Progressive encaement and compression of the retroperitoneal structures by an extensive fibrosing process of uncertain etiology referred to as idiopathic fibrous retroperitonitis was first reported in the English literature in 1948. Since that time, more than 100 cases have been reported, and, in the majority, initial or subsequent symptoms were related to the urinary tract as a result of partial or complete ureteral obstruction. It is not uncommon for the aorto-iliac arteries to be embedded within the proliferating fibrous tissue, although ischemic symptoms have not been a conspicuous aspect of the disease.

Several case reports indicate that partial occlusion of the terminal aorta was an incidental finding at operation or necropsy and, in one case, from an aortogram made to assess the renal vasculature. The following case is presented to alert physicians to the possibility that idiopathic fibrous retroperitonitis can be manifested primarily as an aorto-iliac occlusive disease and to emphasize the relative ease and safety with which the obstruction can be relieved by the proper surgical measures.

Report of Case

A 42-year-old white man was examined at the Mayo Clinic on October 30, 1962, because of vague bilateral inguinal and testicular pain of 10 days' duration. Lifting, straining, and lying supine accentuated the pain, while sitting diminished it. Physical examination gave negative results; and a diagnosis of abdominal wall pain was made. The peripheral pulses were not examined. On December 10, 1962, the patient returned because of a severe cramping pain in his calves, thighs, and hips, which had become progressively worse over the past three weeks. At the time of his return walking no more than 30 feet would precipitate it. The testicular pain had persisted, but there was no impotence, paresthesia, or change in function of bladder or bowel.

The patient was well developed and well nourished. The pulse rate was 78 beats per minute, and the blood pressure was 126 mm. of mercury systolic and 64 mm. diastolic. Examination of the thorax gave negative results. A loud systolic bruit could be heard widely over the abdominal aorta and the iliac arteries. The abdominal aorta was not enlarged to palpation and pulsated normally. The femoral, popliteal, and pedal pulses were markedly diminished bilaterally, but there was little or no ischemia of the skin as determined by the elevation-dependency tests.

The urine was normal with a specific gravity of 1.012. Tests of blood disclosed that the value for hemoglobin was 13.4 Gm. per 100 ml. of blood and the leukocyte count was 14,600 per mm. of blood with a normal differential count. The erythrocyte sedimentation rate was 17 mm. in 1 hour (Westergren method); the concentration of fasting blood sugar (glucose) was 70 mg. per 100 ml. and the blood urea 24 mg. per 100 ml. The prothrombin time (Quick) was 19 seconds. Values for serum protein electrophoretic fractions, and for plasma cholesterol, phospholipids, fatty acids, and total lipids were within normal limits. The VDRL test was nonreactive. Roentgenograms of the thorax showed thickened pleura at the costophrenic angles, which had not been present on a previous roentgenogram in 1958, and roentgenograms of the lumbar and pelvic regions showed no vascular calcification.

A diagnosis of aorto-iliac occlusive disease was made. On December 18, 1962, a percutaneous translumbar aortogram demonstrated that the proximal portion of the abdominal aorta and the renal arteries were normal, but there was diffuse narrowing of the distal segment of the aorta with segmental stenosis of both common iliac arteries (fig. 1). The external iliac and femoral arteries appeared essentially normal bilaterally. On the basis of these data, a diagnosis of aorto-
iliac obstruction by atheromatous plaques was made, and surgical correction of the lesion was deemed feasible.

At operation on December 27, 1962, a grayish-white, woody, hard, fibrous plaque measuring 1.5 cm. in thickness was found which extended from below the left renal vein to the bifurcation of the common iliac arteries where it faded out over the pelvic brim. The process was most prominent near the midline, and its lateral boundaries were indistinct. Aortic pulsations disappeared where the distal segment of the aorta became engulfed by the fibrous mass, and pulses were not palpable in the external iliac arteries. The left ureter was markedly dilated proximal to its entrance into the fibrous mass where it crossed the left common iliac artery. At this point the ureter became bound into the fibrous mass and was severely constricted. The right ureter was not dilated and was drawn only superficially into the mass at the pelvic brim. The inferior vena cava was free from involvement.

The left and the right ureters were dissected free from fibrous mass and displayed laterally. A cleavage plane was developed between the overlying fibrous plaque and the outer aspect of the adventitia of the aorta and the common iliac arteries. With this plane as a guide, the dense scar tissue was excised anterior and lateral to these vessels. This allowed re-expansion and return of pulsatile flow through these vessels. Findings on microscopic examination of frozen sections were compatible with the diagnosis of idiopathic fibrous retroperitonitis (fig. 2). Fat tissue from the sigmoid mesentry was sutured over the raw surfaces in an attempt to cover the iliac arteries and ureters with normal tissue.

The patient made an uneventful postoperative recovery. The peripheral pulses were normal, no bruits were heard, and complete relief from claudication and inguinal and testicular pain was achieved. Results of repeated urinalyses and creatinine and urea determinations on the blood were within normal limits. Excretory urograms made on the tenth postoperative day and repeated 4 weeks later revealed medial deviation of both ureters and left hydronephrosis and hydro-ureter (fig. 3). Excretion of dye was prompt. The patient has returned to full-time employment.

Comment

This case illustrates that idiopathic fibrous retroperitonitis must be considered as a possible cause of aorto-iliac occlusive disease. In contrast to previously reported cases, there
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were no preceding or concomitant complaints or abnormalities related to obstruction of the urinary tract in this case. The testicular and inguinal pain observed early in this and other cases of retroperitoneal fibrosis is probably related to compression of the spermatic vessels. The relative ease with which a cleavage plane was developed that allowed safe excision of the fibrous plaque overlying the terminal aorta and its branches is worthy of emphasis.

The postoperative treatment of choice for prevention of recurrent ureteral and vascular obstruction has not been clearly delineated. Ionizing irradiation has been successful in certain cases but in at least one case, ureteral obstruction recurred 2 years later. Adrenal corticosteroids also have been employed with good results but as with roentgen therapy, not enough patients have been treated to allow definite conclusions. Adrenal corticosteroids were not administered to the present patient because a history of chronic duodenal ulcer was a relative contraindication to their use. Furthermore it was decided that, since the patient lived nearby and his progress could be closely followed by periodic physical examinations and evaluation of urinary function and excretory urograms, additional measures could be instituted if they should become necessary. This case, therefore, offers an opportunity to follow the course after surgical treatment alone. A general clinical impression exists that, like idiopathic fibrous mediastinitis, this disease undergoes prolonged periods of spontaneous remission.

Summary

In the case of idiopathic fibrous retroperitonitis presented, the only symptoms were those of arterial insufficiency of the lower limbs and testicular and inguinal pain of uncertain origin. Relief of claudication and return of normal pulsations were achieved after surgical lysis of the common iliac arteries and the terminal segment of the aorta. Numerous reports in the literature indicate that idiopathic fibrous retroperitonitis is not rare but retroperitoneal arterial obstruction by the fibrous plaque is apparently unusual, and intermittent claudication without urinary symptoms is unique.

Idiopathic fibrous retroperitonitis, therefore, should be included in the differential diagnosis of the causes of intermittent claudication. Although the aortogram may not permit differentiation of arteriosclerosis obliterans from external compression of the aorta by this fibrosing process, the excretory urogram should be helpful in making or excluding the diagnosis. Treatment is primarily surgical with thorough excision of the fibrous mass and with ureteral transplantation; vascular resection or bypass grafting is not necessary. Advisability of postoperative roentgen and adrenal corticoid therapy is not clearly established.
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References


Coarctation of the Aorta

This striking condition of narrowing or even obliteration of the aorta at or immediately below its isthmus, namely the portion between the orifice of the left subclavian artery and the insertion of the ductus arteriosus, was first described in 1789 by Paris, who came upon it in the course of injecting a body for dissection; Laennec had seen three or four cases and referred to four recorded cases of obliteration, including those of Graham, John Bell, and Ashley Cooper. An early case was in a man aged ninety-two years reported by Reynaud in 1828. Peacock collected forty-six cases in 1866, and in 1903 a total of 160 was analysed by Bonnet, who divided them into (i) the infantile or developmental with usually a diffuse narrowing at the isthmus, and (ii) the adult with a sudden constriction at the site of the ductus due to post-natal cicatricial contraction.—Sir Humphry Davy Rolleston. The Harveian Oration, Great Britain, Cambridge University Press, 1928, p. 48.
Intermittent Claudication Secondary to Idiopathic Fibrous Retroperitonitis:
Report of an Unusual Case
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