Acute Idiopathic Retroperitoneal Fibrosis *

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In 1948 Ormond reported two cases of an apparently new disease entity of unknown etiology characterized by compression of both ureters by a dense, fibrotic retroperitoneal process which also involved the aorta and inferior vena cava.19

Since Ormond's report there have been approximately 70 cases reported which can be classified under the broad term "idiopathic retroperitoneal fibrosis." The symptomatology in the chronic stage is vague and insidious until one or both ureters become obstructed; then symptoms related to ureteral obstruction appear. This is the most frequently described manifestation of the disease.

The two cases forming the basis of this report represent examples of this disease seen at an early stage which simulated acute surgical abdominal disease. In its chronic phase the process may lead to compression of the aorta and vena cava as well as the ureters and kidneys.9,14,20,28

Therefore, the general surgeon should be familiar with this condition.

Case Reports

Case 1. N. T., a 23-year-old Negro woman, entered on the medical service of the Los Angeles County General Hospital on September 17, 1959 with a two-day history of constant, sharp pain in her right flank radiating down into the pelvis and into the right thigh. At the onset the patient felt a "popping sensation" in the right side while moving furniture. The pain gradually increased in intensity over a period of two days, was aggravated by motion and coughing, and was accompanied by chills and fever of 39.0° C. Anorexia and nausea were present but vomiting occurred only once. Bowel movements were normal. There was no history of dysuria, nocturia, or other urinary complaints. There was no past history of direct trauma, syphilis, tuberculosis, or other systemic illnesses. Eighteen months prior to entry the patient had influenza.

Physical Examination: The patient was a well-developed, well-nourished Negro woman in mild acute distress. Pertinent findings included a scaphoid abdomen with moderate guarding and tenderness on the right side. Exquisite tenderness was present in the right flank and right costovertebral angle. Bowel sounds were normal. There was a suggestion of a mass in the region of the right kidney. Pelvic and rectal examination showed no masses or tenderness.

Laboratory: Catheterized urine showed albumin and sugar to be absent; microscopic examination showed 4–5 white cells per high power field and 1 to 2 red cells per high power field. The hemoglobin was 10.9 Gm. The white blood cell count was 9,600. Urine cultures were negative on three specimens.

Hospital Course: A tentative diagnosis of acute pyelonephritis with possible perinephritic abscess was made and antibiotics were administered. Each day the temperature rose to 39.0° C. and her pain gradually increased. Urologic consultation was obtained and an intravenous pyelogram showed good bilateral function, with medial deviation of the right ureter near the pelvic brim. There was no evidence of ureteral obstruction (Fig. 1). Needle aspiration of the right perinephric space was negative. The patient failed to improve with antibiotic therapy. The hemoglobin dropped to 8.5 Gm. with no evidence of bleeding. At this time her temperature was 39.5° C. Positive findings were limited to the abdomen and consisted of slight edema over the right lateral chest with exquisite tenderness over ribs 10 and 11 and right costovertebral angle. A

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definite subhepatic mass could be felt in the region of the right kidney.

Repeat chest x-ray films revealed pleural reaction along the surface of the diaphragm with fluid in the right costophrenic sulcus. The diaphragm was mobile at fluoroscopy. Supine and lateral decubitus views of the abdomen revealed good psoas outlines but the renal contours were obscured. There was distended small bowel and colon suggestive of paralytic ileus.

A diagnosis of right subhepatic abscess was made, probably due to ruptured acute appendicitis. At operation on September 29, 1959, a normal appendix was found. Further exploration revealed a retroperitoneal mass in the region of the right kidney. The peritoneum of the posterior abdominal wall was densely adherent to this mass. No purulent material could be aspirated. The mass was incised at the lower pole of the kidney and found to consist of a fibrotic pseudocapsule measuring one cm. in thickness. This fibrotic pseudocapsule was separated from the kidney by a partially organized hematoma. The kidney was partially freed, biopsies were taken, and the area drained through a right flank stab wound. Unfortunately, the right ureter was not examined. Histopathologic studies were reported as chronic inflammatory tissue consistent with perinephric abscess (Fig. 2).

Subsequent Course: The patient was treated with penicillin, chloramphenicol and strepto-
mycin. The patient's temperature immediately dropped to 37.5° C. and remained normal. Subsequent intravenous pyelogram and retrograde studies revealed adequate function with persistent medial deviation of the middle one-third of the right ureter (Fig. 3). Postoperative urine cultures were positive for Klebsiella-aerobactor and Pseudomonas organisms. Cultures from the drain site grew E. coli.

The patient has continued to do well to the present time.

Case 2. J. H., a 40-year-old Negro man, entered the jail service of the Los Angeles County General Hospital on March 7, 1960, with a history for three days of right chest pain, intermittent chills and fever. The pain was also noted on the right side of the abdomen and was aggravated by coughing. Dysuria was noted for two days prior to admission. There was no nausea or vomiting. No bowel movement had occurred in three days. Past history revealed no exposure to trauma, a prior heroin addiction, and, in 1945, syphilis which was treated with penicillin.

Physical Examination: The patient was a well-developed, well-nourished man in mild distress because of chest pain. The blood pressure was 130/80, the pulse was 100, the respiration was 28, and the temperature was 38.5° C. The lungs showed dullness with inspiratory and expiratory rales at the right base. There was marked right costovertebral angle tenderness with a palpable fullness suggestive of a retroperitoneal mass. The abdomen showed mild distention, guarding, and tenderness in the right lower and mid-abdomen. Peristalsis was present but hypoactive. The rectal examination was normal.

Laboratory: The hemoglobin was 13 Gm.; the white blood cell count was 15,000; the urinalysis was negative. A chest x-ray showed a patchy infiltrate at the right base consistent with bronchopneumonia and the right diaphragm was slightly elevated (Fig. 4). Fluoroscopy revealed limited motion of the right leaf of the diaphragm. The supine and left decubitus x-rays of the abdomen showed mild paralytic ileus. The renal and psoas shadows were sharp. The differential diagnosis was: (1) bronchopneumonia with secondary paralytic ileus; (2) rupture retrocecal acute appendicitis with abscess; (3) right subphrenic abscess with compression atelectasis and pneumonitis; and (4) perinephritic abscess.

Hospital Course: Repeat white blood cell count ten hours after admission was 22,000 with 89 per cent polymorphonuclear neutrophils. Subsequent urinalysis and blood cultures were negative. An intravenous urogram on March 8, 1960

Fig. 3. Persistent medial deviation of the middle third of right ureter two months postoperative.

Fig. 4. Chest x-ray demonstrating a patchy infiltrate at the right lung base consistent with pneumonia; also note the elevation of the right hemi-diaphragm.
showed prompt bilateral excretion without evidence of hydronephrosis (Fig. 5). The urologic consultants noted the fullness in the right lumbar region and excluded the genitourinary tract as the source of the problem.

A tentative diagnosis of ruptured retrocecal acute appendicitis and possible subdiaphragmatic abscess was made and operation was performed on March 8 at which time the appendix appeared normal. The left kidney was of normal size and mobile, but the right kidney was involved in a firm mass. The peritoneum was closed and the mass was explored retroperitoneally. The right kidney was firmly encased in a dense mass of fibrous tissue 1.2 cm. in thickness. Specimens on rapid frozen section were reported as "inflammatory fibrotic membrane." The kidney itself appeared normal, and no abscesses were seen. The biopsies were reported as showing chronic inflammation with productive fibrosis of the perirenal fat consistent with idiopathic retroperitoneal fibrosis (Fig. 6).

Postoperative Course: The patient improved rapidly under treatment with penicillin and chloramphenicol. The temperature was 38.5° C. on the first postoperative day and it remained normal thereafter. The patient had an uneventful postoperative course.

A repeat intravenous urogram on March 16, 1960 revealed a normal upper urinary tract
Cystoscopy performed two weeks later was essentially normal. Urine from the right and left ureter cultured pseudomonas aurigenosa. Repeat cultures on April 22, 1960 were positive for paracolon bacillus.

Discussion
This disease has been termed perirenal fascitis, Gerota's fascitis, sclerosing retroperitonitis, sclerosing lipogranuloma, non-specific retroperitoneal inflammation, and idiopathic retroperitoneal fibrosis. Periureteritis plastica, periureteral fascitis, and chronic periureteral fascitis are probably closely related conditions. Idiopathic retroperitoneal fibrosis is the preferred term at present because the exact pathogenesis has not been clarified and structures other than the ureter or kidney may be involved.

Excellent reviews of this subject have appeared recently by Hawk and Hazard (1959) and also by Hutch, Atkinson and Loquvam (1959).\(^{11,18}\) Hutch et al. reviewed the 21 reported cases of perirenal fascitis and added two cases of their own. Hawk and Hazard have grouped perirenal and periureteral fascitis together; they found 45 cases in the English literature and added another five. Additional cases have recently been reported.\(^{11,6,10,14,15,22,25,28}\) Hutch found anuria to be the cardinal symptom of this disease in its chronic form. In eight of the 23 cases complete anuria developed and in two others oliguria developed. They emphasized that the time interval between the first vague symptom and the development of anuria varied from one month to 11 years. The majority of the patients became anuric from one to five months after the onset. The preanuric symptoms were dependent upon the structures in retroperitoneal space which were involved by the fibrotic inflammatory process or plaque. Compression of spermatic vessels may lead to testicular pain; the inflammatory process may produce low back pain; involvement of the aorta may cause claudication; and compression of the inferior vena cava may lead to lower extremity edema. Eventually compression of the ureter leads to oliguria and anuria in approximately 40 per cent of cases.\(^{9,10,11,15,20}\)

Vest and Barelare (1953) attempted to differentiate periureteritis plastica from sclerosing retroperitonitis as described by Ormond.\(^{29}\) They reported four cases in which unilateral hydronephrosis developed; in three of the four cases ureteral catheters could be passed up to the renal pelvis on the involved side. At operation a marked inflammation of the tissue in the periureteral region was found. The indurated tissue varied from one to two cm. in thickness. Because the periureteral tissue reminded them of a linitis plastica of the stomach, they termed this disease "periureteritis plastica." Vest and Barelare postulated that the periureteral lymphatics might become infected and lead to a chronic inflammatory reaction, and that a better designation for this disease might be "chronic periureteral lymphangitis."

Hutch states that perirenal fascitis
should be differentiated from periureteritis plastica for the following reasons: perirenal fascitis is bilateral, usually involves the ureter above the pelvic brim, and frequently produces medial deviation of the involved ureters. In contrast periureteritis plastica is usually unilateral, usually involves the entire ureter, and the ureters remain in their normal position without medial deviation. Clinical differentiation of these entities probably is not important because both processes frequently cause ureteral obstruction, the histologic appearance is similar and, as will be emphasized, the principle of treatment by the conservation of renal tissue is common to both.

Pathogenesis: The pathogenesis of idiopathic retroperitoneal fibrosis is theoretical at present. The lymphatics from many areas drain into the para-aortic retroperitoneal space. Inflammation and neoplasms in the testicle, epididymis, vas deferans, ovary, ureter, kidney and gastro-intestinal tract could lead to involvement of para-aortic nodes. Partially controlled bacterial infection might result in a chronic fibrosing inflammatory process. Acute pyelonephritis with or without perinephritic abscess might lead to this process if the infection is not completely resolved. Ascending infection from the lower genitourinary tract might follow periureteral lymphatics from the bladder, prostate or cervix and gain access to the upper ureter and kidney. These lymphatic pathways have been demonstrated by Kenneth Walker (1913) and by Winsbury-White (1933). Chronic inflammatory diseases such as tuberculosis, syphilis, actinomycosis or other mycotic infections should be considered, but there is no real evidence indicating that these are etiologic factors. Lymphatic pathways from the lung to the paravertebral nodes below the diaphragm have been described as important in the pathogeneses of psoas abscess. These pathways might be important in the development of retroperitoneal fibrosis. Blood borne infections, collagen diseases, and nonsuppurative systemic panniculitis (variant of Weber-Christian disease) have all been suggested as possible causes of this condition. Trauma, through organization of a retroperitoneal hematoma, could conceivably be a cause of this disease.

Diagnosis: The exact diagnosis depends upon surgical exploration and biopsy of the retroperitoneal fibrous tissue. Middle-aged males predominate in the cases reported. The early symptoms in the chronic form are vague, usually consisting of anorexia, nausea and backache; later, as the disease progresses, the structures passing through the retroperitoneal space may become compressed by the inflammatory process. Idiopathic retroperitoneal fibrosis should be considered in all cases of bilateral ureteral obstruction if calculi can be ruled out. The utilization of intravenous and retrograde pyelography frequently shows the characteristic picture of dilatation of the renal pelvis and ureters down to the point of obstruction. The middle third of the ureter is most often involved and typically shows medial deviation. Hutch suggests that an upright view of the pyelogram may show failure of descent of the kidney due to fixation. Catheters can usually be passed beyond the obstruction; this may be followed by a profuse flow of urine, but withdrawal of the catheters usually results in recurrent anuria.

Differential Diagnosis: Many diseases which involve the retroperitoneal space enter into the differential diagnosis of retroperitoneal fibrosis. Pathologically a marked secondary fibrotic reaction occurs in tumors such as Hodgkin's disease, sarcomas, and certain carcinomas. Chronic inflammatory diseases including tuberculosis and actinomycosis should also be considered. Diseases in the genito-urinary tract to be considered include renal neoplasms, nonopaque ureteral calculi, primary ureteral carcinomas, and metastatic
ovarian and testicular neoplasms. Other neoplasms which could involve the retroperitoneal space may originate in the pancreas, gastro-intestinal tract, or arise primarily from this area.

Retroperitoneal fibrosis in an acute stage may suggest a ruptured viscus with involvement of the retroperitoneal space as illustrated by the cases in this report.

Treatment: Talbot and Mahoney have emphasized two main principles of therapy in the chronic stage: the relief of ureteral obstruction, if present, and the preservation of renal tissue. Operation during the acute stage is indicated primarily to clarify the diagnosis, as illustrated by these two cases.

In the cases reported thus far a variety of procedures have been performed: these include nephrectomy, nephrostomy, ureterolysis with transplantation, exploratory laparotomy with biopsy, and various combinations of the above. Nephrectomy should not be used in this disease as an initial procedure since the process is frequently bilateral. Unilateral or bilateral ureterolysis, with or without temporary nephrostomy as a preliminary or simultaneous procedure, gives the best results when ureteral obstruction is present.

Although ureterolysis appears to give excellent long-term results, a number of cases have been successfully treated by exploratory operation, biopsy, and the postoperative use of antibiotics. Adrenocorticosteroids have been used in a few cases with apparent clinical improvement. Certainly no extensive procedure should be undertaken when there is no evidence of ureteral obstruction, as in the two cases presented. These patients may be managed successfully with antibiotics alone and probably with the addition of adrenocorticosteroids. In addition, radiation therapy has been reported to be of value. Signs of ureteral obstruction should be searched for by serial pyelography. Exploration of the retroperitoneal space and ureterolysis should be reserved for cases showing progression of the retroperitoneal fibrotic process with signs of renal, ureteral, aortic, common iliac, or inferior vena caval compression.

Summary

Idiopathic retroperitoneal fibrosis is a disease of obscure etiology which is usually characterized by vague symptomatology until ureteral obstruction occurs as a result of a dense, fibrotic retroperitoneal process. Occasionally the great vessels in the retroperitoneal space also become compressed and this may lead to specific signs and symptoms.

Two cases are presented of this clinical entity which were seen at an early stage and which simulated acute surgical abdominal disease.

The recent literature is reviewed and current concepts in the pathogenesis and the surgical management of this disease are summarized.

Bibliography

9. Furlong, J. H., Jr. and H. V. Connerty: Compression of the Aorta and Ureters by a


