Pelvic lipomatosis

Case report

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Since Engels\textsuperscript{1} first reported five cases of pelvic lipomatosis at least 25 cases have been reported. Pelvic lipomatosis, a term first coined by Fogg and Smyth\textsuperscript{2} in 1968, has also been reported under other titles. Essentially, it is a benign condition in which there is an abundance of fatty tissue in the perirectal and perivesical spaces in the pelvis (Fig. 1). It is important to differentiate pelvic lipomatosis from retroperitoneal fibrosis and pelvic tumors. The clinical course, treatment, and prognosis differ dramatically. In several cases, including this case, the patients had exploratory surgery. However, the disease can be diagnosed from various clinical symptoms, physical findings, and a plain abdominal film. Roentgenographic studies of the urinary tract and the colon will confirm the diagnosis.
Fig. 1. There is increased radiolucency in the pelvis best appreciated in contrast to the radiopaque fluid filling the bladder but present on the preliminary abdominal film (not included here). The increased radiolucency demonstrates the deposition of fat in the perivesical area.

Case report

A 62-year-old man had been treated for various medical problems for approximately 18 years. At this examination the patient complained of chronic cough, dysuria, frequency, and urgency. He also had a history of intermittent hypertension. A urogram showed the distal thirds of both ureters to be displaced medially, and the urinary bladder to be located more cephalad than expected. The bladder had a peculiar appearance with elongation in its vertical dimension (Fig. 2).

The patient's blood pressure at the time of admission was labile and ranged as high as 240/150. An investigation into possible organic causes of hypertension was initiated. Results of tests including a renal arteriogram were normal. Examination of the colon showed tubular narrowing and elevation of the sigmoid out of the pelvis in the midline (Fig. 3). No evidence of obstruction or mucosal alteration was noted. Cystoscopy revealed a mild urethral stric-

ture and deformity of the bladder from extrinsic compression. At laparotomy a large, rubbery mass approximately 9 cm in diameter completely engulfed the prostate gland and the posterolateral walls of the bladder and the rectum. Benign smooth muscle, fibrovascular, and adipose tissue were found in the biopsy specimen, but there was no evidence of neoplasm. The diagnosis of pelvic fibrolipomatosis was made.

Clinical findings

We reviewed 29 reported cases, 27 men and 2 women, ranging in age from 25 to 83 years. Clinical correlations varied, with symptoms of lower urinary tract obstruction (10) and hypertension (7) being the most common. Other symptoms included vague complaints of constipation and nocturia. In most instances the diagnosis was first considered only after roentgenograms had

Fig. 2. The sigmoid colon is straight and elevated from the pelvis, but there is no evidence of mucosal abnormality or obstruction.
been obtained for evaluation of hypertension or nonspecific genitourinary or gastrointestinal symptoms.

Physical findings were not consistent, but an ill-defined mass in the suprapubic region was described most frequently. Prostatic enlargement was mentioned by only three authors.\textsuperscript{1, 3, 4} There was more concern that the prostate was elevated and therefore difficult to palpate.\textsuperscript{2, 5, 6} Elongation of the membranous and bulbous portions of the urethra was noted.\textsuperscript{2, 3, 7}

Several authors mentioned the difficulty in performing cystoscopy because of the elongated bladder or the distorted trigone or both.\textsuperscript{1, 2, 4-6, 8} At cystoscopy, findings ranged from normal\textsuperscript{9} to marked bullous edema.\textsuperscript{8} Biopsy specimens of the bladder obtained in at least five patients showed chronic cystitis, cystitis cystica,\textsuperscript{1, 5} subacute cystitis,\textsuperscript{2} and cystic glandulitis.\textsuperscript{4, 8}

Most authors reported that examinations of the colon showed tubular narrowing and vertical straightening with upper displacement of the rectum and sigmoid colon. Deviation of the sigmoid colon to the right\textsuperscript{4, 10} and to the left\textsuperscript{9} was described infrequently. Sigmoidoscopy, when performed, revealed no intrinsic lesion, but merely a straightening of that part of the colon.

Plain films of the abdomen demonstrated increased radiolucency in the pelvis;\textsuperscript{2, 4-6, 11} the lucency indicated increased fatty tissue.

The bladder has been described by several investigators as “gourd-like,”\textsuperscript{1, 7, 8} “banana shaped”\textsuperscript{4} or “pear shaped.”\textsuperscript{5, 7, 11} These terms describe the vertical elongation of the bladder which is the result of the fatty infiltration in the pelvis. Eight of 12 authors mentioned increased elevation of the bladder, usually in an anterior direction.\textsuperscript{2-7, 11} Hydronephrosis and hydroureter were found only when the fatty tumor impinged upon the pelvic structures to cause obstruction.\textsuperscript{2-8, 11} Four authors reported lateral deviation of the ureter involving the mid-third in two patients\textsuperscript{3, 11} the distal third in one patient,\textsuperscript{4} and marked distal ureteral deviation in one.\textsuperscript{10}

Surgery was performed on at least 20 patients and biopsy specimens were obtained. The typical operative note described a mass of well vascularized fatty tissue in the pelvis with a minimal fibrous component. One patient had fatty deposits localized in the retroperitoneal spaces and the hollow of the sacrum, with no involvement of the bladder, ureter, or rectum.\textsuperscript{12} Other patients had intimate adhesions of the
fatty tissue to the distal ureters, bladder, and rectum so that severe hydroureret and hydronephrosis resulted, necessitating supravesical diversion.2, 6

In two cases reported by Lucey and Smith the patients died of uremia.5 One patient treated with radiation and surgical diversion died 10 years after the initial diagnosis. It was suggested that the diversion procedure was necessary because of fibrosis secondary to radiation therapy. Another patient who did not undergo exploratory laparotomy had two transurethral resections and died of uremia. Lucey and Smith suggested that the patient had a congenital ureterovesical obstruction, since the urogram showed only terminal ureterectasis. It is suggested that the ureterovesical obstruction and not the pelvic lipomatosis was the chief factor contributing to the patient’s death from uremia. No neoplastic tissues were found in biopsy specimens. The typical pathological finding was a group of well-vascularized fat cells with an admixture of fibrous tissue.

Discussion

The cause of this disorder is not known. Although four patients in the series reported by Engels underwent laparotomy and had large amounts of fatty tissue in the pelvis, they also had extensive pelvic adhesions, and he concluded that the latter was the underlying cause of the distortion of the bladder and sigmoid colon.1

Rosenberg et al9 reported the case of a 50-year-old man with sciatica whom they diagnosed as having Dercum’s disease. The patient had painful tender masses on the arms, legs, and right side of the rectum. Barium enema and urographic studies were typical of pelvic lipomatosis, and for this reason the case is included in this review, although the painful nature of the fatty masses is unique.

Fogg and Smyth5 suggest that pelvic lipomatosis is comparable to Weber-Christian disease and sclerosing lipogranulomatosis. Weber-Christian disease is a generalized nonsuppurative panniculitis of the subcutaneous tissues of unknown etiology and sclerosing lipogranulomatosis is a chronic inflammatory reaction to necrotic fatty tissue.

Harrow7 reported two cases he diagnosed as retroperitoneal fibrosis secondary to injection of sclerosing agent. However, the roentgenograms and pathologic findings indicated pelvic lipomatosis.

Lucey and Smith5 investigated one of their cases to determine if various fatty depositions in different anatomic sites had different characteristics, but no pathologic or chemical variations were detected.

Malter and Ornell10 suggested that the disease was a localized manifestation of obesity. However, they were unable to substantiate this theory.

The case reported by Bender and Kass12 was somewhat different in that there were no roentgenographic findings, and the biopsy specimen showed well-encapsulated fat, i.e., lipoma. It is believed that this was an early variant of pelvic lipomatosis occurring before the fat cells extended through the capsule to fill the pelvic space.

Morettin and Wilson11 suggest that fatty deposition is hormonally related, but they offer no convincing arguments.

Differential diagnosis most commonly includes: (1) retroperitoneal
fibrosis, (2) metastatic tumor, (3) pelvic retroperitoneal hemorrhage.

Retroperitoneal fibrosis can easily be differentiated pathologically because of the prominent fibrous element and lack of fatty tissues. It is usually located above the pelvic brim; pelvic lipomatosis lies within the pelvis and distorts the bladder and sigmoid colon. An additional aid is the medical history which frequently includes a history of methysergide maleate (Sansert). At surgery, the predominance of fibrous tissue confirms the diagnosis.

Metastatic pelvic tumors frequently cause lobular extrinsic compression of the bladder and the rectum and usually do not elevate or elongate these organs. Pelvic hematomas are suspected by the clinical history and asymmetric displacement of the rectum and bladder. Most helpful in these cases is the preliminary abdominal film which shows no radiolucency (indicating fatty deposition) in the soft tissues of the pelvis.

Summary

One case of pelvic lipomatosis is reported. Clinical features are discussed. The diagnosis should be suspected in a man when a preliminary abdominal roentgenogram shows radiolucency in the pelvis, the urogram reveals a distorted, elongated urinary bladder, and examination of the colon shows narrowing of the rectosigmoid with elevation out of the pelvis. Cystoscopy is difficult to perform because of the elongated urethra and a deformed trigone. The pathologic findings are well vascularized fatty tissue with a minimal fibrous component.

References