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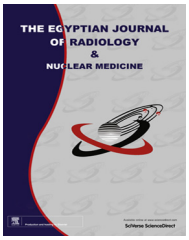

CASE REPORT

Congenital seminal vesicle cyst accompanying with ipsilateral renal agenesis in an adolescent patient: A pediatric radiologist approach to Zinner's syndrome

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Adolescent patient

Abstract A fifteen-year-old boy who had complaints of left sided pelvic pain with known ipsilateral left renal agenesia was referred to pediatric radiology department. Incidentally, his sonography examination revealed a dilated tubular structure located in the retro-vesicular region from cephalic to prostate. Contrast enhanced pelvic MRI showed a huge seminal vesicle cyst which is over 6 cm without a mass effect near the aspect border of the prostate and bladder. The patient was diagnosed with Zinner syndrome. The patient doesn't have new complaint with no definite increase in the diameter of the cyst. In this case presentation we are discussing the Zinner syndrome's imaging findings from a pediatric radiologist approach with a brief review of the literature.

1. Introduction

Seminal vesicle cyst incidence is 0.005% which is extremely rare in the population (1). The exact incidence of the syndrome in the pediatric population is unclear. The mean age of presentation is 30.2 years with right sided predominance (2). In 1914, Zinner et al. described the association of seminal vesicle cyst with ipsilateral renal agenesis. The seminal vesicle cysts can be either in an isolated form or accompanied by upper urinary tract anomalies or associated with autosomal dominant polycystic kidney disease. In Zinner syndrome an additional finding of ejaculatory duct obstruction is required. Related symptoms of duct obstruction such as the male genital system abnormalities including azo-hemospermia could be observed. In adolescent or pediatric age group, the patients are not checked for male reproductive symptoms and therefore this finding explains the late diagnostic age in the literature. In adolescent age group, US is the first choice of imaging modality. Furthermore, including pelvic region masses regarding the adolescent age, contrast enhanced MRI is the second line preferred imaging modality. To figure out the extension borders of the seminal vesicle cyst and to understand the relationship of ductus ejaculatorius, MRI is superior to sonography in

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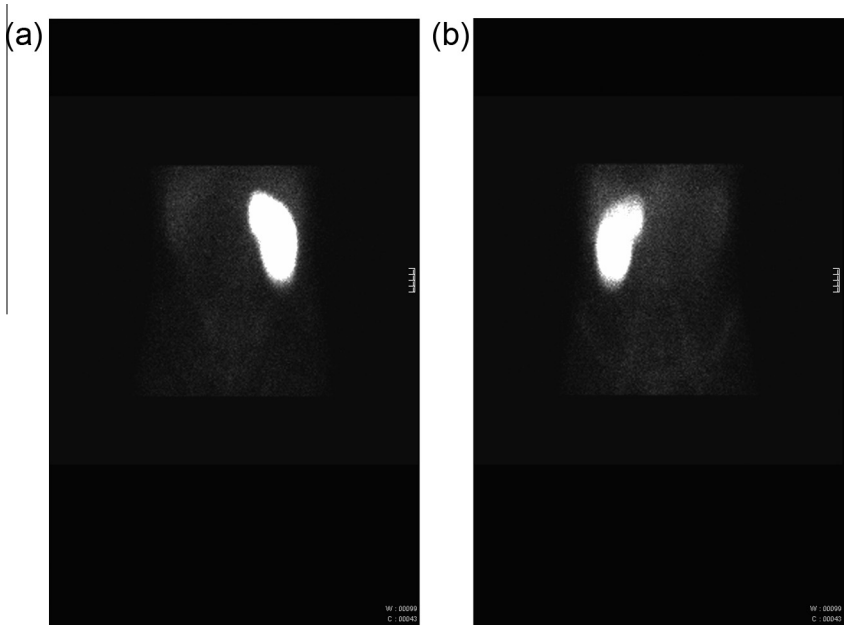

Fig. 1 The fifteen-year-old boy with recurrent pelvic pain has undergone scintigraphy examination: The image (a) shows the absence of left kidney from anterior view, (b) as same as the image a, the posterior view of the scintigraphy confirms the absence of the left kidney.

pediatric age group. In this case presentation we discuss the Zinner syndrome imaging findings from a pediatric radiologist approach with a brief review of the literature.

2. Case

A fifteen-year-old male patient was referred to our clinic with pelvic pain from a pediatric nephrologist. The left renal agenesia was demonstrated by renal scintigraphy when he was ten years old (Fig. 1). He had pain symptoms in the pelvic region but the physical examination and blood tests did not show underlying cause of a significant pathology. Sonography examination revealed dilated tubular structures located in the retrovesicular region from cephalic to prostate. Patient was subjected to pelvic MRI for further evaluation. Pelvic MRI demonstrated round cystic masses with $84 \times 74 \times 52$ mm in the left seminal vesicle with contents of high signal intensity in T1-weighted images and low signal intensity in the T2-weighted series. Diffusion weighted images did not show any restriction in the lesion. After contrast enhancement, neither the wall of the cystic structure nor the inner elements show enhancement (Fig. 2). Within the revert of the patient to rule out the underlying cause in male reproductive system scrotal ultrasonography examination was done. It revealed that the volume and sonoechogenity of the both testes were in the normal limits. No definite dilatation of vas deferens or ejaculatory duct or any abnormality in the seminal vesicle was seen on the right side. The normal left seminal vesicle was not identified. There was a slight compression over the left ejaculatory duct. The distal part of vas deferens presented normal caliber. The vas deferens was dilated along its superior course in spermatic cord. There was a slight hydrocele in the left scrotum (not shown). Regarding the left ureter, there was no evidence of ectopic insertion or blind ended seen. The patient is still on follow-up for one year without any additional complaint. There was no change in the cyst size or appearance.

3. Discussion

The association between seminal vesicle cyst and ipsilateral renal agenesis was firstly described by Zinner et al. in 1914. The seminal vesicle cyst usually found in the second to third decade due to the actual reproductive life complaints begins. They are found incidentally during routine procedure within the developing imaging procedures (3). The cyst less than 6 cm usually does not have mass effect and causes obstructive symptoms on either the vesicle or prostate (4).

In the late adolescent age group, small percentage of presentation is the palpable abdominal mass or palpable fluctuant mass arising from the prostate. In this group of patients there are usually accompanying symptoms of epididymitis, prostatitis, azo-/hematospermia causing infertility problems. For this aim, usually transrectal ultrasound is useful to determine the exact localization and able to get seminal vesicle aspiration as also a method of treatment indeed (5). The cyst greater than 12 cm is termed as 'giant' which has a definitive causative symptoms of mass effect on bladder and colon (2). In our case, although the cyst size was larger than 6 cm, the pelvic imaging modality did not show any compression over the colon segments. However, the left ductus ejaculatorius findings could not serve as the results of the mass effect in relation within the seminal vesicle cyst. These findings are the results of the embryologic evaluation process.

In the etiology of the congenital seminal vesicle cyst accompanying with the renal agenesis, insufficient development of the lower mesonephric duct and the differentiation of the metanephric blastema are considered as the basic mechanisms of the disease. If the ureteral bud arises more in cephalic position to the mesonephric duct blastema there will be delayed absorption of the caudal mesonephric duct and there will be emptying problem of the mesonephric duct derivate such as the duct of ejaculatorius or seminal vesicles (2,4,6). Therefore by time, the inclusion of the secretions will be accumulated

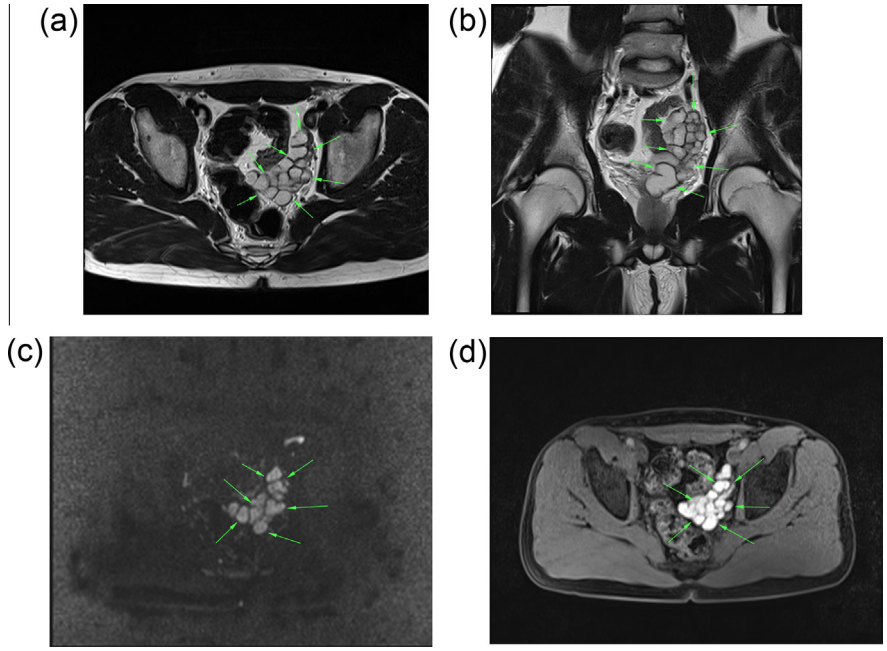

Fig. 2 The fifteen-year-old boy with pelvic pain at magnetic resonance imaging examination; (a) axial-T2-weighted sequence shows the dilated tubular structures, (b) coronal-T2-weighted hyperintense sequence shows dilated tubular structures in the left pelvic region superior to colon segments with thin linear septations. (c) Diffusion weighted images on coronal plane sequence do not demonstrate diffusion restriction. (d) On T1-weighted contrast enhanced sequences, dilated tubular structures do not enhance after contrast enhancement in axial images. All the lesions are demonstrated by large arrows.

in the seminal cyst because of the drainage problem. Usually the renal agenesis is unilateral and ipsilateral. There are only three cases demonstrated at the contralateral side in the literature (6–8).

Bilateral renal agenesia association of seminal vesical cyst is common in the patients with autosomal dominant polycystic kidney disease. This incidence of bilaterally is described as up to 43% in the literature (9). The ectopic ureter insertion into the seminal vesicle is extremely uncommon reported with accompanying by reflux and obstruction and therefore a primary adenocarcinoma could cause this (10).

Magnetic resonance imaging is an accurate diagnostic tool for identifying the upper and lower genitourinary system abnormalities. In particular in the child and adolescent age group because of its radiation free-dosage protocol it is preferred than computerized tomography (CT). Additionally MRI can show the ectopic insertion of the ureter with a high resolution (3,4,10). On MRI seminal vesicle cysts have variable signal intensity depending on the protenious content of the fluid (3,6,11). They have hypo to isointense on T1-weighted images and hyperintense on T2-weighted images. They do not enhance after intravenous gadolinium administration. On diffusion weighted images there is not any diffusion restriction because of the basics of the sequence.

In differential diagnosis of cysts and cystic dilatation of seminal vesicles, several reasons can be considered, such as the cystic structure of prostatic prostate gland, prostatic utricle cysts, ejaculatory duct cysts, mullerian duct cysts, hydronephrotic pelvic kidneys, bladder diverticula, and ureteroceles. The main differentiation is usually based upon the location such as median, para-median, or lateral located. Mullerian duct cysts and ejaculatory duct cysts are midline in location. Diverticulosis of ampulla of vas deferens and ectopic ureterocele are more laterally located. The presence of spermatozoa in the aspirate may differentiate seminal vesicle cysts from mullerian duct cysts. However mostly symptomatic cases are detected at the early stage of adulthood or late adolescent ages. Usually they are demonstrated by the analysis of spermiogram. MRI is also helpful for accurate preoperative surgical planning for seminal vesicle cyst excision. Surgical excision of a seminal vesicle cyst depends on the size and location of the cyst and the presence of clinical symptoms.

A unique pentad of mesonephric duct abnormalities on imaging, including cystic dysplasia of rete testis, seminal vesicle cyst, ipsilateral renal agenesis, partial hemitrigonal development, and epididymal dilatation were described by the Casey et al (12). This pentadology includes an extension into the testicular and epididymal structures.

The treatment methods include ultrasound guided cyst aspiration, transurethral unroofing and laparoscopic cyst excision. There are several case series successfully treated with laparoscopic method (1,2,6,7). Transuretral cyst excision is the most preferable method in the early adulthood and late adolescent age group.

In conclusion the Zinner syndrome is the triad of mullerian duct abnormality comprising of unilateral renal agenesis, ipsilateral seminal vesicle cyst, and ejaculatory duct obstruction. It is an uncommon but important diagnostic consideration in young age especially in the late adolescent and early adulthood ages when the patient presents with recurrent urinary symptoms and unsolved chronic pelvic pain. The modern-day imaging techniques have facilitated the early diagnosis of this entity.

In adolescent and pediatric age group if a cystic lesion is diagnosed in the pelvic region and in close relation with the prostate and bladder, congenital seminal cyst should be

considered in diagnosis. Within the high probability of accompanying of the upper urinary system abnormalities to the seminal vesical cysts both ipsilateral and contralateral genital systems should be checked for congenital abnormalities.

Conflict of interest

The author attributes that there is no conflict of interest.

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