Partial nodular transformation of the caudate lobe of the liver presenting with ascites

M. S. FLETCHER
F.R.C.S.*

D. G. D. WIGHT
M.A., M.R.C.Path.

Addenbrooke's Hospital, Cambridge

Summary

Partial nodular transformation is a rare condition which affects the perihilar portion of the liver and may cause chronic portal hypertension. This case report describes an elderly woman in whom the condition was confined to the caudate lobe, which had become greatly enlarged and, by external compression of the portal vein at the hilum of the liver, had caused portal vein thrombosis and subsequent acute ascites.

Introduction

Many causes of extra-hepatic portal vein thrombosis have been implicated in adults, although a large group often remain idiopathic (Thompson and Sherlock, 1964; Turcotte and Child, 1972). Minor variations in size and configuration of both lobes of the liver are common but major anomalies are rare (Pujari and Deodhare, 1976). The caudate lobe may become enlarged in the Budd-Chiari syndrome (Tavill *et al.*, 1975) but massive enlargement confined to the caudate lobe and causing marked compression and thrombosis of the extra-hepatic portal vein has not previously been described.

Case report

A previously fit 90-year-old spinster was admitted with a 3-day history of progressively severe lower abdominal pain, loose stools, anorexia and increasing abdominal swelling. On examination she was dehydrated and had marked abdominal distension due to ascites. There was considerable lower abdominal tenderness and bowel sounds were scanty. There were no stigmata of chronic liver disease nor other signs of cardiac failure. Investigations on admission were as follows: Hb, 13.3 g/dl; WCC, $8 \times 10^9/l$; PCV, 0.394; ESR, 6 mm in the first hr (Westergren); urea and electrolytes normal; total proteins, 57 g/l; albumin, 31 g/l; serum bilirubin, 22 $\mu\text{mol/l}$; alkaline phosphatase, 62 i.u./l. Urine contained no albumin. Chest X-ray showed some left basal shadowing, and abdominal X-rays

suggested the presence of free fluid with several small bowel fluid levels.

After rehydration with i.v. fluid and a period of observation, the abdominal pain became more severe, rebound tenderness was noted and bowel sounds were absent. At laparotomy 3 litres of clear, pale yellow peritoneal fluid were found and there was venous engorgement of the small intestine, which was moderately dilated throughout its length. The liver was small and a large smooth mass approximately $12 \times 9 \times 8$ cm was found to project forwards and downwards from the caudate lobe, causing marked compression of the portal vein just proximal to its bifurcation (Fig. 1). Proximal to this the vein was dilated and thrombosed. Several biopsies were taken from this mass, but in view of the patient's age and general condition no further procedure was undertaken. Postoperatively, in spite of diuretic therapy and sodium restriction the patient's condition deteriorated and she died on the 10th post-operative day.

At post-mortem, although the liver weighed only 940 g (total body weight 58 kg), the caudate lobe was grossly enlarged and had compressed the extra-hepatic portal vein at its origin from the superior mesenteric and splenic tributaries. On section this resembled the remainder of the liver and had no clear boundary or capsule. There was no other evidence of portal hypertension and the inferior vena cava and hepatic veins were normal. The spleen weighed 168 g.

Histologically, the appearances were somewhat variable. Both in the enlarged caudate lobe and in the remainder of the liver there was evidence of chronic passive congestion with mild centrilobular sinusoidal dilatation and hepatocyte atrophy. In addition within the caudate lobe there was a striking nodularity which had not been apparent macroscopically. There was nodular hyperplasia of the liver cells, largely centred upon portal triads, and here most of the liver cell plates were 2 or more cells thick. These nodules had a pushing margin apparently compressing the adjacent atrophied centrilobular cells (Figs 2 and 3). Portal tracts and efferent

* Present address: Department of Surgery, King's College Hospital, Denmark Hill, London SE5.

FIG. 1. Post-mortem photograph of liver showing a massively enlarged caudate lobe (C) causing marked compression of the portal vein (P).

FIG. 2. Lower power view of the caudate lobe showing marked nodularity of the liver, but a complete absence of fibrosis (HE, $\times 40$).

FIG. 3. Higher magnification of the caudate lobe showing thinning of liver cell plates in the periportal regions with compression of the thinned hepatocytes in the centrilobular region (centre of photomicrograph) (HE, $\times 102.5$).

veins retained their normal relationships and there was no evidence of increased fibrosis and no bile duct proliferation. The appearances were thus of localized nodular hyperplasia as seen in partial nodular transformation, together with relative atrophy of the remaining liver.

Discussion

Major anatomical anomalies of the liver, apart from the well recognized Riedel's lobe (Riedel, 1888), are unusual and are often only discovered incidentally at operation or post-mortem. Accessory lobes of the liver, if pedunculated, may become symptomatic if they undergo torsion and infarction (Omanik and Jablonsky, 1972; Pujari and Deodhare, 1976). Massive enlargement of the caudate lobe is, however, most unusual.

It is now recognized that disproportionate enlargement of the caudate lobe may occur in the Budd-Chiari syndrome (Tavill *et al.*, 1975) and this may be apparent on hepatic scintiscanning with predominant central localization of radio colloid, and on inferior vena cavography which shows a characteristic antero-posterior narrowing. The separate venous drainage of the caudate lobe, directly

into the transhepatic inferior vena cava, is usually preserved when the main hepatic veins are occluded and hypertrophy of the caudate lobe occurs because of its retained perfusion. In this case there was no evidence of hepatic vein thrombosis.

Benign tumours and tumour-like conditions may also affect the caudate lobe. Areas of focal nodular hyperplasia and hepatic adenomas may be 20 cm or more in diameter (Phillips *et al.*, 1973; Gold, Guzman and Rosai, 1978). Lesions of focal nodular hyperplasia have expanding margins but are not encapsulated, and characteristically have a depressed central scar. Microscopically, the latter contains proliferating bile ducts. Similarly, hepatic cell adenomas have pushing margins and at most only a partial capsule but they differ from focal nodular hyperplasia in that they lack fibrosis and bile duct structures.

Most of these tumours present either as incidental findings or with localizing symptoms. However, a separate condition has been identified called partial nodular transformation (Sherlock *et al.*, 1966; Classen *et al.*, 1970; Dick and Gresham, 1972) which may cause portal hypertension. In this, the nodules appear to be relatively restricted to the

perihilar portion of the liver where they may cause extrinsic compression upon the hepatic vascular bed. Like the other tumours, this condition is observed almost exclusively in women of childbearing age. It has a somewhat variable histological appearance but has been clearly distinguished from both focal nodular hyperplasia and nodular regenerative hyperplasia (Sherlock *et al.*, 1966). Nodular regenerative hyperplasia is a diffuse nodularity of the liver cells with no or only minimal fibrosis (Steiner, 1959). The condition was reviewed recently by Rougier *et al.* (1978) who found only 11 well documented previous cases and added 6 further cases of their own. Although all their own cases presented with portal hypertension this was found in only about 50% of those previously reported. However, in all cases it was a diffuse disease, apparently affecting all parts of the liver equally. Nevertheless, the changes seen in the caudate lobe in this case were histologically indistinguishable from nodular regenerative hyperplasia, but the main part of the liver was unaffected. The present case thus appears to be a localized form of nodular regenerative hyperplasia and is similar in this respect to the previously reported cases of partial nodular transformation. It is uncertain whether this condition is reactive, neoplastic or a malformation and the current case sheds little light upon this problem. Most of the previously reported cases have been in women of childbearing age and this is thus an exception in this respect. The most common predisposing conditions associated with the diffuse form of nodular regenerative hyperplasia are rheumatoid arthritis and congestive heart failure. There was no evidence of rheumatoid disease in this patient but, although there was little clinical evidence of congestive heart failure, there was centrilobular sinusoidal dilatation in the liver together with moderate cardiac enlargement (470 g) attributable to essential hypertension.

The main clinical feature in this case was the rapid development of ascites in a previously fit patient which caused severe abdominal pain and peritonism. This suggests that the portal vein thrombosis was the cause of the ascites and a recent event attributable to extrinsic compression by the enlarged caudate lobe. The absence of splenic enlargement and other signs of long-standing portal hypertension would support this. In contrast, in the previously reported cases of partial nodular transformation it was

considered that a post-sinusoidal block was the probable mechanism of the long-standing portal hypertension (Sherlock *et al.*, 1966; Classen *et al.*, 1970).

The current case thus appears to be a unique example of partial nodular transformation of the liver in an elderly woman, which presented when it caused extrinsic pressure upon the portal vein.

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