

## Idiopathic Retroperitoneal Fibrosis Involving the Ureters [*Abridged*]

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THE first examples of this disease were reported by Ormond (1948). In 1956 the author reported 7 cases (Raper, 1956); there were then only 10 previously reported cases. There are now 50 reported cases and personal reports to the author of almost another 70 suggest that this disease is not so uncommon as it at first appeared. With such an increased amount of information available a description of the disease entity becomes possible.

*Site of the disease.*—A consistent and puzzling feature of the disease is the site of the fibrosis. Although its extent may vary, its centre is almost always at the level of the fourth and fifth lumbar vertebræ, overlying the aortic bifurcation. The ureters are at the edge of the lesion and are always pulled towards the mid-line (Fig. 1). The width

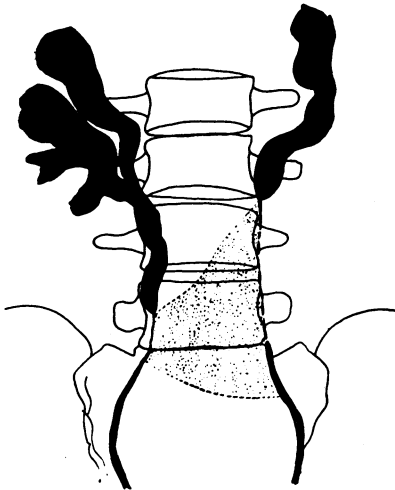


FIG. 1.—Diagram traced from a radiograph of a patient with retroperitoneal fibrosis showing its characteristic position with the ureters involved at each edge.

of the fibrotic area is sometimes no greater than the width of the vertebral bodies. To describe it as peri-ureteric fibrosis ignores the fact that the disease probably arises in the mid-line. The thickness of the fibrous tissue varies from one patient to another and may form a palpable swelling, but more often is described as a plaque or fibrous mat. It may be noticed during a laparotomy as white scars in the base of the mesentery. It may compress or distort the aorta or vena cava, and in one case reported by the author obstructed the common bile duct.

The histology of the fibrous tissue has been described already by Dr. Pugh.

*Symptoms.*—Most patients have been under

the care of a doctor for a few weeks or months before a diagnosis is made. The common symptoms are ill-defined pains and some of the early symptoms such as nausea, weakness and loss of weight may be the result of developing uræmia. Eventually and sometimes with disconcerting speed the patients develop renal pain with or without oliguria. Until these final symptoms, disturbances of micturition are uncommon. In retrospect other symptoms may be considered significant such as claudication or œdema of the legs due to interference with the circulation. Intermittent testicular pain is not uncommon and may be due to involvement of the spermatic vessels.

*Clinical examination* does not usually aid the diagnosis but there may be a palpable tender kidney or central abdominal tenderness.

*Diagnosis by radiography.*—This disease has a characteristic pyelogram, the essential features being an obstruction of one or both ureters in their middle third with the ureter or ureters drawn towards the mid-line in the obstructed area (Fig. 2). If the ureter is incompletely obstructed there will be a hydronephrotic kidney above the block, whereas if the block is complete the kidney is often small and functionless (Fig. 3).

*Treatment.*—Cortisone: The fibrinolytic properties of the corticosteroids make them an attractive group of drugs to use in a disease of this type. However, there are few reports of their use and even these do not conclusively prove their value. I am indebted to Mr. J. F. S. Withycombe for his detailed personal reports of 2 patients in whom prednisone was used to such good effect that it seems only reasonable to suggest its use for other patients. The first patient had previously had a left nephrectomy and was in hospital for investigation of a right hydronephrosis due to retroperitoneal fibrosis when he developed a very painful attack of herpes. To relieve this pain he was given 20 mg of prednisone daily for six weeks. Another intravenous pyelogram showed very considerable improvement in the right hydronephrosis. There has been no deterioration during the following eighteen months. The second patient was admitted to hospital three months after a partial gastrectomy because that operation had not relieved his symptoms and he was persistently vomiting. After a short time in hospital he developed oliguria and renal failure and was too ill for complete investigation. On a provisional diagnosis of polyarteritis nodosa he was given prednisone (80 mg daily dropping to 15 mg daily). There was a dramatic improvement in

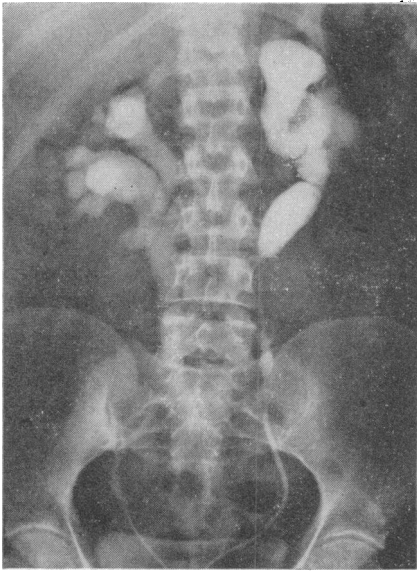


FIG. 2.—Bilateral ascending pyelogram showing bilateral hydronephrosis with a narrow segment at about the centre of each ureter, this area of ureter being closer to the mid-line than normal.

his condition with a diuresis and fall in the blood urea. This allowed a full urological investigation which revealed no function of the left kidney and a right hydronephrosis. Later exploration of his left ureter confirmed the diagnosis of retroperitoneal fibrosis, and with further steroid therapy the right hydronephrosis returned to normal.

**Relief of the uræmia:** In these patients an attempt will probably be made to get a catheter up one or other ureter when investigating the sudden onset of anuria. The catheter may pass up to the kidney without much difficulty for the envelopment of the ureter in a rigid tunnel seems to prevent conduction of urine even before a complete mechanical obstruction has occurred. If a catheter can be passed to the kidney and a good flow of urine results it should be left there to relieve the uræmia. If no catheter can be passed the uræmia should be relieved by dialysis with an artificial kidney or a nephrostomy on the tender kidney.

Ureterolysis is a satisfactory way to treat the ureteric obstruction and in most patients the ureter can be dissected from its bed of fibrous tissue. An approach from the flank is usual but mid-abdominal exposure has the advantage that both ureters can be freed at the same time. It also reveals more accurately the exact site of the disease, and from further observation of this it may be possible to learn more about its cause.

When the ureter has been set free from the fibrous tissue it should be fixed in the iliac fossa

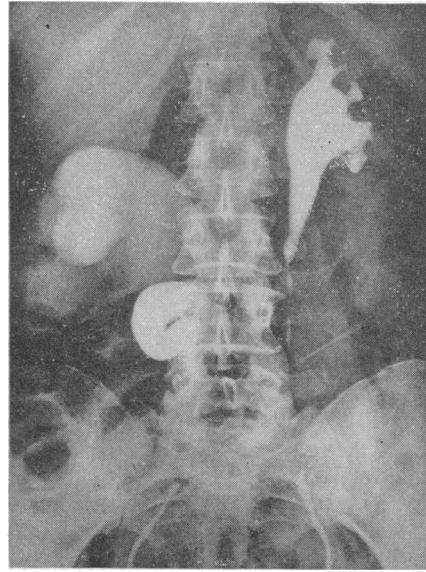


FIG. 3.—Bilateral ascending pyelogram showing a large right hydronephrosis and a small left kidney. Fifteen months earlier both kidneys were hydro-nephrotic but the left has now ceased to function. The medial position of the narrow segment of the ureters is again seen.

by stitches between the posterior peritoneum and the muscles of the posterior abdominal wall medial to the laterally placed ureter. The ureter may be brought within the peritoneal cavity and so far this does not seem to have been followed by any trouble. In one patient, after dissecting out the ureter, there remained a few nodules of fibrous tissue in the wall of the ureter. These nodules have not grown in the succeeding seven years.

**Prognosis.**—The prognosis of this disease can only be assessed by observations over many years. One of the author's patients died from renal failure eleven years after the first symptoms of the disease. Others previously reported (Raper, 1956) are still alive and in none has renal failure or ureteric occlusion advanced. One has had a leg amputated because of severe ischæmic pain and it is possible that vascular occlusion by the retroperitoneal fibrosis played some part in this. Another has developed severe claudication in both legs at the age of 49.

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