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Pelvic Lipomatosis: A Rare Case with a Good Surgical Outcome

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ABSTRACT

Pelvic lipomatosis is defined as a condition characterized by the diffuse proliferation of normal mature fatty tissue in the perirectal and perivesical region. We report a case of a 40-year-old, obese male who presented with marked lower urinary tract symptoms. Imaging studies and blood tests revealed bilateral hydroureteronephrosis and right non-excretory kidney with deranged renal function. Diagnosis of the pelvic lipomatosis was established on the basis of clinical and radiological findings. The patient was successfully managed by simple cystectomy and ileal conduit. After 6 years of follow-up, the patient is doing well with normal renal parameters.

CASE REPORT

A 40-year-old, obese, normotensive, euglycemic patient presented in 2005 with increased frequency, urgency, urge incontinence, nocturnal enuresis, straining to void, and poor stream for 15 years. Symptoms had aggravated over the last year (2004). There was a history of low backache and erectile dysfunction for the last 6 months. His bowel function was normal and there was no edema of the lower limbs.

He had a history of circumcision and urethral dilatation in 2002. The physical examination showed that the patient was of average build, had a protuberant abdomen, and he was circumcised, with xerotic glans but an adequate meatus. The digital rectal examination revealed a flat prostate with nodules over the right lobe.

His serum creatinine level was 2 mg/dl. An ultrasound revealed bilateral gross hydronephrosis, hydroureter, and a thick-walled bladder. An intravenous pyelogram (IVP) done back in 1994 demonstrated a B/L excretory kidney with a high pear-shaped bladder, with translucency in the pelvis (Figure 1). An IVP in 2005 showed right non-excretory and left excretory kidneys with marked hydronephrosis and hydroureter up to the lower end (Figure 2). A retrograde urethrogram showed an elongated prostatic urethra (Figure 3), and a micturating cystourethrogram revealed a pear-shaped bladder (Figure 4). Uroflowmetry reported a Qmax of 8 ml/sec for a voided volume

KEYWORDS: Pelvic lipomatosis, pear shaped bladder, simple cystectomy, cystitis glandularis

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Figure 1. IVP in 1994 demonstrated B/L excretory kidney with high pear-shaped bladder with translucency in pelvis.

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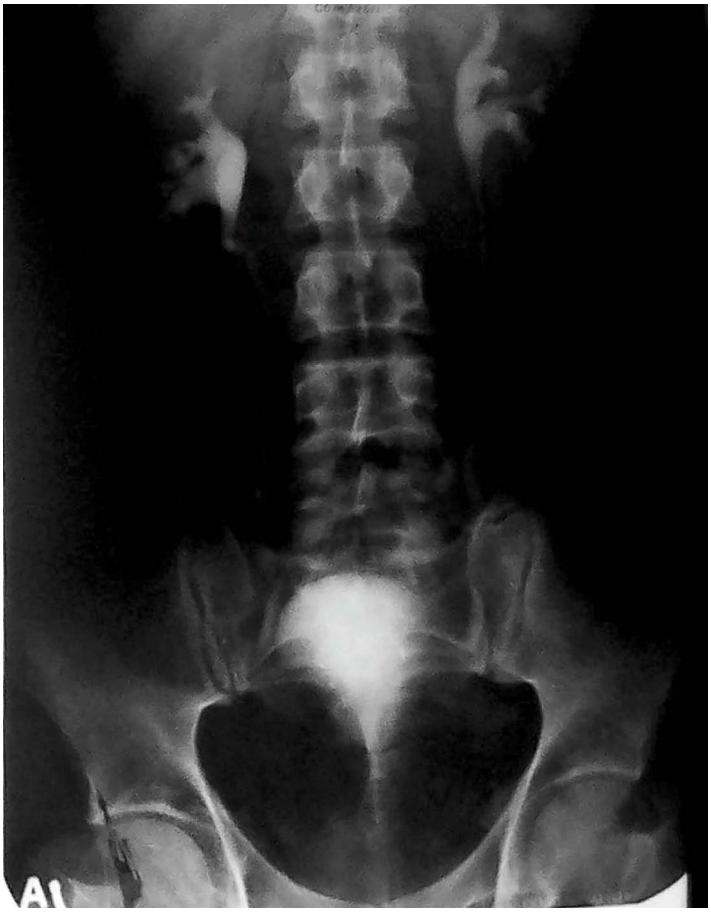

Figure 2. IVP in 2005 showed right non-excretory and left excretory kidney with marked hydronephrosis and hydroureter up to lower end.

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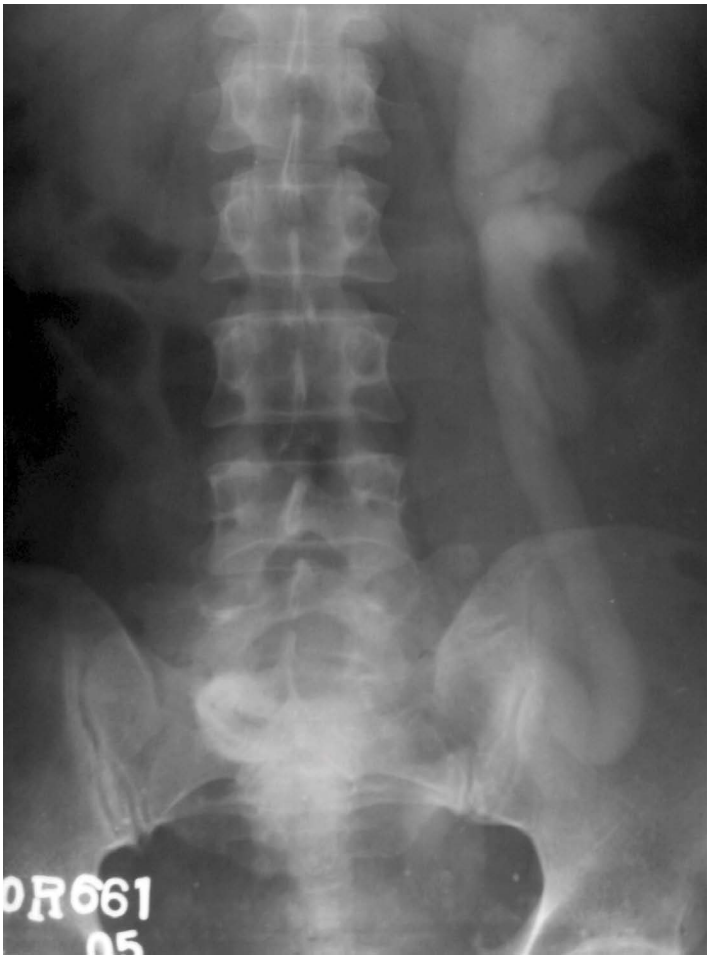

of 170 ml. A urodynamic study demonstrated a small capacity, poorly compliant bladder. A cystoscopy revealed an elongated prostatic urethra, angulated bladder neck, a non-obstructive prostate, and normal bladder mucosa with a small capacity. A non-contrast CT scan demonstrated marked fat densities in the pelvis, encasing the bladder and rectum with a thick wall, resulting in a bladder with a small capacity (Figure 5). An MRI on the T-1 weighted image demonstrated abundant perivesical and perirectal hyperintense signals, and an elevated, distorted bladder base and a compressed rectum (Figure 6). On the basis of clinical and radiological findings, a diagnosis of pelvic lipomatosis was made. In the meantime, the patient's renal function deteriorated and his serum creatinine rose to 3 mg/dl.

Bilateral percutaneous nephrostomy was done, and when his nadir serum creatinine value reached and remained static at 1.3 mg/dl, a simple cystectomy and ileal conduit was done. There were dense fatty masses adhering all around the bladder, and his ureters were encased in fatty tissue. No attempt was made to release the ureters. Some of the fatty mass was excised for a biopsy, which revealed a benign lipomatosis (mature fat cells with chronic non-specific inflammation). A histopathology of the cystectomy specimen revealed proliferative cystitis in the bladder mucosa, with cystitis glandularis and urothelial atypia near the left ureteric orifice.

Figure 3. A retrograde urethrogram showing an elongated prostatic urethra.
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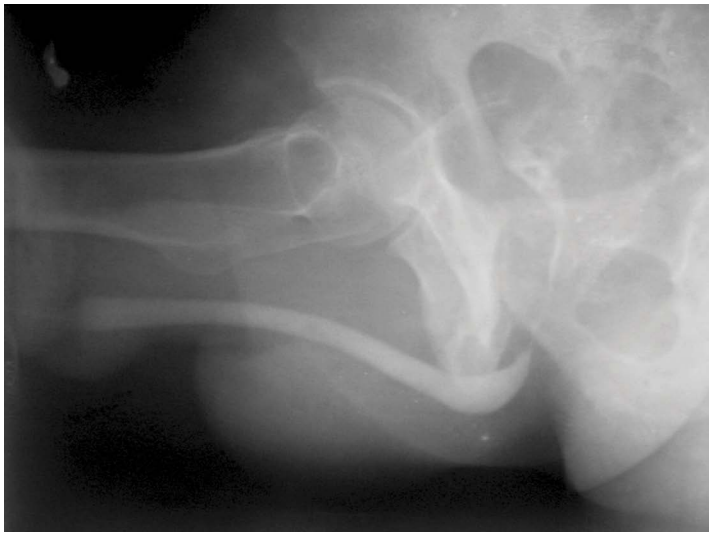

Figure 4. A micturating cystourethrogram revealed a pear-shaped bladder.
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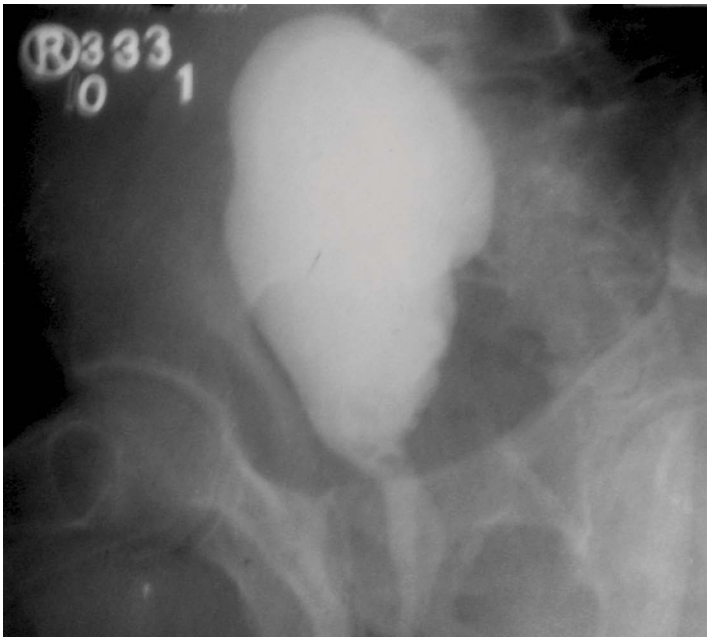

The patient had uneventful recovery. He is asymptomatic after 6 years of follow-up, with a normal level of serum creatinine (1 mg/dl). His erectile dysfunction did not improve after surgery but he is performing well on 5-phosphodiestersae inhibitors.

DISCUSSION

Pelvic lipomatosis is a rare condition characterized by diffuse exuberant pelvic overgrowth of non-malignant but infiltrative adipose tissue in the perivesical and perirectal space. It was first reported by Engels in 1959 in 5 patients [1]. Fogg and Smyth first coined the term pelvic lipomatosis in 1968 [2]. Pelvic lipomatosis had been reported in association with retroperitoneal lipomatosis [24]. The mean age of presentation in the largest review (130 patients) was 48 years [3]. Carpenter described 2 patients. The first is a young stocky male who is more susceptible to progressive ureteral obstruction [4]. The second is an older male with an indolent disease course.

The incidence of pelvic lipomatosis is 0.6 to 1.7 per 100 000 hospital admissions [3]. There is a strong male predominance, with a male-to-female ratio of 1.8:1. There is also a racial disparity: 67% of African Americans have pelvic lipomatosis compared to 33% of Caucasion patients with the disease. [3]. A possible genetic etiology is suggested by abnormality in the chromatin-regulating HMGA proteins [5]. HMGA2 overexpression and HMGA1 under expression are associated with increased fatty tissue deposits in murine models [6-8]. The etiology of this disorder is unknown, but it is suggested that obesity plays a role, and it has been found in half of the patients afflicted with obesity [4,9]. Approximately half of the patients present with lower urinary tract symptoms, and about a quarter present with bowel complaints. Suprapubic pain or lumps, backache, flank pain, or perineal discomfort may be other clinical manifestations [10]. Hypertension is common, occurring in 35 to 74% of patients [3]. Hydronephrosis develops in at least 1/3 of the patients evaluated. It often presents with bilateral hydronephrosis in 19% of patients. The cause of hydronephrosis is distal encasement of the ureter by lipomatosis lesions. Renal failure occurs in 6% of all reported cases. This small incidence of renal failure is attributed to a gradual, progressive course of pelvic lipomatosis, and a high grade of obstruction [11]. A life threatening complication of pelvic lipomatosis is thromboembolism expressed by either deep venous thrombosis or pulmonary embolism (7%). Venous stasis is due to the compression of iliac veins or vena cava [11-13]. Physical findings include suprapubic masses, a high-riding prostate, and an indistinct pelvic mass [10].

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Figure 5. A non-contrast CT scan demonstrated marked fat densities in the pelvis, encasing the bladder and rectum within a thick wall.

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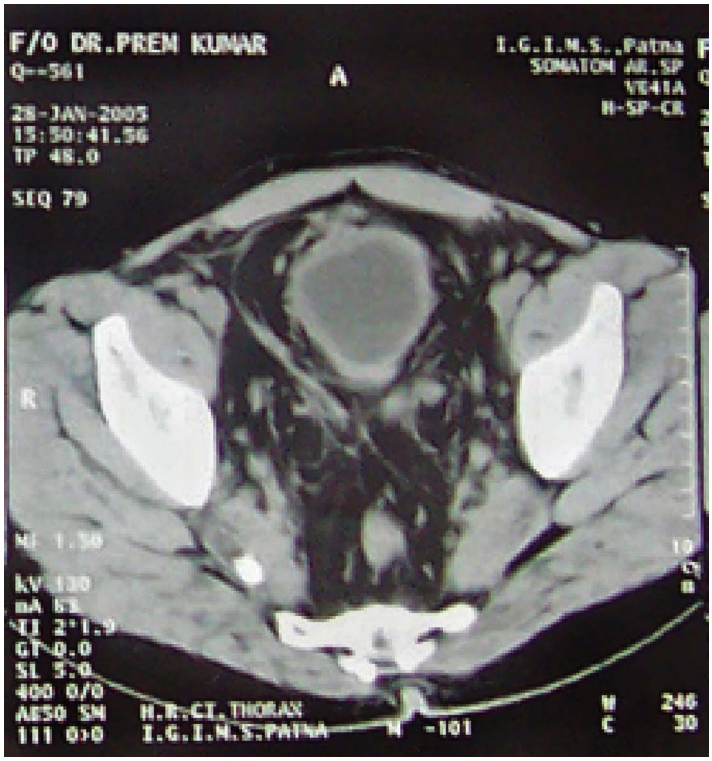

Figure 6. An MRI on a T-1 weighted image showed abundant perivesical and perirectal hyperintense signals, with an elevated, distorted bladder base and compressed rectum.

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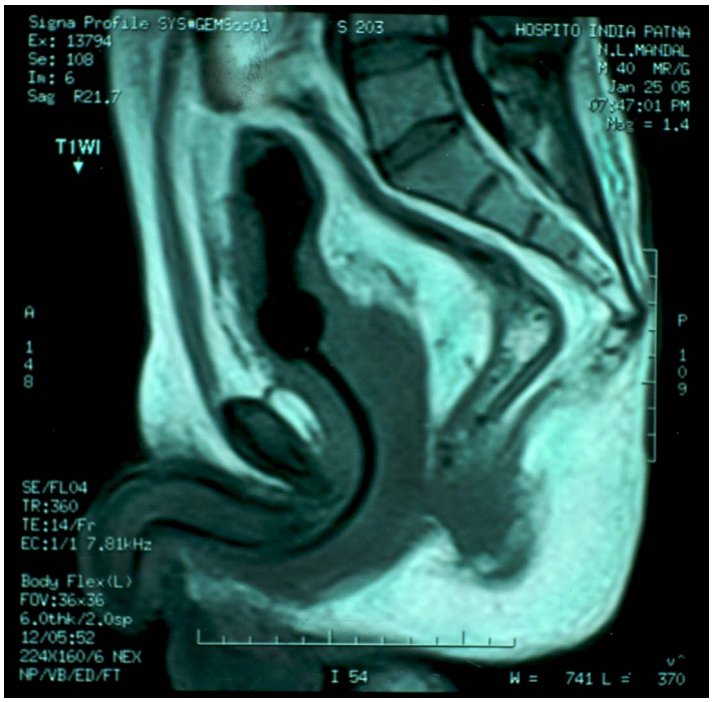

The diagnosis is based on a large index of suspicious clinical and radiological findings. The first radiological indicator is marked translucency in the perivesical area on plain X-ray (KUB) film. On excretory urography, the bladder characteristically assumes a pear or gourd shape, with an elevated bladder base and a medial or lateral deviation of the ureters due to symmetric compression [2,14,15]. Hydronephrosis is also seen [3]. On pelvic and abdominal CT, the bladder and rectosigmoid are surrounded and displaced by homogenous tissue with low attenuation (-40 to -100 HU), signifying fat content. The CT establishes a diagnosis of pelvic lipomatosis [16,17]. An MRI can also be used for diagnosis, as it permits the characterization of fat planes, and it provides the delineation of an elevated bladder base and the elongation of a posterior urethra [18]. The fat may extend laterally within the pelvis to involve the blood vessels, as well. The stretching and thinning of the femoral veins are easily discernable with venography. However, because contemporary imaging methods suffice well enough to characterize any vascular sequelae from lipomatosis, venography is typically no longer employed [22].

Evaluation by cystoscopy and the biopsy of any suspicious lesion should be taken because of an association with adenocarcinoma of the bladder [19]. Cystoscopy detects abnormalities in 75% of patients and most often includes cystitis glandularis and cystitis cystica [19].

Pelvic lipomatosis can be managed conservatively, unless obstruction of the urinary tract, cystitis glandularis, or adenocarcinoma occurs. Steroids, antibiotics, and radiation therapy have been tried without any success [20]. There is controversy regarding excision of lipomatous tissue because of the obliteration of normal anatomic planes, and the increased vascularity within fatty masses and the intimate association of vascular structures. Ultrasonic-assisted lipectomy has been

suggested but long-term follow-up data are lacking [21]. Resulting ureteral obstruction with deteriorating renal function can be managed with nephrostomy, ureteric re-implantation, or simple cystectomy and ileal conduit. Pelvic lipomatosis may remain dormant for many years. Radiological evaluation and serum creatinine estimation should be regularly performed. Adenocarcinoma of the bladder has been reported in the second transurethral resection of the bladder, occurring 1 month after the first cystitis glandularis operation associated with pelvic lipomatosis. Close follow-up has been suggested [23]. There is no specific recommendation, but it would be reasonable that patients undergo annual or biannual cystoscopies unless storage symptoms or hematuria develop, leading to more frequent examinations.

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