Seminal Vesicle Cysts with Unilateral Renal Agenesis and Contralateral Ureteral Stenosis in a β-Thalassemic Patient: An Unknown Association by Incomplete Development of the Mesonephric Duct

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Introduction

Seminal vesicle cysts are rarely reported in the literature, especially when associated with other abnormalities. Van den Ouden et al. [1] analyzed 52 cases of seminal vesicle cysts combined with unilateral renal agenesis.

Seminal vesicle cysts may be congenital or acquired [1–4]. If congenital, they are generally associated with other congenital anomalies of the urinary tract, like unilateral renal agenesis with an incidence between 60 and 68%. However, the association with congenital anomalies like deferent vessels or agenesis of the ejaculator duct and insertion in an abnormal ureteral location [1–4] are very rare events.

The patient who came to our observation presented not only seminal vesicle cysts and unilateral renal agenesis but also contralateral ureteral stenosis with hydronephrosis (2nd degree) and β-thalassemic trait; this complex congenital defect is due to the common embryonic origin of these structures from the mesonephric duct.
Case Report

A 13-year-old male patient came to our observation for a colic pain on the right hip. There was no family history of colic pain and renal alterations. Renal function was normal after routine examinations. Since the mean red cell volume was 58.33, hemoglobin electrophoresis screening was performed and it showed β-thalassemic trait.

Abdominal and pelvic sonography showed right kidney agenesis with a pelvic mass characterized by reduced echogenicity. Urography confirmed agenesis of the right kidney (fig. 1a) and showed hypertrophy of the left kidney with moderate ureteral enlargement of the distal third due to an insertion defect in the bladder, suggestive of primary segmental megaureter without obstruction (fig. 1b). MR and renal angioscintigraphy imaging confirmed these findings. The latter showed normal elimination of the radionuclide after furosemide. MR studies also showed multilocular cysts of the right seminal vesicles.

One year later the renal and pelvic MR examination showed an increase in volume of the cyst in the right seminal vesicles with ramifications through the ejaculator ducts (fig. 2a, b). Genetic and chromosome analyses in the patient and in his parents were negative. Radiological examination of the head and cervical vertebrae considering Mayer-Rokitansky-Küster syndrome was negative too. During pregnancy the patient’s mother did not take any drugs likely to cause alterations in fetal development.

Surgical intervention was made for removal of seminal vesicles; histological examinations showed protein-mucous material mixed with blood.

Discussion

Cysts of the seminal vesicles are rarely observed and can be congenital or acquired. If congenital, they may be associated with other anomalies of the urogenital system like homolateral renal agenesis (60–68%) and rarely with changes in ureteral insert in bladder as in our patient.

Alteration in the development of the distal mesonephric duct during embryological life has been reported to be the main cause of this complex disease.
The metanephros starts its development around the 4th week of the embryonic period, connecting to the mesonephric duct or wolffian duct [5].

Seminal vesicles appear like ramifications of the distal segment of the distal duct during the 13th week of the embryonic period. Renal development starts from the metanephric blastema, but the link with the mesonephric duct is essential for complete and final development of the urogenital system. The metanephric blastema releases a substance, that acts as an inducer and promotes the development of the ureteral bud, whose exit is off the wolffian duct. The ureteral bud releases substances that promote the formation and development of secretor tubules once it reaches the metanephric blastema.

Generally, seminal vesicle cysts have variable size, reaching 5 cm in transversal diameter, with an asymptomatic course during the first years of life, becoming symptomatic with micro/macrohematuria, dysuria, and rarely hemospermia, that may be confused with orchitis, epididymitis after the first decade of life.

The diagnosis is exclusively based on radiological examination. First choice examination should be sonography that detects cysts in the posterolateral (or rarely mesolateral) bladder area. Abdominal ultrasound is generally used for evaluating the size of cysts and their relation with adjacent organs.

CT and MRI are important for the differentiation of cystic contents that, on some occasion, may be hemorrhagic [2, 6]. The differential diagnosis of pelvic cysts includes cysts of müllerian ducts, prostatic utricular enlargement, cysts of prostatic gland, cysts of Gartner’s duct, and in our case the Rokitansky-Kuster-Hansen (RKH) syndrome [7, 8].

Nevertheless, the latter syndrome, that generally affects females (with vaginal agenesis, bicornate uterus and uterus didelphys), and when it affects males it is generally associated with seminal vesicle alterations and skeletal changes in the cervical vertebrae and the head, is due to an autosomal dominant disorder, as well as to all müllerian malformations [9].

In our patient, with suspected RKH syndrome, in order to make a correct differential diagnosis, we performed...
genetic and chromosome analysis, together with radiological evaluation of the skeleton and the head, both with negative results.

This case represents a very rare association of seminal vesicle cysts, unilateral renal agenesis, and alteration in contralateral ureteral insertion in the bladder responsible for hydronephrosis (2nd degree) and β-thalassemia trait. Moreover, it was not necessary to correct, with surgery, the defect of ureteral insertion since a previous radiological examination (renal angioscintigraphy) showed a complete elimination of the drug through the bladder after stimulation with furosemide.

The removal of the right seminal vesicle was necessary due to the occurrence of tumor degeneration in 70–80% of cases and to avoid the risk of kidney failure due to hydronephrosis in a patient with single kidney.

Surgical intervention through laparoscopy is to be preferred to endoscopy because microinvasive techniques may be responsible for secondary erectile dysfunction [10]. Further studies will help to understand the role of minor β-thalassemia, found in our case that, at present, is likely to be occasional. Another aspect to be taken into account is the fast increase of the cyst; patients that show these cystic alterations must be frequently examined through sonography.

One year after surgical intervention, the patient had normal renal function on routine examination.

References