurological disorders do not occur until hepatic function becomes severely impaired (Issami, Pack, Miller, Vanamee, Randall, and Roberts 1956; Liebowitz, 1959). These facts seem to imply that the neurological symptoms, which are presumably due to ammonia intoxication, are related to a decrease in the hepatic clearance of ammonia rather than to the direct consequence of the portacaval shunt (Coppo and Agnolucci, 1968; Léger and Henriot, 1971). However, our patient still demonstrated a certain capacity for urea synthesis, because the hepatic vein level of ammonia was lower than found elsewhere in the body, and removal of infused ammonia was still possible.

An interesting question is the pathogenesis of the liver alteration that we observed. Although the microscopic studies of the liver showed lesions suggestive of an alcoholic origin, we were reluctant to ascribe them to this aetiology only. Several facts—such as the macroscopic aspect of the liver, the absence of nodular formations, and the age of the patient—support the concept that chronic portal bypass of the liver may have a functional significance. Most of the blood appeared in fact to be diverted through the fistula. Perhaps our patient's subnormal mentality itself resulted from chronic cerebral ammonia intoxication due to the congenital fully functioning shunt. Severe hepatic fibrosis has been observed with types of shunt other than Aranti's duct (Price et al., 1963); the absence of any ethanol intoxication could be verified in some cases (Raskin et al., 1964; Darnis et al., 1966). Experimental studies support the conclusion that chronic portal bypass of the liver leads to fibrosis and atrophy of the gland. This fact is presumably related to the diversion of hepatotrophic substances from portal blood origin or to the decrease of blood flow itself through the liver (Fisher, Fisher, and Lee, 1967).

Lastly, we should like to emphasize the existence of systemic haemodynamic abnormalities. A hyperdynamic condition was demonstrated in our patient, with an increased blood volume and a high cardiac output. These changes appeared to be related to the huge, longstanding fistula, although it was a venovenous shunt. Predominance of the dynamic condition upon the hepatic alteration was indicated by decreased aldosterone secretion.

Conclusion

Our patient seems to represent an exceptional case of patent duc tus venosus. The vascular malformation appears to have had an important role from the very beginning of the patient's life, with possible consequences for his mental state. As a clinical demonstration of an Eck's fistula, it is postulated that chronic diversion of the portal blood from the liver can lead to atrophy of the gland and sensitize it to secondary alcoholic damage.

References


Fontan, C. L. M. J. (1911). Le canal veineux d'Arantius. (Thesis (Lille), no. 26), Lille.


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