Zinner's Syndrome

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Abstract

A 52-year-old male was subjected to an F-18 fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) study for the evaluation of newly diagnosed non-Hodgkin's lymphoma. An incidental non-FDG avid urinary bladder mass was detected, as well as an absent kidney. Ureterocele was suspected, but subsequently a seminal vesicle cyst was confirmed on a CT urogram.

Keywords: F-18 fluorodeoxyglucose positron emission tomography/computed tomography, renal agenesis, seminal vesicle cyst, ureterocele, Zinner's syndrome

Introduction

A patient, newly diagnosed to have lymphoma, was found to have an asymptomatic mass in the urinary bladder. This is the first known case of seminal vesicle cyst, characterized by F-18 fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT). We also provide the characteristics of each differential diagnosis for bladder masses.

Case Report

A 52-year-old male presented with symptoms of fever, weight loss, and night sweats which had been progressing slowly over a period of past 1 year. He was found to have white blood cell count of 45,000/mcL. A CT of the abdomen showed hepatosplenomegaly and lymph node enlargement in the porta hepatis area. A bone marrow biopsy was performed which established the diagnosed of non-Hodgkin's lymphoma. Subsequently, an FDG PET/CT [Figures 1 and 2] was done to stage the disease and evaluate its extent. PET/CT detected several hypermetabolic lymph nodes, splenomegaly, absent left kidney and an incidental bladder mass. CT urogram [Figures 3–5] was later ordered, which confirmed the diagnosis of seminal vesicle cyst.

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Discussion

In this report, we describe a rare case of a congenital seminal vesicle cyst with ipsilateral renal agenesis. This is the first case known to us that has been detected by PET/CT in the background of widespread lymphoma. Although there was a bladder mass, the patient denied any symptoms of dysuria, hematuria, hesitation or incontinence. One month after the PET/CT was completed, a CT urogram was ordered to further evaluate the urinary bladder mass.

The seminal vesicle cyst is a rare entity and may be congenital or due to chronic inflammation or obstruction.[1] It is usually detected in the 3rd to 5th decade of life,[2,3] but may be detected earlier with the increasing use of CT and magnetic resonance imaging (MRI).[1] Patients may be asymptomatic or have vague perineal pain, dysuria, or infertility (37.5%). The cyst may be large enough to protrude into the bladder as in this case or may even prolapse into the bladder neck.[4] Seminal vesicle cysts as large as 6 cm have been reported to be asymptomatic.[1]

Often confused with ureterocele, the distinguishing factor of a seminal vesicle cyst is that it is associated with other congenital genitourinary anomalies. The association of a seminal vesicle cyst with ipsilateral renal agenesis was first reported in 1914 by Zinner and subsequently came to be known as Zinner's syndrome.[2] Ipsilateral renal agenesis is associated with the congenital form of the seminal vesicle cyst in 70–80% of cases,[3] but absent ureter, trigone, testicular ectopia may also

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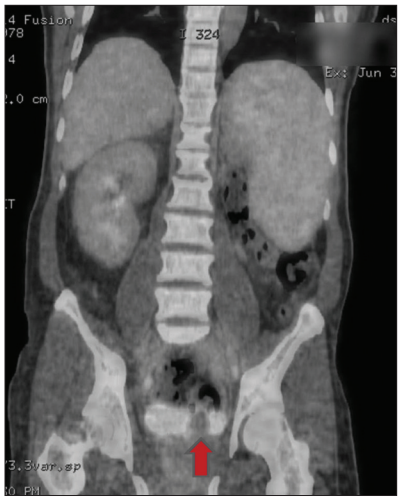

Figure 1: F-18 FDG PET/CT study, showing coronal section of abdomen and pelvis. The scan shows an enlarged spleen with relatively more FDG uptake than the liver. In addition, there are metabolically active inguinal, iliac, periaortic, prevertebral lymph nodes, as well as increased bone marrow activity. The left kidney is absent with compensatory hypertrophy of right kidney

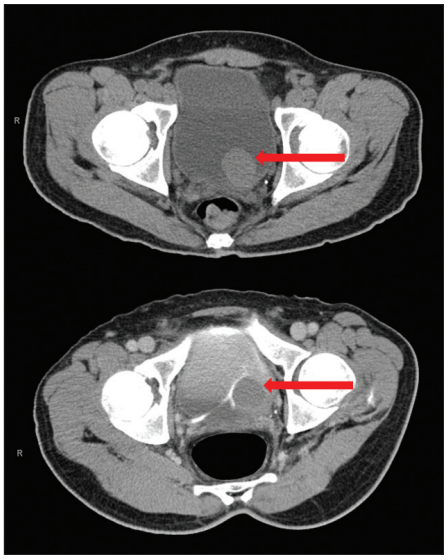

Figure 3: CT urogram shows a urinary bladder mass having higher attenuation density with the pre-contrast images. The lesion does not demonstrate any significant enhancement

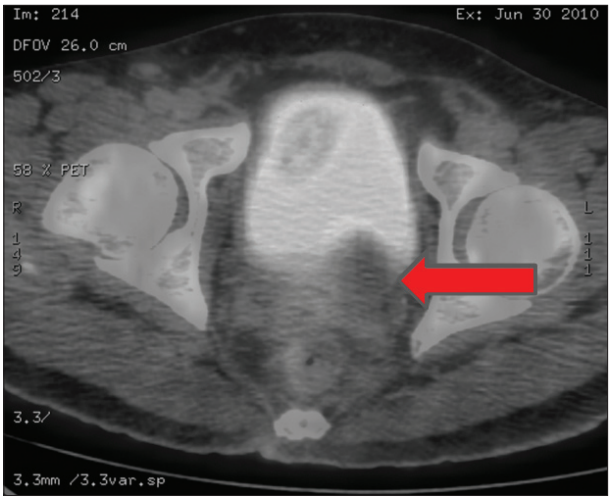

Figure 2: F-18 FDG PET/CT study, showing an axial section of pelvis. An incidental non-FDG avid urinary bladder mass is noted on the left side posteriorly (arrow)

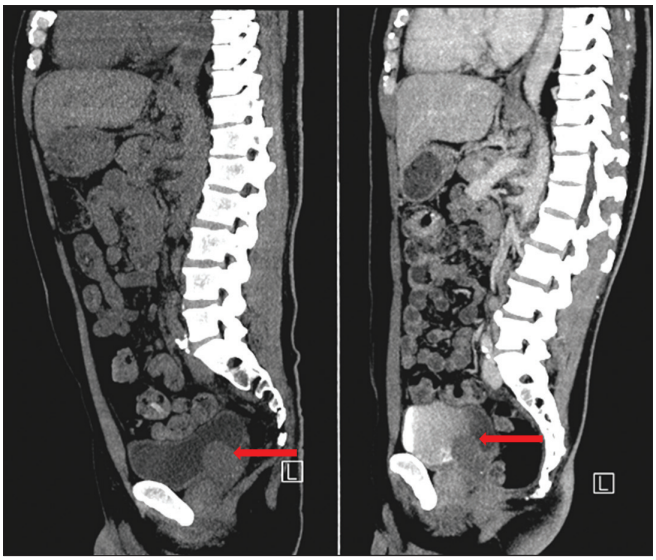

Figure 4: A sagittal view of the CT urogram with and without contrast, showing the seminal vesicle cyst

be associated with this condition.[2,4] In 27% of cases, a ureteral bud remnant may also exist.[3]

The embryogenesis of this condition is believed to occur between weeks 4 and 7 of gestation when the ureteral bud sprouts from the mesonephric duct. Usually, the ureteral bud must rise into the center of the metanephric blastema to form the kidney. However, the ureteral bud may form too early and then migrate too cranially. Subsequently, it could miss the metanephrogenic blastema which would cause a dysplastic or absent kidney.[2] Furthermore, during this prolonged period, the ureters may expand and implant ectopically or form into an ureterocele. Normally, the ejaculatory ducts are derived off the mesonephric system. However, the ejaculatory duct forms abnormally in these cases and seminal vesicle fluid builds up inside forming a cyst.[1,2]

Vaso-vesiculography, which involves aspiration of cyst and injection with contrast, is the diagnostic test of choice; however, this is rarely done unless the patient is symptomatic, and to avoid complications.[3] A more commonly recommended method is intravenous urography or rectal ultrasonography. On CT, the cyst will have high signal intensity because it contains proteinaceous material.[4] Laparoscopic excision is the treatment of choice but should only be performed if the patient is symptomatic.

Ureterocele may mimic this diagnosis. Ureterocele is a

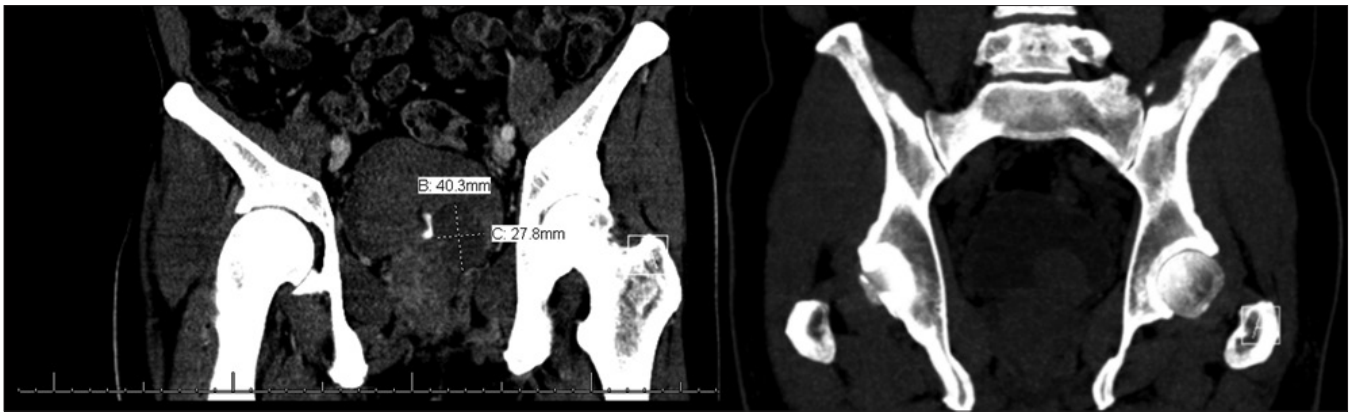

Figure 5: A coronal view of the mass, with and without contrast. Note the bright ureter in the midline of the bladder. This ureter originates from the right kidney

sacculation at the terminal portion of ureter and may be caused by congenital malformation of the ureter or chronic inflammation/trauma.[5] In adult males, the ureterocele is usually unilateral and inserts into the normal position of the trigone. It may also contain a stenotic portion which would cause the ureterocele to look like a "cobra head" on an urogram.[6] In the CT pelvis, the mass does indeed seem like an ureterocele since it is round inside the bladder wall and is unilateral. The position of insertion matches where the ureter would have inserted. Ureterocele may be asymptomatic or cause symptoms of dysuria, urgency, and recurrent urinary infections. The formation of ureterocele is equivocal and no single theory explains all the types of ureterocele. In a normal embryo, the ureteral bud branches off the mesonephric duct and rises cranially to join the metanephric blastema to form the nephrons. At this time, there may be an incomplete breakdown of the ureteral membrane between the ureteral bud and the mesonephric duct. This membrane that breaks down is also known as Chwalla's membrane and needs to be patent for ureter to function correctly. Otherwise, there will be obstruction of the ureters which will lead to an ureterocele.[4]

Mullarian duct cyst occurs in the midline, arising behind the verumontanum and extending above the base of prostate. It usually has a beak-like projection into the bladder, shaped like an inverted pear or simple oval.[4] Unlike seminal vesicle cyst, it is not located laterally and it also contains no spermatozoa or fructose.[3,4] Only 10% of these cases are associated with renal agenesis or dysgenesis.[3]

Prostatic cysts are usually smaller (0.75–3cm) and contain no sperm.[3,7] They are usually localized at the level of the verumontanum and can be either midline or lateral. These cysts are unilocular, have thin wall and are hypointense with respect to the prostate.[7]

Gartner's duct is a remnant in female development of the mesonephric ducts, which should have atrophied after guiding the Mullerian duct to form the vagina and uterus. Like seminal vesicle cyst, Gartner's duct cyst is also a pelvic cyst associated with unilateral renal dysgenesis. Likewise, the embryologic malformation is thought to result from too-cranial migration of the ureteral bud from the mesonephric duct.[8] The mesonephric duct persists and develops into a cyst which may retain urine if attached to the ureter. These cysts are located near the cervix, wall of the vagina, or lie on the broad ligament posterior to the bladder (50% of cases). It is not uncommon to find them protruding into the bladder and simulating an ectopic ureterocele.[4]

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