

Inflammation and infection

Difficult cystoscopy due to “Elongated” urethra- think of pelvic lipomatosis

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ARTICLE INFO

Keywords:

Cystitis cystica
Pear-shaped bladder
Pelvic lipomatosis

ABSTRACT

Pelvic lipomatosis is a rare, proliferative disease with unknown etiology involving an overgrowth of normal fat in the pelvic retroperitoneal space.¹⁻⁵ Cystitis glandularis or cystitis cystica can be observed in 75% of patients with pelvic lipomatosis.⁵ We describe a 44-yr-old chronic smoker with suspicion of bladder mass who was referred to us following an inconclusive cystoscopy due to an “elongated” urethra and was later diagnosed to be a case of pelvic lipomatosis with cystitis cystica. Because the association of this condition with bladder adenocarcinoma and the possibility of obstructive uropathy later, we have kept the patient on close follow-up.

Introduction

Pelvic lipomatosis is a rare condition characterized by a non-malignant overgrowth of normal fat in the perirectal and perivesical spaces.¹ Though it's a recognized clinicopathological entity, many cases probably remain unrecognized or unreported because diagnosing such a rare entity needs a high index of suspicion. It has been demonstrated that approximately three-quarters of cases with pelvic lipomatosis have histologic evidence of proliferative cystitis, such as cystitis cystica and cystitis glandularis, although its cause remains unknown.¹ In the present report, we describe a case of pelvic lipomatosis associated with proliferative cystitis who was referred to us following an inconclusive cystoscopy due to an abnormally long urethra.

Case report

A 44-year-old chronic smoker with body mass index of 32.6, presented with occasional obstructive voiding symptoms for more than one year. He gave a history of having undergone a cystoscopy outside for evaluation of bladder mass and though the details weren't available he mentioned specifically that the surgeon had informed him that his urethra was abnormally “long” which made it difficult to evaluate his bladder. Physical examination, blood and urine workup were normal. Ultrasonography shows bilateral minimal calyectasis with ill-defined echogenic lesions in posterior bladder wall. Uroflowmetry showed a bell-shaped curve with Q_{max} of 10 ml/sec. Contrast enhanced computed tomography demonstrated large amounts of fatty tissue occupying the pelvis from the bottom of the pelvic cavity, causing extrinsic

compression and elevation of the bladder neck, besides enhancing lesions in the posterior bladder wall (Fig. 1). A micturating cystourethrogram revealed a pear-shaped bladder with elevation of bladder neck typical of pelvic lipomatosis (Fig. 2). Cystoscopy showed elongated prostatic urethra with multiple cystic lesions in prostatic urethra and over bladder neck which were resected and histopathologically confirmed as cystitis cystica (Fig. 3). The patient was explained about the possibility of progression of disease and risk of malignancy also and advised strict follow up.

Discussion

Pelvic lipomatosis is a rare disease, first described in 1959 by Engles, that is characterized by overgrowth of mature fatty tissue in the perivesical and perirectal space and absence of delimitation by a capsule.¹ The etiology of pelvic lipomatosis is unknown. However various theories have been proposed for its genesis ranging from a localized manifestation of generalized obesity to an inflammatory response to repeated urological infections. Additionally, other authors suggest that it might be a localized response to hormonal and metabolic phenomena, while some others based on the observation of the highest incidence in black men, suggest the possibility of a genetic basis for the disease.¹

The exact incidence of this disease still remains undetermined, but it is known that it is most prevalent in men and individuals with dark skinned phenotype, most frequently at the third or fourth decade of life. The clinical manifestations result from the extrinsic compression of the structures comprising the urinary system, the lower intestinal tract and the vascular system. Thus, the occurrence of poor stream of urine,

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<https://doi.org/10.1016/j.eucr.2020.101492>

Received 13 October 2020; Received in revised form 5 November 2020; Accepted 8 November 2020

Available online 12 November 2020

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dyuria, nocturia, hematuria (less frequently), urgency, urinary incontinence and retention, besides repeated urological infections may be observed. Equally, constipation, tenesmus, diarrhea, lower limbs edema and thrombophlebitis, low back pain, suprapubic and perineal pain, painful ejaculation, epididymitis and orchitis may be observed.^{1,2} Although pelvic lipomatosis is considered a benign disease, distal encasement of the ureter by lipomatosis lesions it can cause upper tract obstruction and subsequent renal failure which has been seen in 6% of reported cases.² On physical examination, one may observe pain at abdominal palpation, presence of a palpable mass in the hypogastric region, urinary retention, elevation of the prostate at digital rectal examination, lower limbs edema and arterial hypertension.^{1,2}

The diagnosis is based on a high index of suspicion along with corroborative clinical and radiological findings. Plain X-ray film may show marked translucency in the perivesical area.³ Contrast radiography procedures like excretory urography and micturition cystourethrogram can reveal a "pear or gourd" shaped bladder, with an elevated bladder base and a medial or lateral deviation of the ureters due to

symmetric compression with associated hydronephrosis.³ Computed tomography (CT) is the cornerstone of the diagnosis of pelvic lipomatosis and shows the bladder and rectosigmoid are surrounded and displaced by homogenous tissue with low attenuation (-40 to -100 HU), signifying fat content. A pear-shaped bladder is a common finding on CT urogram that is a characteristic indicative of pelvic lipomatosis.³ Magnetic resonance imaging (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.³ Evaluation by cystoscopy detects abnormalities in 75% of patients and most often includes proliferative processes involving the bladder mucosa like cystitis glandularis and cystitis cystica.⁴ The biopsy of any suspicious lesion should be taken because of an association with adenocarcinoma of the bladder.⁴ Further, it is observed that cystoscopy in these patients often can be difficult because of the elongated bladder neck and posterior urethra.¹

The ideal management of pelvic lipomatosis is not well established due to the lack of literature and follow-up information. The

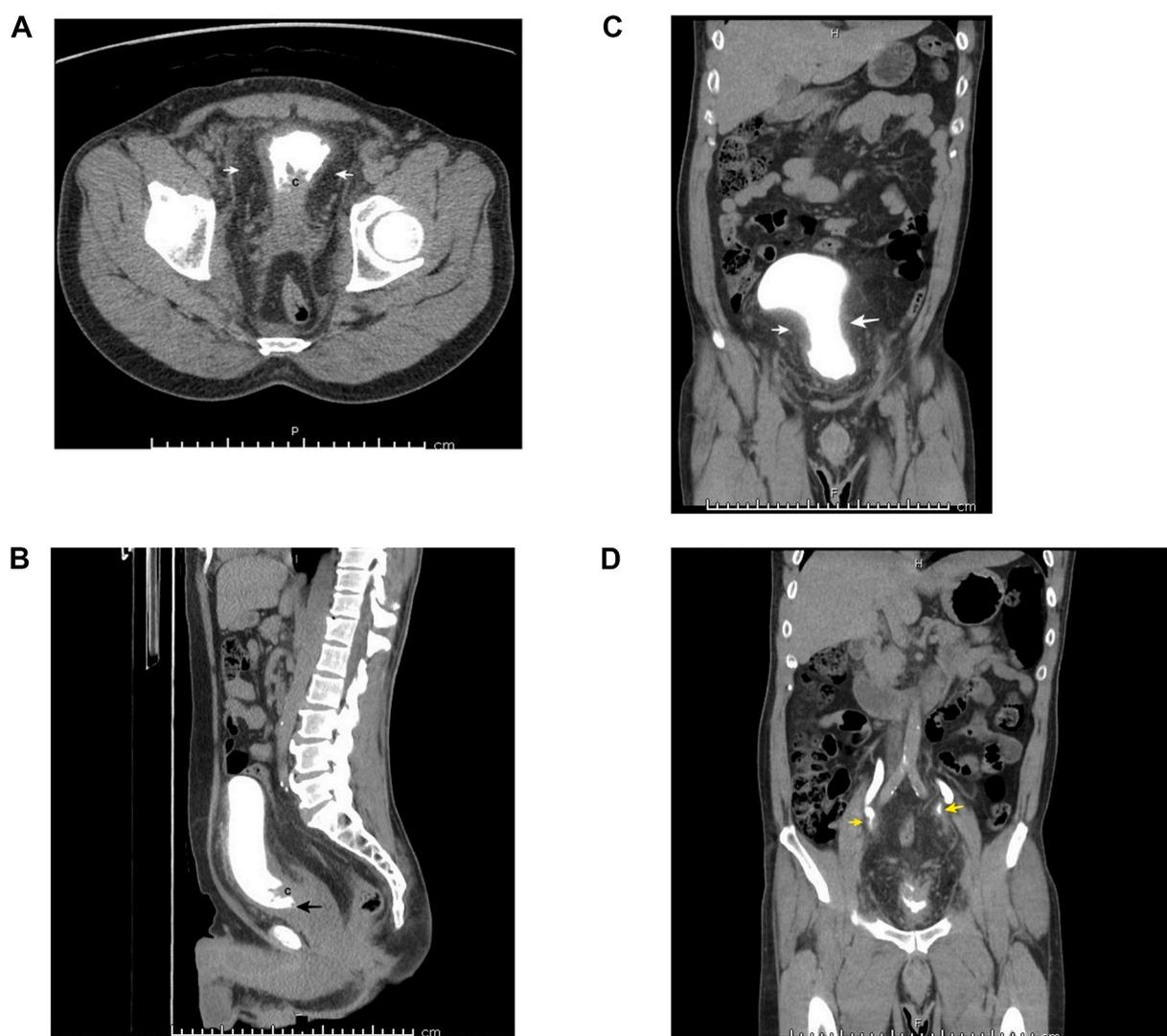


Fig. 1. Contrast enhanced computed tomography images.

A. Axial cut showing the presence of unencapsulated hypodense fat (white arrows) in the pelvis, causing extrinsic pressure of the bladder and proliferative cystitis within bladder (C).

B. Sagittal cut displaying the anatomical deformity of the bladder neck (black arrow) being pushed above due to the deposition of hypodense fatty tissue around bladder neck along with proliferative cystitis changes (C).

C. Coronal imaging showing a 'pear'-shaped bladder surrounded by hypodense fatty tissue (white arrows).

D. Coronal cut showing the ureters entering the proliferative pelvic fat (yellow arrows) with mild proximal ureteric dilatation. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

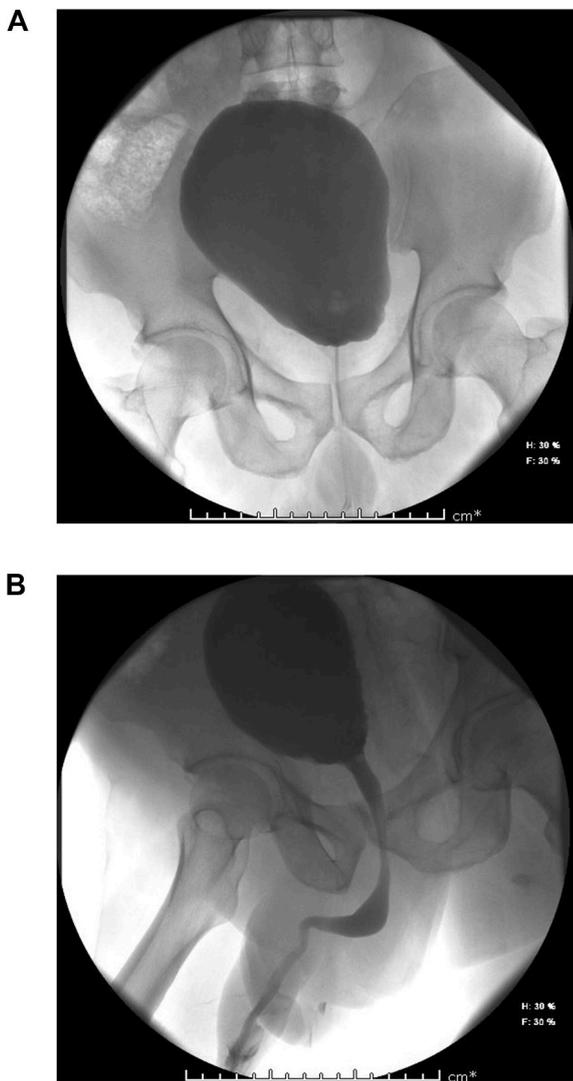


Fig. 2. Micturition cystogram showing A. Pear shaped bladder B. Elongated prostatic urethra with high bladder neck.

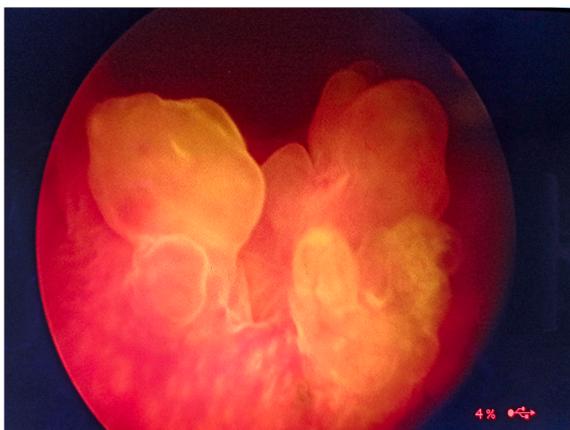


Fig. 3. Cystoscopic images of cystitis cystica in the prostatic urethra and posterior bladder wall.

recommended treatment options vary from watchful waiting to radical

surgical options, including total cystectomy and urinary diversion.⁵ 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. 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 but surgical excision has been described with successful results. Resulting ureteral obstruction with deteriorating renal function can be managed with nephrostomy, ureteric re-implantation, or simple cystectomy and ileal conduit.^{2,5}

Pelvic lipomatosis may remain dormant for many years. Though cystitis cystica and cystitis glandularis are considered benign lesions, the possibility of development of vesical adenocarcinoma should always be kept in mind, justifying the need for periodic follow-up. Though, there is no specific recommendation, it would be reasonable that patient's follow-up annually or biannually for radiologic and serum creatinine evaluation, with more invasive investigations in those who develop worsening symptoms or hematuria.⁵

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Declaration of competing interest

None.

Acknowledgments

None.

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