The tumour recurred in 24 cases, in most instances within one year and only rarely after five years. Seven of these recurrences proved fatal despite further treatment in four of them.

The causes of recurrence are discussed. It is thought that inadequate removal of the tumour is the usual explanation but that some late "recurrences" may represent fresh primary growths.

The more radical type of excision is advocated for mixed parotid tumours as a routine, and a technique is described for the early identification of the trunk of the facial nerve at the stylomastoid foramen to minimize the dangers of injury to the nerve and its branches during the course of the operation.

I should like to thank Dr. C. J. P. La Touche, Director of the Department of Radiotherapy, Leeds General Infirmary, for permission to publish details of these cases, which were treated under his care.

REFERENCES

- Adson, A. W., and Ott, W. O. (1923). *Arch. Surg. (Chicago)*, **6**, 739.
 Ariel, I. M., Jerome, A. P., and Pack, G. T. (1954). *Surgery*, **35**, 124.
 Billroth, T. (1859). *Virchows Arch. path. Anat.*, **17**, 357.
 Kaltschmied, C. F. (1752). *Jenae Lit. Tennemannianis*.
 Kaufmann, C. (1881). *Arch. klin. Chir.*, **26**, 672.
 McFarland, J. (1936). *Surg. Gynec. Obstet.*, **63**, 457.
 Martin, H. (1952). *Surgery*, **31**, 670.
 Patey, D. H. (1930). *Brit. J. Surg.*, **18**, 241.
 Plaut, J. A. (1942). *Ann. Surg.*, **116**, 43.
 Shucksmith, H. S., Boyle, T. McM., and Walls, W. K. J. (1951). *British Medical Journal*, **2**, 830.
 Sistrunk, W. E. (1921). *Surg. Clin. N. Amer.*, **1**, 1515.
 Virchow, R. (1863). *Vorlesungen über Pathologie*, Bd. 2. Hirschwald, Berlin.
 Willis, R. A. (1948). *Pathology of Tumours*. Butterworth, London.

PORTAL HYPERTENSION DUE TO EXTRAHEPATIC OBSTRUCTION

BY

A. DAS, M.B., M.R.C.P.

Professor of Medicine

AND

A. K. BASU, M.S., F.R.C.S., F.A.C.S.

Professor of Surgery

Nilratan Sircar Medical College and Hospital, Calcutta

Portal hypertension may be due to intrahepatic or extrahepatic causes. Amongst the latter, the more important recognized aetiological factors may be congenital or acquired stenosis of the portal system of veins; compression by tumours, cysts, or cicatrix; thrombosis due to inflammation or traumatic causes; and, lastly, by what is called cavernomatous transformation of the portal vein. Extrahepatic obstruction of the portal venous system of this nature may produce the syndrome of splenic anaemia with recurrent attacks of haematemesis as its chief symptom. So far, the exact nature and site of the obstruction could not be determined by clinical investigation; only at necropsy, or sometimes at operation, was this possible.

Of the four cases which form the basis of this communication, one was due to obstruction of the splenic vein by a cicatrix over the middle of the anterior surface of the pancreas and two were due to cavernomatous transformation of the portal vein. A fourth case was caused by obstruction of the splenic vein by a pancreatic mass involving the body and tail of the pancreas. In three of these cases the exact diagnosis of the site of obstruction of the portal system of veins was arrived at by splenoportal venography and was later confirmed in the first case at necropsy and in the other two cases at operation.

Case 1

A woman aged 35 was first admitted to hospital with several attacks of haematemesis on August 7, 1953. She had also vomited blood three months prior to this attack. Her past history was not significant. She had no history of abdominal pain or acidity.

The only abnormal findings on admission were severe anaemia and a palpable spleen (two fingerbreadths below the costal margin). She was investigated after her general condition and anaemia had improved. There was no ulcer in the stomach and duodenum on barium-meal examination and no varices could be detected in the oesophagus. Other investigations proved negative. She was discharged, and at this time the spleen was three fingerbreadths below the costal margin. She refused splenectomy. The clinical diagnosis was splenic anaemia with haematemesis.

The patient was readmitted on December 7 with a history of two severe haematemeses on the same day. Physical examination was negative except for evidence of severe haemorrhage and an enlarged spleen, which was just palpable. Hb was 30% and the red-cell count 1,320,000 per c.mm. She was resuscitated by several blood transfusions. The bone marrow showed normoblastic hyperplasia and was otherwise normal. W.R. was negative and thymol turbidity was 10 units. Her anaemia improved with treatment, and the spleen was enlarged to four fingerbreadths below the costal margin and firm in consistency.

She was reinvestigated radiologically. No ulcers were present in the stomach or duodenum, but oesophageal varices were demonstrated. A splenic venogram revealed obstruction of the splenic vein at about its middle and numerous collaterals were seen (Fig. 1). We thought that the haematemeses came from oesophageal varices due to obstruction of the splenic vein, and a splenectomy was decided upon. Just before her transfer to the surgical side she had several bouts of haematemesis and collapsed. Repeated blood transfusions were given to her, but unfortunately she died on February 22, 1954.

A partial post-mortem examination was done. The gross appearances were as follows: Liver was not cirrhotic, but looked slightly fatty and enlarged. The spleen was enlarged

FIG. 1.—Splenic venogram (Case 1) showing obstruction of the splenic vein and upwardly directed collaterals.

FIG. 2.—Photograph of necropsy specimen (Case 1).

to about five times its normal size (Fig. 2). On dissection, the splenic vein was found embedded in adhesions in the middle of its course and was surrounded by what appeared to be dense fibrous tissue. The rest of the splenic vein was normal in appearance. The portal vein beyond the obstruction was patent with normal walls. The stomach and duodenum showed no ulcer. The small portion of the oesophagus removed at partial necropsy did not show any obvious varices. The splenic vein near the pedicle of the spleen was opened; a rubber catheter was inserted into it and tied, barium solution was injected under pressure, and a skiagram of the specimen was taken (Fig. 3). It showed

FIG. 3.—Skiagram of necropsy specimen with barium solution injected into splenic vein (Case 1).

an obstruction of the barium solution at about the site where the dye had failed to pass on percutaneous splenic venography. The catheter was next pushed further into the splenic vein and was obstructed at the same site. The histological appearances showed, amongst other findings, that the liver was not cirrhotic, and that the pancreas was normal. The spleen showed normal sinuses, with some hyperplasia of the pulp and increase of eosinophil and reticular cells. The fibrous tissue around the splenic vein was confirmed histologically as such.

Case 2

A woman aged 21 was admitted on July 5, 1954, with a history of recurrent attacks of haematemesis from the age of 4 years. The last haematemesis occurred six weeks before admission. Some of these attacks had required several blood transfusions for resuscitation. On admission there

FIG. 4.—Splenic venogram (Case 2) with upwardly directed collaterals and diffusely distributed dye at the site of the portal vein (cavernomatous transformation).

was slight anaemia but no jaundice and no oedema or ascites. The spleen was palpable four fingerbreadths below the costal margin. The liver was not palpable. Findings in the heart suggested pulmonary stenosis, which was confirmed by radiological and electrocardiographic appearances. She was two months pregnant at that time. Haematological investigation had been carried out systematically for one and a half years before admission and had showed a persistent moderate degree of anaemia, the haemoglobin ranging from 8.4 to 9.6 g. per 100 ml., and red blood cells numbering 2,500,000 to 3,800,000 per c.mm. There was also persistent leucopenia during this period; the total white blood cell count varying between 1,500 and 3,000 per c.mm. The platelets were also diminished. The bone marrow showed normoblastic hyperplasia. Other features were compatible with hypersplenism. Thymol turbidity was 4 units. Renal function tests were normal. W.R. was negative.

Radiological investigations showed a varicose condition of the oesophagus and a part of the upper portion of the stomach. On splenoportal venography a cavernomatous state of the portal vein with development of collateral circulation was demonstrated; the intrahepatic channels were not seen (Figs. 4 and 5).

An operation was performed on August 7, the findings being as follows. The spleen was enlarged up to the umbilicus. It was soft in consistency. The liver was normal in appearance. The portal pressure was 330 mm. of saline. There were large tortuous veins crossing the lesser omentum and directed towards the oesophagus. These were mostly situated near the right edge of the lesser omentum. In addition to these there were numerous venous tributaries at the site of the portal vein which could not be identified as a single entity. The intrasplenic pressure was 310 mm. of saline. The spleen was removed, but, as there was a hypotensive phase after this, lieno-renal anastomosis was not performed. The weight of the spleen was 930 g. On the fifth day after operation she had an attack of severe haematemesis which required two blood transfusions. After this she had an uneventful post-operative course and gave birth to a full-term child seven months later.

FIG. 5.—Diagram of Fig. 4, showing collateral channels of the (a) short gastric and (b) the left gastric venous systems.

Case 3

A man aged 24 was admitted on December 1, 1954, with a history of five attacks of haematemesis and melaena within the last one and a half years. The first attack was in April, 1953, and the last in July, 1954. Each haematemesis was large in amount and was followed by melaena. After each attack he became severely anaemic. There was no significant past history. The patient had been operated upon elsewhere on May 13, 1953, but the surgeon could not find any cause for the haematemesis, and the abdomen was closed.

On admission he was pale and emaciated. There was no jaundice, oedema, or ascites. The spleen was palpable two fingerbreadths below the costal margin. The liver was not palpable. Examination of other systems was negative. The bone marrow showed active normoblastic erythropoiesis. On radiological examination there was no evidence of peptic ulcer but the lower two-thirds of the oesophagus showed varices. The patient's anaemia and general condition improved with medical treatment, and splenectomy with lieno-renal anastomosis was performed on December 9, 1954. The

portal vein was absent, and overlying the lesser omentum there were numerous tortuous collateral venous channels coursing towards the liver and oesophagus. The portal pressure in the gastro-epiploic vein was 350 mm. of saline. The liver appeared normal both in size and in consistency. The weight of the spleen was 420 g. The histology of the liver and spleen were normal. The patient had a stormy post-operative course for the first few days, with several attacks of haematemesis and melaena, and required blood transfusions. Since then he has improved in health and has had no further attacks of bleeding. Post-operative pyelography has shown good elimination of dye through both kidneys. The oesophageal varices are much diminished in size.

Case 4

This patient, aged 55, was admitted on March 25, 1955, with a history of occasional pain in the epigastrium for 10 years and recurrent haematemesis and melaena since 1952. An exploratory abdominal operation had been performed in July, 1954, it being recorded that large dilated and tortuous veins involved the gastro-epiploic system, and that there was a mass involving the body and tail of the pancreas. The abdomen had been closed without any intervention.

On admission the patient was very anaemic. The spleen was two fingerbreadths below the costal margin. The liver was not palpable. There was no tenderness on palpation of the abdomen and no mass could be felt. There was no ascites. Hb was 45%; R.B.C., 2,400,000 per c.mm.; W.B.C., 3,100 per c.mm. The bone marrow showed normoblastic erythropoiesis. W.R. was negative. Radiological investigation showed no oesophageal varices. On spleno-portal venography (Fig. 6), the dye in the splenic vein

FIG. 6.—Splenic venogram (Case 4) showing obstruction of the splenic vein near the hilum, and dilated collaterals.

abruptly changed direction and coursed upwards towards the under surface of the left leaf of the diaphragm through several dilated and tortuous channels. No dye was found in the rest of the splenic vein or in the portal vein. The appearance suggested obstruction of the splenic vein close to the hilum of the spleen.

At operation the spleen was found slightly enlarged. There were large tortuous veins involving the gastro-epiploic system. From these dilated veins two large tributaries coursed towards the hilum of the spleen. They joined together close to the hilum and then joined the main splenic vein. Pressure in the gastro-epiploic system was 160 mm. of saline. The liver was normal in appearance. Two large masses were found to involve the anterior surface of the body of the pancreas and pressing on the splenic vein. They were mostly firm in consistency though soft at places. Splenectomy was performed. The dilated collaterals were ligated and partly excised. Pancreatic biopsy showed evidence of chronic inflammation. The histological appearance of the liver was normal, but the spleen showed dilated sinuses with erythrophagocytosis.

Discussion

It is now realized that, in a fair number of cases of splenic anaemia syndrome with haematemesis and melaena as their chief features, the cause of the portal hypertension is some form of mechanical obstruction in the portal system of veins outside the liver. Diagnosis of the cause of bleeding in many obscure cases of haematemesis and melaena, and management of this form of obstruction, would be facilitated if it were possible, pre-operatively, to determine the exact anatomical pattern of the portal system of veins together with the site and the nature of the obstruction. In the course of our investigations on splenomegalies of different types we have found splenoportal venography a valuable method of accurately visualizing the portal system of veins. In three of the cases here reported, such investigation enabled us to locate the obstruction in the portal system of veins at different sites and gave us some information about its nature.

It will be seen from Fig. 1 that the dye is obstructed abruptly about the middle of the splenic vein. The portal vein and the intrahepatic channels are not seen at all. The existence of portal hypertension is also suggested by the numerous collaterals found coursing both above and below the splenic vein. It will also be evident that the main upwardly directed collateral belongs to the short gastric system of veins obviously passing to the greater curvature area of the stomach.

In Case 2 spleno-portal venography (Fig. 4) showed the splenic vein in its entirety. At the expected site of the portal vein the dye was diffusely and irregularly spread out, thus definitely suggesting cavernomatous transformation of the avascular channel at this site. That this cavernomatous transformation had produced marked portal hypertension is shown (Fig. 5) by the prominent upwardly directed collateral channels—of both the short gastric (a) and the left gastric (b) venous systems—the latter passing up by the side of the spine towards the cardiac orifice of the stomach.

In Case 4, the dye in the splenic vein was obstructed very near the hilum, and the splenic vein distal to this was not seen at all. The dye was directed instead through several dilated and tortuous channels, presumably collaterals, towards the left leaf of the diaphragm. It was thus possible to assume that the splenic vein was obstructed near the hilum and that the bleeding was due to diversion of blood which could reach the portal or the systemic circulation only through the vasa brevia or the gastro-epiploic system.

The site of obstruction, the nature of the anatomical abnormality, and the course of the collaterals were subsequently confirmed in Case 1 at necropsy, and in the other three cases at operation. The liver was normal in appearance in all four cases, and its histology did not show evidence of any cirrhotic change. Portal pressure taken at operation in Cases 2 and 3 was considerably raised. In Case 4 it was slightly raised. Both these findings point to the possibility that operation directed towards reduction of such portal hypertension would be adequate to relieve haematemesis, which is the common cause of death in these cases.

At necropsy in Case 1 the cause of obstruction of the splenic vein was found to be a cicatrix, and it can be suggested that this was produced by localized subacute pancreatitis. In Case 2 the anatomical abnormality in the portal system was due to cavernomatous transformation, which is likely to be a congenital condition. The existence of congenital pulmonary stenosis in this case is interesting and lends support to the view that the abnormality in the portal vein could also be of the same nature.

In Case 3, it was not possible to observe the portal vein adequately by venography. But our clinical diagnosis of extrahepatic obstruction was confirmed by the finding of cavernomatous transformation of the portal vein at operation. The portal pressure was high, and splenectomy with

lieno-renal shunt has stopped further attacks of haematemesis and has led to considerable regression of the oesophageal varices.

At operation in Case 4, it was found that the splenic vein was embedded in a pancreatic mass, which was subsequently proved by histological examination of a biopsy specimen to be due to chronic inflammation. The large tortuous collaterals seen in the venogram actually belonged to the gastro-epiploic system of veins and obviously accounted for the bleeding.

Summary

Four cases of extrahepatic obstruction of the portal system of veins with repeated severe haematemesis and melaena are described. Two were due to cavernomatous transformation of the portal vein and two to obstruction of the splenic vein by chronic inflammatory pancreatic masses.

In three cases splenoportal venography clearly demonstrated the sites of obstruction and their possible nature. Both these findings were subsequently confirmed.

In addition to the routine methods of investigation, the advantages of splenoportal venography in such cases, both in diagnosing the cause of haematemesis and in planning the operation, are commented upon.

We are thankful to Dr. A. K. Datta Gupta, Principal, Nilratan Sircar Medical College, Calcutta, for allowing us to use the records of the cases here reported. We also acknowledge the help given to us by Professor S. Basu and Dr. R. Roychoudhury, of the radiology department, in the radiological investigations. Two of these cases were initially referred to us by Dr. R. N. Chaudhuri, Director of the School of Tropical Medicine, Calcutta, and we wish to express our grateful thanks to him.

REFERENCE

Das, A., and Basu, A. K. (1954). *J. Ass. Physicians India*, 2, 1.

CARCINOMATOSIS OF THE MENINGES

SOME CLINICAL AND PATHOLOGICAL ASPECTS

BY

K. W. G. HEATHFIELD, M.D., M.R.C.P.

Temporary Assistant Neurologist, St. Bartholomew's Hospital, London

AND

J. R. B. WILLIAMS, M.D.

Senior Registrar, Department of Pathology, St. Bartholomew's Hospital, London

Diffuse carcinomatous spread in the meninges was first described by Saenger in 1900, although the neurological features seen in the terminal stages of malignant cachexia had been observed previously by Oppenheim (1888). Since the early German papers there have been many publications on this subject, and Jacobs and Richland (1951) were able to collect 52 cases. Fischer-Williams, Bosanquet, and Daniel (1955) have recently reviewed the condition.

It is probable that carcinomatosis of the meninges is commoner than the literature would lead one to suppose, for there is often no naked-eye abnormality, and unless histological sections are examined the diagnosis is not made. Furthermore, among Jacobs and Richland's cases there were no fewer than ten in which no primary neoplasm was found, and some cases may be missed altogether at necropsy and death be attributed to bronchopneumonia or other cause.

We describe two cases of carcinomatosis of the meninges which demonstrate different aspects of the disease and the possibility of improvement with treatment. The long survival of one—three years from the onset of symptoms—is unique.

Case 1

A labourer aged 68 was admitted to hospital on November 9, 1954, complaining of blindness. Four months previously he felt generally unwell, lacked energy, and had some pain in the neck. He later developed severe attacks of occipital headache, followed by anorexia and constipation. Two weeks before admission he found that his vision was deteriorating and his relatives noticed that he was groping about. On the day of admission he admitted that he was totally blind and had partly lost his hearing. In the past he had suffered from no serious illness apart from a paralysis of the left leg from infancy.

On examination he was confused, with marked neck stiffness and positive Kernig's sign, but afebrile. There was complete anosmia. Both eyes showed no perception to light, but the optic fundi were normal. The pupils were small and irregular, and did not react to light, though they reacted to attempted convergence. A rotary nystagmus was present with weakness of both external rectus muscles and partial dissociation of conjugate ocular gaze. There was slight right facial weakness of central type with partial bilateral deafness. No other abnormal neurological signs were found apart from the old flaccid left foot-drop. General examination showed mild hypertension (blood pressure 150/90 mm. Hg), the lungs and abdomen showed nothing abnormal, but the prostate was slightly enlarged.

The haemoglobin was 14.5 g. per 100 ml., the white cells 1,800 per c.mm., and the sedimentation rate (Westergren) 5 mm. in one hour. X-ray examination of the skull and chest showed no abnormality. Lumbar puncture gave a clear fluid under 300 mm. pressure, containing 28 lymphocytes per c.mm., with clumps of large mononuclear cells. In a Leishman film these had the characters of malignant cells. The fluid also contained protein 160 mg. per 100 ml., chlorides 700 mg. NaCl per 100 ml., and sugar 70 mg. per 100 ml.; the Wassermann reaction was negative. A left carotid angiogram showed separation of the vessels, suggesting internal hydrocephalus. A ventriculogram revealed dilated lateral and third ventricles, the fourth ventricle being normal. An electroencephalogram was not helpful.

The patient remained blind, with severe frontal and occipital headaches, but was still mentally alert until three days before death. He then became very drowsy and died on December 3, 1954.

Necropsy Findings

There was a primary diffuse infiltrating carcinoma of the stomach affecting all layers of the gastric wall, extending from the cardia to the pylorus and involving the lymph nodes of the lesser curvature. No other visceral metastases were found.

The brain showed a fine fibrinous exudate over the base, especially around the optic chiasma, and milky-white thickening of the arachnoid extended all along the spinal cord. The brain after fixation showed no visible metastases. The lateral and third ventricles were dilated, but the fourth ventricle was normal in size.

Histology.—The gastric tumour was a mucus-secreting, poorly differentiated carcinoma composed of sheets of "signet-ring" cells and clumps of small irregular acini. Sections of the base of the brain, including the optic chiasma and hypothalamus, of the cerebral and cerebellar cortices, and of the mid-brain, pons, and medulla showed a fine deposit of "signet-ring" carcinoma cells with a little fibrinous exudate scattered diffusely in the pia-arachnoid. Over the cerebrum the deposits were greatest between the convolutions. Similar tumour tissue was present in the arachnoid of the spinal cord and extended in the perineural