PELVIC AND RETROPERITONEAL LIPOMATOSIS: CASE REPORT

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SUMMARY – A rare case of pelvic and retroperitoneal lipomatosis lasting for more then three decades is presented. Although the disease is benign, it is usually progressive and may lead to renal insufficiency. On the other hand, treatment options are limited and with questionable effects. The clinical features, intravenous urography, computed tomography and magnetic resonance images, differential diagnosis and therapeutic aspects of this rare disease are presented.

Key words: *Lipomatosis – complications; Lipomatosis – pathology; Lipomatosis – radiography; Urography; Pelvis; Case report*

Introduction

Pelvic lipomatosis is a rear disease, but urologists should be aware of it and together with radiologists should assume primary responsibility in suggesting and then establishing the diagnosis of pelvic lipomatosis.

Case Report

A 73-year-old man presented with complete urinary retention lasting for one day. Urinary catheter was placed and approximately one liter of urine was collected. He was known for having 'kidney problems' from 1973, but was unsure of what kind. He came without any medical documentation. He had no fever, blood pressure was normal, serum creatinine level was 178 μmol/L (normal range, 79-125 μmol/L), urea was 10.3 mmol/L (normal range, 2.8-8.3 mmol/L) and C reactive protein was 140.4 mg/L (normal range <5.0

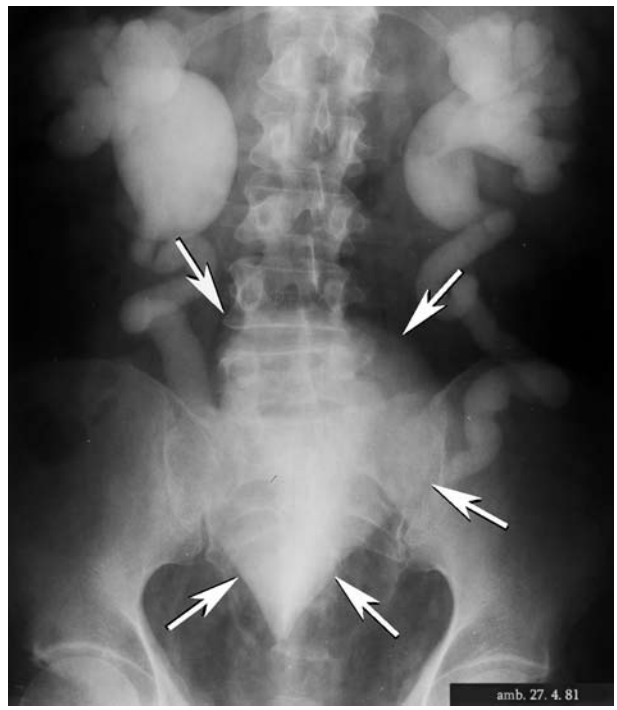

Fig. 1. Intravenous urography taken in1981 shows an increased pelvic radiolucency, bilateral dilatation of the renal collecting system, tortuosity of dilated ureters, and an inverted tear-drop configuration of the superiorly displaced urinary bladder (arrows).

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Received April 16, 2010, accepted in revised form September 8, 2010

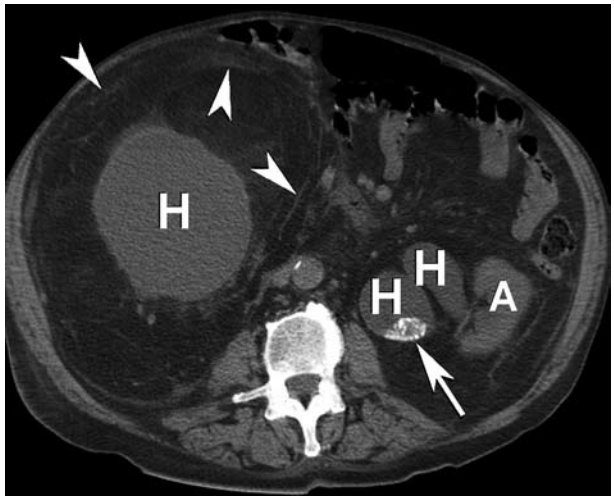

Fig. 2. Unenhanced transverse computed tomography scan obtained through renal pelvis shows extreme bilateral hydronephrosis (H) with renal parenchymal atrophy (A) and left-sided calculi (arrow). Excessive fat accumulation in perinephric spaces, more pronounced on the right side, with streaky areas of increased density (arrowheads).

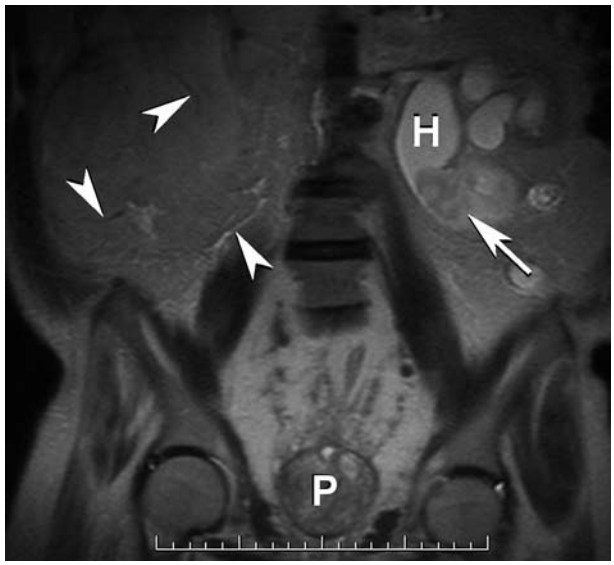

Fig. 3. Coronal HASTE respiratory gated magnetic resonance image (repetition time 1040 ms, echo time 80 ms, flip angel 1500, one signal acquired, section thickness 8 mm, field of view 400 mm, matrix size 192x256) at the level of anterior aspect of the lumbar spine demonstrates hydronephrosis (H) with renal parenchyma atrophy and calculi in the left kidney (arrow). Enlargement of the prostate (P) is shown (volume of 110 ccm). Abundant tissue with signal intensity similar to that of subcutaneous fat, with linear foci of fibrosis is seen in retroperitoneum (arrowheads).

mg/L). Urine analysis showed an increased number of red blood cells and leukocytes. On digital rectal examination, the prostate could not be palpated. Later on the same day, the patient's family brought intravenous urography findings, taken in 1981 (Fig. 1) and the patient was referred for further workup. Computed tomography (CT) (Fig. 2) and magnetic resonance (MR) (Fig. 3) of the abdomen and pelvis were performed.

Discussion

Pelvic lipomatosis is a rare, benign disease that causes ureter, bladder, sigmoid colon and rectum deformities secondary to an increased deposition of fat in the pelvis. The disease, first described by Engels in 1959, is characterized by overgrowth of nonencapsulated, nonmalignant but infiltrative adipose tissue[1]. Histologic examination reveals normal, mature adipose tissue. Occasionally, mild fibrosis and inflammatory cells can be noted. Pelvic lipomatosis is usually symmetrically distributed and confined to the pelvis, however, retroperitoneal involvement may sometimes occur[2,3]. The etiology of pelvic lipomatosis is unknown; it can be related to obesity[4] but also to genetic causes[5]. Approximately 70% of the patients affected are male African-Americans, average age 48[6], however, pediatric patients aged only 9 have been described[7]. Pelvic lipomatosis is usually asymptomatic. Clinical symptoms that are associated with the advanced stage of the disease, as well as physical findings are usually nonspecific and unremarkable, and the disease is usually detected accidentally. The patient may present with urinary tract infection and obstruction, constipation, abdominal and pelvic pain. The possible late complications are obstructive renal failure, proliferative cystitis[8] and bladder adenocarcinoma[9]. Physical findings may show obesity, a suprapubic mass, and on digital rectal examination the prostate could be elevated or even unable to palpate due to the bladder base elevation, as in our patient. Cystoscopy may be difficult to perform to elongation of the prostatic urethra, elevation of the bladder, and pelvic fixation.

Changes in the pelvis and retroperitoneum can be seen by intravenous urography, or by CT and/or MR. All three methods can show superior displacement of the bladder base, 'specific' changes in bladder shape, dilatation and displacement of one or both ure-

ters, hydronephrosis and narrowing of rectosigmoid colon. Increased pelvic radiolucency can be seen on pelvic roentgenograms, which allows for visualization of the muscular planes and better definition of the bony detail of the sacrum[10]. Excretory urography demonstrates ureterohydronephrosis and deviation of ureters to the medial in 80% or to the lateral in 20% of patients[7]. Urinary bladder is extrinsically compressed and vertically elongated and elevated from the pelvic floor. Its shape is described as 'banana', 'pear-shaped', 'gourd-shaped', or as in this case 'inverted tear-drop' with its narrow end at the base (Fig. 1). The lack of prostatic indentation at the base of the bladder is an important differential point in the exclusion of prostatic enlargement as the cause of bladder elevation[7], although most patients already have prostatic enlargement that is probably due to their age.

The presence of fat is easily recognized owing to its low attenuation at CT or its high signal intensity at T1-weighted MR imaging with loss of signal intensity on fat-suppressed images[11]. The fat tissue in the pelvis and retroperitoneum is extensive, homogenous, nonencapsulated, and usually bilateral and symmetrically distributed.

Differential diagnosis for lesions mostly composed of the fat in the retroperitoneum and pelvis would include pelvic lipomatosis, lipoma and liposarcoma. Lipomas are benign tumors composed of mature fat cells and a small amount of connective tissue. On CT, lipomas have density equal to that of normal fat; they are homogeneous, well defined and consist almost entirely of fat, although within the fatty tissues they can have linear, streaky densities. They appear noninvasive, causing displacement of adjacent structures. Even tumor capsule can be seen on CT scan[12]. Lipoma cannot be distinguished radiographically from pelvic lipomatosis but they are usually unilateral, encapsulated, and less extensive comparing to pelvic lipomatosis, which is usually symmetrical and bilateral. Liposarcoma is a malignant tumor of mesenchymal origin and the most common type of primary retroperitoneal malignancy. There are histologically different types of liposarcoma and even in the same tumor there can

be a mixture of different histologic subtypes. Depending on their histology, they can have different proportions of malignant cells and connective tissue, which makes their CT and MR imaging appearance variable[12]. If they are well differentiated, they usually contain an appreciable amount of fat and on CT and on MR they resemble lipomas. This can be a diagnostic problem and additional examinations and clinical data may be necessary to exclude malignant disease. If they are high-grade liposarcoma, they do not have to demonstrate the appreciable amount of fat and may appear similar to other sarcomas[11]. For diagnosis and follow up of patients with lesions mostly composed of fat, history and clinical data are needed. Fatty masses that remain stable in size, together with long time history of the disease, can help in excluding malignant disease.

There is no definitive treatment for pelvic lipomatosis. Complete surgical eradication of the adherent fatty tissues is difficult and seldom possible with questionable clinical effects. Other methods of treatment such as weight loss, radiation therapy and chemotherapy were used in the past, but did not show significant results. Although the disease is progressive and can lead to obstructive renal failure, our patient shows that even when the disease is present for a long period of time (more than three decades), kidney function need not be significantly interrupted and watchful waiting can be the method of patient surveillance. After two weeks, the patient was discharged from the hospital in stable condition, with urinary catheter and normal levels of serum creatinine and urea.

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Sažetak

ZDJELIČNA I RETROPERITONEALNA LIPOMATOZA: PRIKAZ SLUČAJA

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Prikazan je rijedak slučaj zdjelične i retroperitonealne lipomatoze koji traje više od tri desetljeća. Iako je ova bolest u svojoj naravi dobroćudna ona uglavnom s vremenom napreduje te može dovesti do bubrežne insuficijencije, a mogućnosti liječenja su ograničene s upitnim rezultatima. Opisuju se kliničke značajke, slike intravenske urografije te kompjutorizirane tomografije i magnetske rezonance, kao i diferencijalna dijagnoza te terapijske mogućnosti liječenja zdjelične i retroperitonealne lipomatoze.

Ključne riječi: *Lipomatoza – komplikacije; Lipomatoza – patologija; Lipomatoza – radiografija; Urografija; Zdjelica; Prikaz slučaja*