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Medical Memoranda

Retroperitoneal Fibrosis Causing Localized Portal Hypertension

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A single case of portal hypertension due to portal vein compression by retroperitoneal fibrosis has been recorded (Eiseman and Yeoh, 1962). We present here the first report of retroperitoneal fibrosis causing localized portal hypertension.

CASE REPORT

This male patient first presented at another hospital in 1964 at the age of 44 complaining of intermittent pain in the left loin. Investigations showed: haemoglobin 6.7 g./100 ml.; serum iron 10 mg./100 ml.; barium meal and barium enema examinations negative, and blood urea 36 mg./100 ml. Intravenous pyelography showed no excretion of contrast medium on the left side, and a retrograde pyelogram showed a left hydronephrosis. A left nephrectomy was therefore performed. Before discharge he had a haematemesis, but repeat barium studies showed nothing abnormal.

In 1967 he was transferred to St. Thomas's Hospital for investigation of oedema of his legs. There was no clinical evidence of cardiac failure. Examination showed haemoglobin 6.9 g./100 ml.; E.S.R. 54 mm./hour; serum iron 35 mg./100 ml.; iron-binding capacity 385 mg./100 ml.; blood urea 62 mg./100.; plasma creatinine 3.5 mg./100 ml.; and creatinine clearance 30 ml./minute. Liver function was normal.

A barium meal examination showed no abnormality, and in particular no evidence of oesophageal varices. An intravenous pyelogram showed dilatation of the calices of the right kidney, and the right ureter was displaced medially. The histology of the left kidney was reviewed. Fibrous tissue was seen to be infiltrating the renal capsule, and a diagnosis of retroperitoneal fibrosis was made. An inferior vena cavagram showed pronounced narrowing at the level of the second lumbar vertebra.

While under investigation he had two further haematemeses, and laparotomy was therefore performed. This revealed a large fibrous plaque obstructing the inferior vena cava, dilated veins over the pylorus and greater curve of the stomach, a small spleen, but no demonstrable splenic vein. A portal venogram through the gastroepiploic vein showed no evidence of obstruction, and the portal venous pressure was 120 mm. of saline. Despite these findings, it seemed that this situation could be best explained on the basis of portal hypertension. A repeat cavagram showed that the inferior vena cava was not suitable for a shunt, and so the patient was managed conservatively.

During the next year he had several more haematemeses requiring transfusion, but otherwise remained well and continued at work. In October 1968 a barium meal showed oesophageal varices. A coeliac axis arteriogram (see Fig.) showed the splenic artery to be tortuous and grossly dilated. A transsplenic portal venogram showed total occlusion of the splenic vein as far as its junction with the portal vein, with extensive varices around the spleen, fundus of stomach, and lower oesophagus.

At a second laparotomy very extensive retroperitoneal fibrosis was found involving the left gastric pedicle, the posterior wall of the stomach, the posterior half of the spleen, and all the vessels in the splenic hilum. There were massive varices over the stomach. A portal venogram showed occlusion of the splenic vein, with

a patent superior mesenteric vein entering the portal vein. The portal pressure was again normal.

The stomach was mobilized with great difficulty; it could not be separated from the spleen, and a splenectomy was therefore carried out. Portoazygos disconnection was then achieved by gastric transection. He made a good initial recovery but later developed peritonitis, and subsequent hepatic and renal failure. These did not respond to medical treatment, including peritoneal dialysis, and he died two weeks after operation.

Necropsy showed breakdown of the suture line of the gastric transection. The liver was macroscopically normal. The splenic artery and vein ran through dense fibrous tissue around the pancreas; the splenic vein was practically obliterated by external compression. The superior mesenteric and portal veins were normal. There was a hydronephrosis of the right kidney, and the ureter was involved in fibrous tissue from just below the kidney. The abdominal aorta and inferior vena cava were surrounded by fibrous tissue from the level of the coeliac axis to just below the pelvic brim. Histology confirmed widespread intra-abdominal retroperitoneal fibrosis. The liver showed evidence of bile stasis, but there were no other gross pathological changes.

COMMENT

In this patient, the rare situation of localized portal hypertension followed involvement of the gastric and splenic veins by retroperitoneal fibrosis with sparing of the portal vein.

Initially the cause of the gastrointestinal haemorrhage could not be established as barium studies were repeatedly negative and oesophageal varices were not seen until very late. While at the first laparotomy a diagnosis of portal hypertension seemed likely, the site of the portal obstruction was not found, and not until a year later, at a second laparotomy, was the diagnosis fully established. The disease process

Selective coeliac axis arteriogram showing considerably dilated and tortuous splenic artery.

was then so far advanced as to make surgery extremely difficult.

The hypertension found in the upper half of the stomach and lower oesophagus was probably due to (1) the increased inflow from the large spleen and its enlarged tortuous artery; (2) obstruction of the left gastric, splenic, and left gastroepiploic veins by fibrosis; and (3) high pressure in the normally low-pressure azygos system, since these veins were acting as collaterals for the venous return from the legs. No previous report of this complication could be found on reviewing the literature.

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Multiple Sclerosis in the African

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There is some doubt about whether multiple sclerosis occurs in indigenous Africans (Gelfand, 1957; Trowell, 1960; Haddock, 1965; Dean, 1967). During the period 1952-69 we have encountered only two cases, and these we report here.

CASE 1

The patient, a man aged 30, was admitted to the Kenyatta National Hospital on 9 July 1968 with a five-month history of difficulty in walking and weakness of the lower limbs. There were no constitutional symptoms. The onset of the disease was fairly rapid, occurring over a period of five days, starting with tremors in the left leg, unsteadiness of gait, and weakness in both legs, though he was still able to walk. His legs became progressively weaker and he noted burning sensations in his fingers. Ten days before admission he complained of being unable to control his urine. In June 1967 he had been in another hospital with similar complaints. He had stayed there for a month, making a slow but incomplete recovery.

Examination showed nystagmus, with a quick movement to the left, diplopia on looking to the right, and a doubtful left facial weakness. The optic discs were normal. Cerebellar ataxia and hypotonia were present in both arms, but more pronounced on the left. There were no sensory changes in the arms. The abdominal reflexes were absent. The legs were weak and spastic, with brisk reflexes and extensor plantar responses. There was some loss of vibration sense and slightly impaired position sense in the feet, with no loss of light touch or pain sensation. There was inco-ordination of movement in both legs of a cerebellar type and he was unable to stand. He was incontinent of urine.

Nystagmus, diplopia, and incontinence disappeared after a few weeks. All the other signs, however, remained static, though he was gradually able to walk a little with the aid of a stick. He was transferred to another hospital on 4 February 1969, and since then we have had no record of his progress.

Routine investigations were negative. The C.S.F. showed gross abnormalities compatible with multiple sclerosis. Other investigations showed: total phospholipid 99.4 µg/100 ml., lysocleithin 5.9%, sphingomyelin 27.6%, lecithin 54.2%, serine 3.5%, ethanolamine 12.7%, and phosphatidic acid 5.1%. Total protein 63.5 mg./100 ml., alpha-globulin 13.6 mg./100 ml. (21.4% of total); Lange curve 4433321000; and W.R. negative Acrylamide disc electrophoresis: prealbumin 4.6%, albumin 59.3%, beta-globulin 6.1%, IgB 10.6%, and IgA 2.1%.

CASE 2

This patient, a 25-year-old woman, from Machakos district, was seen in January 1966. She gave a two-year history of weak-

ness, heaviness, and paraesthesiae of the left leg, failing vision, recurrent attacks of pain behind the eyes, and incontinence of urine. Her gait was both spastic and unsteady. There was pallor of the optic discs, more pronounced on the temporal side, but no nystagmus or other cranial nerve involvement. The tendon reflexes in the lower limbs were abnormally brisk, particularly the left knee jerk. The plantar responses were doubtfully extensor. The abdominal reflexes were absent. She had an intention tremor of both upper limbs. There was a loss of proprioception over the left leg. Sensation to pain and touch were intact. The C.S.F. showed: protein 25 mg./100 ml., Lange curve 5543211000, and a negative Kahn test. She was discharged after six weeks, when her walking had improved.

Eighteen months later she was readmitted ataxic. She gave a history that she had improved after her return home, but that since the birth of a child three months previously she had been unable to walk and was incontinent. Examination showed bilateral optic atrophy, no nystagmus, and a scanning speech. All deep reflexes were increased and she had left ankle clonus. The abdominal reflexes were present. The plantar responses were extensor. There was pronounced ataxia of the cerebellar type, affecting the legs more than the arms. Investigations were again negative, apart from those of the spinal fluid, which showed an abnormal Lange curve (555222000), a negative Kahn test, and a slightly raised protein (41 mg./100 ml.).

She was given a course of parenteral vitamin B injections. After four months in hospital the clinical picture had not appreciably altered, except that there was some improvement in the control of her bladder.

COMMENT

In these two cases the clinical features conform well with those of multiple sclerosis, though the definite diagnosis is made only at necropsy. Both patients were young, with evidence of involvement of the pyramidal tracts and ataxia of the cerebellar type with minimal sensory changes. They had disseminated lesions in the central nervous system, showing fluctuations in the clinical manifestations, and, lastly, the C.S.F. findings in each case were compatible with the diagnosis. We therefore feel that the diagnosis is justified in both these patients.

Dean (1967) produced evidence to show that the incidence of multiple sclerosis is higher among immigrants to South Africa from Europe than among the white population born in South Africa. He suggested that multiple sclerosis is a viral gastrointestinal infection of infancy and that lack of exposure to the virus in early life predisposes to the development of multiple sclerosis later on. Kuru (Mathews, *et al.*, 1968) has been shown to be due to a slow virus and presents as a progressive subacute cerebellar ataxia. A type of progressive cerebellar ataxia for which no aetiological cause has been established is seen not infrequently in Kenya. It could be that this may be the more usual response in the African to a slow virus if such be the cause of multiple sclerosis. Dietary and immunological factors have been considered as predisposing to the disease, but the applicability of these in Africa, where large sections of the population have little or no medical coverage, is hard to assess. The low incidence of multiple sclerosis in the indigenous African is by no means certain or accounted for.

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