Giant paraovarian cyst in a child complicated with torsion

Džinovska paraovarijumska cista komplikovana torzijom kod devojčice

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Abstract

Background. A variety of benign cyst may occur in and around the ovary and broad ligament and simulate serous cystadenomas. The majority of broad ligament and paraovarian epithelial tumors are serous neoplasms of low malignant potential and presented with a pelvic mass with or without ascites or pain, but without involvement of the ovary. Ovarian torsion and paraovarian serous cystadenoma are rarely reported. Case report. We presented a case of giant paraovarian cyst in a 14-year-old girl, with characteristics of serous cystadenomas grossly and microscopically, and complicated with double adnexal torsion. A computed tomography scan showed large hypodense cystic mass (measuring 30 × 26 × 12 cm), occupying the whole abdominal cavity, with no adhesion to the surrounding organs. Conclusion. Precise clinical data as well as pathological examinations based on immunohistochemical stainings were important in making the diagnosis. These rare cystic lesions of para/mesoovarian location in children and their unclear histogenesis might be a histopathological diagnostic problem.

Key words: ovarian cyst; cystadenoma; torsion; diagnosis; immunohistochemistry; adolescent.

Introduction

A variety of benign cyst may occur in and around the ovary and broad ligament and simulate serous cystadenomas both grossly and microscopically. Cystic lesions of the ovary are most common during infancy and adolescence, which are hormonally active periods of development. Cysts are mostly nonneoplastic in children and could be categorized as follicular, simple, and corpus luteum cysts. The rete ovarii rarely give rise to cysts and to benign and malignant tumors. These are mostly found in postmenopausal women and only rarely in children. The most common clinical presentation of ovarian cysts are abdominal pain, nausea and vomiting, and a history of previous episodes of similar pain and low grade fever.

One of the most intriguing aspects of ovarian epithelial neoplasms is their histogenesis. A suggestion is made that components of the secondary Müllerian system, which include parovarian/paratubal cysts, rete ovarii, endosalpingiosis, endometriosis, and endomucinosis, merit some consideration as to their possible role in ovarian tumorigenesis.

We presented a case of giant paraovarian cyst in a 14-year-old girl, with characteristics of serous cystadenomas both grossly and microscopically, and complicated with double ovarian torsion.

Case report

Clinical data

A 14-year-old girl (strongly obese), was presented to the Department of Surgery, Military Hospital Niš, with lower
right quadrant abdominal pain for the previous 24 hours, of moderate intensity and periodical characters, and not accompanied by nausea and vomiting.

Computed tomography (CT) showed a large hypodense cystic mass (measuring $30 \times 26 \times 12$ cm), occupying the whole abdominal cavity (Figure 1). Laboratory analysis showed increased erythrocyte sedimentation rate – 32 mm/h (one hour), mild neutrophilia (white blood cell – WBC : $11.8 \times 10^9$/l; neutrophil leucocytes 83.4%). There were also signs of mild anemia syndrome with serum ferum/iron level decrease of 6.6 nmol/l. The remaining biochemical analyses showed normal values.

At laparotomy, a smooth cystic mass that originated from the right paraovarian tissues and extended to the upper abdomen was found, but without taking the right ovary. Double right adnexal torsion was found, too. There was no adhesion to the surrounding abdominal organs. Aspiration from cyst was evacuated around 6 l of clear serous fluid. Complete excision of the cyst with the hemorrhagic infarcted right ovary was performed. Contralaterally, there was paraovarian serous cyst (measuring $7 \times 4 \times 4$ cm), thin-walled, translucent and filled with clear watery fluid. Complete excision of the cyst was performed, and left adnexa was conserved. Pathologic examinations of the excised surgical material were performed.

Pathological findings

Giant empty cystic tumor had following characteristics: 15 cm in greatest diameter, smooth and wrinkled, glistening, white-greyish the external surface (Figure 2). On section, the unilocular cyst showed ragged and wrinkled greyish inner surface (Figure 3). The wall of the cyst was thickened and toughed, in some place separated and with hemorrhages.

Formalin-fixed, paraffin-embedded tissues samples were sectioned at 5 µm thick sections and stained with hematoxylin and eosin (HE), Alcian Blue – Periodic Acid Schiff (AB-PAS) and Masson Trichrome. Representative materials were stained with a panel of antibodies using the labeled streptavidin-biotin-peroxidase method according to the manufacturer’s instructions (LSAB2 Kit, Dako). The primary antibodies used included estrogen receptor (ER) (clone 1D5), progesterone receptor (PR) (clone PgR636), vimentin (VIM) (clone V9), cytokeratin (CK) (clone AE1/AE3) and smooth muscle actin (SMA) (clone 1A4). The chromagen was 3,3'-diaminobenzidine (DAB), and the slides were lightly counterstained with Meyer’s hematoxylin. All reagents were from Dako Company (Denmark, Copenhagen).

Microscopically, the cyst wall was composed of fibrovascular tissue containing bundles of smooth muscle (Figure 4). The lining of cyst was composed of a single layer of tubal-type columnar epithelium with ciliae (Figure 5). A single layer of tubal-type columnar epithelium was strong immunoreactive for PR (Figure 6), VIM and CK. Immunoreactivity for ER was negative.

In the mesoovarian region some neurovascular elements were noted, while the hilus cells were not found despite the multiple analysis of several tissue samples. Microscopically, cystic and rare atretic follicles were discovered inside the membrane of the ovarian tissue sample, as well as rare corpus luteum in regression and morphological elements of hemorrhagic infarction.

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Discussion

The majority of broad ligament and paraovarian epithelial tumors are serous neoplasms of low malignant potential and presented with a pelvic mass with or without ascites or pain, but without taking the ovary. One of the most intriguing aspects of ovarian epithelial neoplasms is their histogenesis. The most common subtypes of these tumors are morphologically indistinguishable from neoplasm arising from those organs of the female genital tract that are embryologically derived from Müllerian ducts. Thus, the serous subtype is similar to tumors arising in the fallopian tubes. The currently favored hypothesis is that ovarian epithelial tumors arise from single cell layer lining the ovarian surface, which is often referred to as surface epithelium. This cell layer, which is continuous with the mesothelial lining of all pelvic and abdominal structures, is morphologically very similar to the mesothelial lining of peritoneal surface even away from the ovary. In addition, Müllerian-lined cysts are common not only in the ovary, and are also frequently seen in paraovarian tissues with no apparent direct connection to the ovary. Laughlin used the term “secondary Müllerian system” to designate structures lined by Müllerian epithelium found out of the uterus, cervix, and fallopian tubes.

A significant positive diffuse immune reactivity of the cystic epithelium for CK and VIM in this case can contribute to the suggested theory about mesothelial histogenesis of these tumors, and their Müllerian epithelium origin.

The rete ovary, the ovarian analogue of the rete testis, is a network of anastomosing tubules lined by flat, cuboidal, or columnar nonciliated cells with scanty eosinophilic or clear cytoplasm and located at the hilus of the ovary. Rete cysts are typically located in the ovarian hilus, most are unilocular, although occasionally they are multilocular. Rutgers and Scully described 16 cases of rete cysts. In this series, the ages of the patients ranged from 23 to 80 years (mean, 59 years); all but 4 were postmenopausal. The cysts had a mean diameter of 8.7 cm (range 1–4 cm). In addition to their hilar location, clues to the origin of the cysts are an irregular contour of their inner surface with small crevice-like outpouchings and a wall that often contains bundles of smooth muscle and hyperplastic hilus cells.

The rete epithelium is immunoreactive for CK, VIM, and desmoplakin, as well as low levels of ER and PR.

The most common clinical presentations of ovarian cysts are abdominal pain, nausea and vomiting, and a history of previous episodes of similar pain and low grade fever. Sudden pain and mild hyperthermia were the clinical manifestations in our case, but without ascites. After the evacuation of 6 l of serous fluid, the paraovarian location of the cyst was clearly distinguished from the surrounding organs and the ovary itself.

Because infundibulopelvic pedicle is longer in a child, torsion is a significant risk for larger cysts. Ovarian torsions are rare in the pediatric age group. In addition, atrophy of the ovary and other complications are common. In our case, the giant paraovarian cyst was accompanied by some complications including a double torsion, and initial hemorrhagic infarction of the ovary in question.

Conclusion

Our case report shows that the ovarian epithelial cystic neoplasm of paraovarian location may develop in children.
The cystic neoplasm of paraovarian location may reach a large size and cause numerous complications, the most frequent of which is adnexal torsion, which could be prevented by an early diagnosis and surgical treatment. These rare cystic lesions of paraovarian/mesoovarian location in children and their unclear histogenesis might be a histopathological diagnostic problem. According to our opinion, in making the diagnosis of these cystic neoplasms it is necessary to make (immuno)histochemical analyses on big series of tissue samples, as well as to know precise clinical data.

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