Annals of Clinical Case Reports

Case Report
Published: 23 Sep, 2021

Inguinal Hernia Containing a Fallopian Tube and Ovary with Giant Cyst in Patient with Mayer-Rokitansky-Küster-Hauser Syndrome

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Abstract

Inguinal hernia represents the most common congenital anomaly in childhood, significantly more often encountered in boys. About 15% to 20% of the inguinal hernias in girls contain uterus with adnexa, and are commonly associated with congenital anomalies of the genitourinary system such as the Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome. We report a case of a large ovarian cyst which herniated into the right inguinal canal, in combination with MRKH-2 syndrome.

Keywords: Hernia; Ovarian cyst; MRKH; MURC

Introduction

Inguinal hernia represents the most common congenital anomaly in childhood, significantly more often encountered in boys. About 15% to 20% of the inguinal hernias in girls contain uterus with adnexa [1,2], and are commonly associated with congenital anomalies of the genitourinary system such as the Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome [3]. Upon impact on genitalia only the condition is defined as MRKH type-1 syndrome. Type 2 MRKH or MURC-associated syndrome includes renal anomalies (unilateral agenesis, ectopic kidney, horseshoe kidney), skeletal anomalies (fused vertebrae, scoliosis), and more seldom-hearing loss or cardiac defects. The type of inheritance is autosomal-dominant with incomplete penetrance and variable expression. A rare case of a ruptured hemorrhagic ovarian cyst within an incarcerated inguinal hernia has been described [4].

We report a case of a large ovarian cyst which herniated into the right inguinal canal, in combination with MRKH-2 syndrome.

Case Presentation

We present a 30-year old female patient with mental retardation and amenorrhea, hospitalized in our clinic due to accidentally established lump in the right inguinal area, asymptomatic and without any history of trauma at this area. Physical examination revealed a rounded swelling with smooth surface, about 10 cm in diameter, fixed and relatively superficial. Ultrasound imaging findings included a cystic formation in the right abdominal half with an augmented internal echoes and size 98 mm/87 mm, possibly originating from the right ovary; agenesis of the right kidney and slightly increased size of the left kidney with dilated parenchymal area. CT-scan confirmed the agenesis of the right kidney (Figure 1) with left-sided lateralization of the uterus without other pathological alterations. A herniated thin-walled (2 mm) cystic structure was described along the course of the inguinal canal with intra- (33 mm/27 mm) and extra-abdominal (103 mm/97 mm) component, in close proximity to an ileum intestinal loop, but there was no evidence of any connection between them (Figure 2). No nutrient vessels to the structure were spotted, and it remained relatively hypodense during the contrast phases. The structure described exerted pressure on the femoral vein in the inguinal canal to almost complete obturation of the vein in the contact segment. As an additional finding, a marked S-shaped scoliotic deformation due to congenital vertebral alterations was established-wedge-shaped L2, block vertebra of L1, Th12 and Th11; a wedge-shaped vertebrae at the level of Th10; and a sacrococcygeal deformation. Due to the proximity of the above-described formation to the great vessels we also performed a Doppler examination, establishing pervious deep veins of the lower extremities bilaterally.

Through a right-sided parainguinal approach we reached a large cystic formation with 10 cm

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Received Date: *01 Sep 2021*
Accepted Date: *20 Sep 2021*
Published Date: *23 Sep 2021*

Citation:
Shumarova S, Maslyankov S, Gribnev P, Sokolov M. Inguinal Hernia Containing a Fallopian Tube and Ovary with Giant Cyst in Patient with Mayer-Rokitansky-Küster-Hauser Syndrome. Ann Clin Case Rep. 2021; 6: 1995.

ISSN: 2474-1655

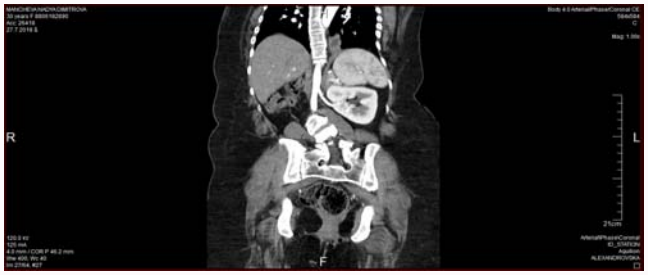

Figure 1: CT scan confirmed the agenesis of the right kidney.

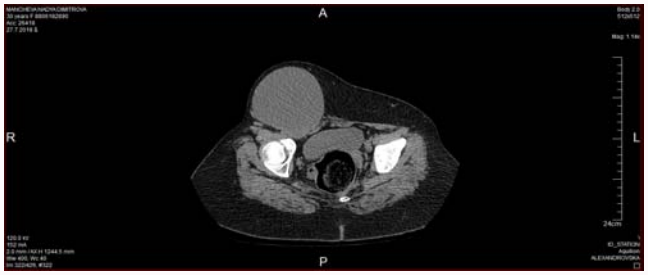

Figure 2: CT scan confirmed ovarian cyst into the inguinal canal.

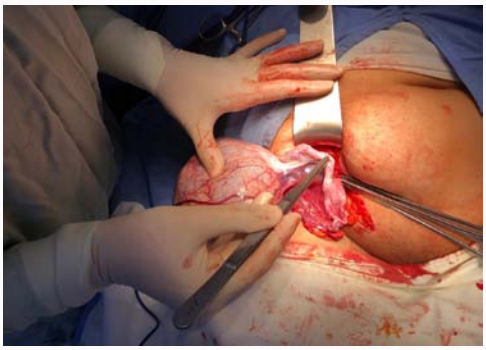

Figure 3: Large ovarian cystic formation with 10 cm to 11 cm diameter herniated together with the right uterine tube into the right inguinal canal.

to 11 cm diameter, originating from the right ovary and herniated together with the uterine tube into the right inguinal canal (Figure 3). We performed adnexectomy together with the cyst extirpation, followed by plastic reconstruction of the posterior inguinal wall. Histological evaluation revealed a cystadenoma of the right ovary with fibrous capsule with multiple endophytic papillary structures, covered by single-layer cuboidal epithelium, and hyper-cellular stroma containing elongated cells and collagen.

Discussion

Male and female reproductive systems have common initial development. Reproductive glands are initially situated into the abdominal cavity, and later both the testicles and the ovaries descend, but to a different degree. The testicles descend following their gubernacula by invagination of the parietal peritoneum called vaginal processus, and by the end of the 8th month of the pregnancy are localized in the scrotum. Normally after the birth the vaginal processus obliterates. In girls the non-obliterated vaginal processus postnatally is referred to as the canal of Nuck [2] and represents a cause for ovarian herniation, sometimes together with uterine tubes.

Although seldom, indirect inguinal hernias are observed in women in reproductive age as well [3]. In such cases the etiopathogenesis includes a Muller anomaly, caused by fusion anomalies of the Muller canals to Muller aplasia and Mayer-Rokitansky-Küster-Hauser syndrome (MRKH). A recent study of Deng et al. [5] compared the percentage of both types of MRKH among various Chinese race groups, as well as between them and the Caucasians. Their analysis included a total of 274 Chinese patients: 197 (71.9%) with type 1 MRKH syndrome and 77 (28.1%) with type 2 MRKH syndrome. They established renal anomalies in 53.1% and skeletal anomalies in 40.8% (51/125) of the cases, with the highest percent of them for scoliosis 84.3%. Type 2 MRKH syndrome is more common in the European population compared to the Chinese, where the percentage of renal anomalies is lower. We have not studied the chromosome model of our patient, but the renal agenesis and the skeletal deformations combined with inguinal herniation of a large ovarian cyst are a fact, which makes the case casuistic.

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