Presumptive Ormond's Disease: Remission Following Steroid Therapy

R. E. Reinhard
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Remission Following Steroid Therapy
R. E. Reinhard, M.D.*

Many cases of idiopathic retroperitoneal fibrosis have been reported since Ormond's original description of the disease in 1948. Most clinicians have preferred ureterolysis to other modes of therapy.†,‡

Figure 1
Delayed kidney function on the left side.

*Division of Urology
The following is a case report in which steroids were used primarily on a patient with radiographic changes consistent with idiopathic retroperitoneal fibrosis.

**CASE REPORT**

(L.P.) — A 57 year old colored female was first seen in the Urology Clinic on September 21, 1962, with a 10 day history of intermittent, non-radiating LCVA pain. Physical examination was unremarkable. Urinalysis was negative for sugar and albumin; however, a low grade pyuria was noted on microscopic examination. An intravenous pyelogram showed an essentially normal right pyelogram, but there was marked delay of function and hydronephrosis on the left side (Figure 1). At this time it was thought that the patient most likely had a non-opaque left ureteral calculus and she was placed on sulfa, and followed conservatively. She returned two weeks later asymptomatic and with a clear urine. However, an intravenous pyelogram revealed non-visualization on the left side, and she was admitted to the hospital for further investigation. Past history was significant in that the patient had a cholecystostomy, followed by a cholecystectomy in 1960. She had been treated for hypertension in 1960. Review of systems with particular regard to the gastrointestinal, cardiorespiratory, and genitourinary systems was otherwise non-revealing.

Examination revealed a markedly obese, colored female in no particular distress. The blood pressure was 160/70, with a normal pulse rate and oral temperature. Examination

![Figure 2](image)

**Figure 2**

Narrowing and medial deviation of left ureter.
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of the head, neck and thorax was negative. On abdominal examination, the spleen was felt to be palpable by some observers. No other abdominal masses or organomegaly were felt. Routine laboratory data revealed a hemoglobin of 9.9 grams/100 cc., leucocyte count of 7,850 per cubic centimeter of blood with a normal differential. A urine culture was reported as negative. The VDRL was non-reactive. Serum uric acid 4.5 mg. per cent, serum calcium 9.3 mg. per cent, and serum phosphorus 4.2 mg. per cent. The fasting blood sugar was 75 mg. per cent and BUN 24 mg. per cent. The serum creatinine was 1.1 mg. per cent. A chest x-ray was normal. A cystogram was also normal. Cystoscopy was unremarkable. A ureteral catheter was passed into the left renal pelvis and 30 cubic centimeters of urine, containing leucocytes and erythrocytes, was obtained. A pyeloureterogram demonstrated mild dilation of the pelvis and calyces. The ureter appeared essentially normal except for mild medial deviation.

The patient was discharged on sulfa and followed in the Outpatient Department where a subsequent intravenous pyelogram again showed non-visualization on the left, and a mild right hydronephrosis. At this time a urinalysis showed a low grade pyuria, and she began to notice increasing edema of both lower extremities.

She was readmitted for further urologic evaluation. Physical examination was essentially unchanged except for the presence of 2+ pitting peripheral edema. The hemoglobin 9.2 grams/100 cc., leucocyte count of 7,550 per cubic centimeter of blood with a normal differential. A blood smear revealed a mild microcytic normochromic anemia. The sedimentation rate was slightly elevated. Serum protein electrophoresis showed a slight elevation

Figure 3
Narrowing and medial deviation of right ureter.
of the beta and gamma globulin fractions. The BUN was 14 and serum creatinine 1.3 mg. per cent. Three 24-hour urine specimens for AFB were negative. Bulb pyelograms were done and showed bilateral hydronephrosis with narrowing and medial deviation of both ureters at L-4 (Figures 2 and 3).

The patient was started on prednisone 10 mg. q.i.d., tetracycline 250 mg. q.i.d., and a modified ulcer regime. She was maintained on this program for two weeks, during which time her edema subsided. Subsequent pyelograms have shown gradual improvement in the hydronephrosis (Figure 4). She has been asymptomatic, and her urine has remained clear six months after the onset of symptoms.

Figure 4
Intravenous pyelogram taken six months after onset of symptoms and subsequent improvement in hydronephrosis.

Comment
Radiographically, idiopathic retroperitoneal fibrosis characteristically produces marked narrowing and compression of the ureters with slight medial displacement. In contrast neoplasia, the most common cause of extrinsic ureteral obstruction, generally causes marked distortion of the ureters with lateral and anterior displacement and minimal compression.
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This patient's x-rays were very suggestive of Ormond's disease. Because of her marked obesity, we preferred not to undertake bilateral ureterolysis. Several authors have reported improvement following steroid therapy;\textsuperscript{11-14} therefore, we treated her initially with prednisone.

We did not have a tissue diagnosis, and we are aware that other conditions may radiographically mimic retroperitoneal fibrosis.\textsuperscript{15} Some of these may respond initially to steroids.

It is generally believed the longer standing lesions of idiopathic retroperitoneal fibrosis consist of dense fibrous tissue with some fat and cellular infiltration. In the earlier stages of the disease, the tissue is looser with considerably more fat cells, polymorphonuclear leucocytes, and eosinophils.\textsuperscript{2,4} If this is true, and if the disease is self-limiting as many believe,\textsuperscript{7,10} steroids could conceivably be of value in the earlier stages.

SUMMARY

A patient with a presumptive diagnosis of idiopathic retroperitoneal fibrosis has been presented. She is still in a remission six months after a course of steroid therapy.

REFERENCES


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