CAVERNOUS TRANSFORMATION OF THE PORTAL VEIN FOLLOWING UMBILICAL SEPSIS

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The interpretation of splenomegaly in childhood is often difficult. When infective conditions, blood dyscrasias, reticuloses, lipoidoses, and cirrhosis of the liver have been excluded, there remain some cases of splenomegaly in which no obvious disorder can be demonstrated. A number have formerly been diagnosed as Banti's disease, the existence of which as an entity is doubtful. The syndrome is no longer regarded as a primary condition (Sherlock, 1955). A variety of conditions resulting in portal hypertension—for example, cirrhosis of the liver and obstruction of the portal vein—may produce it. The use of the descriptive term "chronic congestive splenomegaly" is therefore preferable.

Whipple (1945) classified portal hypertension into two major categories—those in which obstruction to portal blood flow is intrahepatic and those in which obstruction is extrahepatic. Cirrhosis of the liver is by far the commonest cause of intrahepatic portal obstruction.

Extrahepatic portal obstruction is commonly due to thrombosis of the portal vein. Less commonly, cavernous transformation of the portal vein may cause extrahepatic portal obstruction. The condition has often been reviewed, notably by Klemperer (1928) and more recently by Gibson and Richards (1955) and Parker and Seal (1955), and with reference to the surgery of portal hypertension by Learmonth (1951), Walker (1954), Hunt and Whittard (1954), and Schuckmell, Grove, and Remenchik (1955).

Pick (1909) first postulated that the lesion was an angioma, and Walker (1954) stated that it was congenital in origin, a true venous angioma. Gibson and Richards (1955) reject the previously used term "cavernomatous" transformation in preference for cavernous transformation, which was used by earlier German writers. They state that the condition is seldom if ever a hamartoma and never a true neoplasm. Together with Parker and Seal (1955), on the basis of post-mortem findings, they regard thrombosis of the portal vein and its tributaries as of major importance in the pathogenesis of cavernous transformation, recanalization, and the development of collateral circulation resulting in the cavernous picture. Although historical evidence was lacking in their cases, they concluded that thrombosis was probably a complication of past intra-abdominal infection—for example, umbilical sepsis in the neonatal period and appendicitis—a theory suggested by Wallgren (1927). The possibility of extension of the normal obliterative process in the umbilical vein into the portal vein after birth was not entirely excluded.

Van Crefeld and Levy (1930) suggested that cavernous transformation in their patient, a 4-year-old boy, resulted from the transmission of umbilical infection along the umbilical vein to involve the portal veins. Cavernous transformation of the portal vein in the case reported here, proved at percutaneous splenic venography, is of interest because of the positive history of umbilical sepsis

and of the presentation as splenomegaly without any of the complicating features of portal hypertension, particularly bleeding from oesophageal varices.

Case Report

A girl of 7½ years was first seen after an exacerbation of upper abdominal pain which she had had for the previous two years. The spleen was palpable three-fingerbreadths below the costal margin and she was referred to Great Ormond Street.

The history on admission was that she had been born of a normal pregnancy and there had been no neonatal jaundice. Apart from mild bouts of abdominal pain she had been an active healthy child. Her growth and development had been normal.

She had never been abroad, and there was no past history suggestive of hepatitis, nor was there a family history of blood disorder. Splenomegaly was the only abnormal physical finding. Routine haematological investigations and liver-function tests were within normal limits. X-ray examination of the bones revealed no infiltrative lesion. The Wassermann reaction was negative. Cirrhosis of the liver was thought to be a possible diagnosis.

She was readmitted for review eighteen months later, when aged 9 years. Since her previous admission she had bruised easily and had had occasional mild abdominal pain. Apart from a cold associated with enlarged cervical lymph nodes for the past fortnight, she had remained well. There had been no episodes of haematemesis or melaena. On direct questioning the mother stated that the child when 17 days old had been admitted to a local hospital with an inflamed anterior abdomen. On inquiry, the hospital reported: "She was suffering from cellulitis . . . arising from sepsis of the umbilicus." She had been treated with penicillin and apparently recovered uneventfully.

On examination she looked well, apart from a few recent bruises on the legs. There was no jaundice, anaemia, or purpura. She had no vascular spiders. The posterior cervical lymph nodes were enlarged and tender on the right side. The throat looked healthy. A firm, non-tender spleen was palpable four-fingerbreadths below the left costal margin. The liver was not palpable, and there was no ascites.

Haematological investigations showed: haemoglobin, 11.6 g. per 100 ml. (79%); film, normal; reticulocytes, 1.2%; white cells, 3,800 per c.mm. (neutrophils 80%, eosinophils 3%, lymphocytes 16%, monocytes 1%); platelet count, 222,000 per c.mm.; E.S.R., 17 mm. per hour. Coagulation time (Lee and White), 3 minutes at 37° C.; bleeding-time, 7 minutes (Duke's method); prothrombin time (1 stage), 11 seconds; red-cell fragility, normal. Bone-marrow examination was normal. Biochemical tests revealed: bilirubin, less than 0.5 mg. per 100 ml.; thymol turbidity, 2 units; zinc sulphate turbidity, 6 units; alkaline phosphatase, 5 (K.A.) units; plasma proteins, albumin 3.91 g., globulin 3.11 g. per 100 ml.; electrophoresis, normal; blood urea, 25 mg. per 100 ml.; urine, no excess urobilin. Barium swallow with Valsalva manoeuvre revealed no oesophageal varices, and stools were negative for occult blood. Repeat x-ray examination of the bones revealed nothing abnormal.

The cervical lymph nodes subsided in a few days, confirming their infective origin. Further blood counts continued to show mild anaemia (haemoglobin, 79 to 84%), leucopenia (W.B.C., 2,300 to 3,800 per c.mm.), and thrombocytopenia (platelets, 159,000 to 184,000 per c.mm.). The cause of the splenomegaly remained uncertain. A provisional diagnosis of portal hypertension probably due to extrahepatic portal venous obstruction was made. Cirrhosis of the liver seemed an unlikely cause because of her normal development and absence of clinical stigmata of the disease. She was referred to Dr. Sheila Sherlock at Hammersmith Hospital, where a percutaneous transplenic portal venogram

was done under general anaesthesia. At the same time a liver biopsy was obtained, the consistency of which felt normal and microscopically showed no abnormalities.

At portal venography, as described by Atkinson, Barnett, Sherlock, and Steiner (1955), splenic pressure was 10 mm. Hg (normal 3–17 mm. Hg). Dr. R. E. Steiner's report on the venogram stated that the film clearly demonstrated extensive cavernous transformation of the portal vein. Some of the contrast medium had in fact reached the liver and a lot of blood was shunted through extensive gastric varices and mediastinal veins into the azygos vein (see Fig.).

Percutaneous transsplenic venogram showing extensive cavernous transformation of the portal vein.

To conclude, this patient had extrahepatic portal obstruction, doubtless resulting from thrombosis of the portal vein following neonatal umbilical sepsis.

Discussion

Cavernous transformation of the portal vein is most probably a sequel to portal vein thrombosis, which begins as a complication of one or other intra-abdominal infection. Umbilical sepsis in infancy could certainly cause portal thrombosis without recognizable signs at the time. In later life it might be difficult, if not impossible, to prove that this has occurred.

A history of umbilical sepsis was wanting on first admission. Even after portal venography a congenital anomaly was thought a possible explanation for the cavernous transformation. The documented history of neonatal umbilical sepsis was not learnt until later. Despite adequate treatment it had been followed by portal venous obstruction. The importance of umbilical sepsis as a cause of intrahepatic portal obstruction is particularly significant in the paediatric age group.

The clinical diagnosis of extrahepatic portal venous obstruction was confirmed by portal venography. Until recently extrahepatic portal obstruction could be diagnosed only at laparotomy. In the last five years percutaneous transsplenic portal venography has become a recognized procedure in the clinical investigation of suspected cases of portal hypertension (Atkinson *et al.*, 1955). It is possible to make accurate diagnosis and locate an obstruction or other anomaly of the portal vein. A greater proportion of oesophageal varices are revealed in this way than either by barium swallow or by oesophagoscopy alone or in combination. With careful technique it is free of serious risk in selected cases.

With development of surgical operations for the relief of portal hypertension, portal venography is advisable to diagnose the site and nature of obstruction. The venogram obtained determines the most appropriate procedure. Thrombotic occlusion of the portal vein or its tributaries virtually precludes porto-splenic anastomotic operations, as the part of splenic or portal vein remaining patent is usually inadequate for an anastomotic operation.

Haematemesis from bleeding oesophageal varices is a common complication of portal hypertension from extrahepatic portal obstruction. Because the patient has had none, portal hypertension is unlikely to be marked. The impression is confirmed by a normal intrasplenic pressure at venography, and it looks as if the colossal collateral azygos circulation has relieved any portal hypertension that might have been present previously.

Although varices were outlined at venography in the above case, it is possible that the child might never suffer from haematemesis.

Chronic neurological complications are particularly common in association with portal venous thrombosis (Sherlock, Summerskill, White, and Phear, 1954). Fortunately, again, the above patient did not show any of the neurological symptoms and signs of porto-systemic encephalopathy.

The blood picture of mild anaemia, leucopenia and thrombocytopenia, and prolonged bleeding-time suggests hypersplenism. She will certainly have to be closely watched with regard to her blood picture.

Abdominal pain of one sort or another is a common symptom in cases of cavernous transformation, and may even suggest intestinal obstruction. Gibson and Richards (1955) offer as explanation minor attacks of thrombosis in peripheral radicles of the portal vein. In three of their cases sudden massive thrombosis of the superior mesenteric vein resulted in fatal intestinal infarction. The attacks of upper abdominal pain in the above case, though mild and transient, need careful observation, but it would be difficult to prove whether or not these are due to minor episodes of thrombosis.

Summary

A case of cavernous transformation of the portal vein following neonatal umbilical sepsis, proved on percutaneous transsplenic portal venography, is described in a 9-year-old girl who first presented with abdominal pain and splenomegaly.

The diagnosis and complications of extrahepatic portal obstruction are discussed with reference to recent views on the aetiology and clinical picture of cavernous transformation of the portal vein. A greater recognition of neonatal umbilical sepsis as a cause of portal hypertension in childhood is indicated.

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