

Mini-Review

Management of Large Ovarian Neoplasms in Pediatric and Adolescent Females

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ABSTRACT

The overwhelming majority of ovarian cysts in pediatric and adolescent girls are physiologic; however, large simple and complex ovarian lesions often require surgical intervention due to the increased risk of neoplasia. In this review article, we discuss the preoperative evaluation and intraoperative management of large ovarian neoplasms. We review the current literature regarding long term ovarian function and fertility, rates of recurrence and residual disease, and novel surgical approaches. Managing large ovarian neoplasms in the pediatric and adolescent population requires careful preoperative and intraoperative care to optimally resect neoplasia while maximizing fertility and minimizing pain.

Key Words: Ovarian mass, Ovarian neoplasms, Ovarian cysts, Neonates, Children, Adolescents, Large pelvic mass

Introduction

Ovarian neoplasms may be cystic (simple with a solitary fluid collection), complex (cystic with solid components or septations), or solid. Ovarian cysts may be physiologic and associated with pregnancy, polycystic ovarian syndrome, and ovulation and associated with infections (tuboovarian abscesses). The majority of neoplasms in prepubertal and adolescent females are benign, the most common being mucinous cystadenomas, mature teratomas, and serous cystadenomas. The most common neoplasms identified in this age range are listed in [Table 1](#). In general, the larger and more complex an ovarian lesion is, the more likely that it is a neoplastic process necessitating surgical removal.¹ The purpose of this review is to provide the practicing provider with a review of the current literature and a suggested preoperative evaluation and surgical approach for management of large ovarian lesions suspicious for ovarian neoplasms. This review aims to address common questions and focus primarily on ovarian lesions larger than 8 cm in adolescents and larger than 5 cm in prepubertal females.

Initial Evaluation

Most young females are diagnosed with an ovarian cyst by ultrasonography (US), performed for abdominal pain or a

palpable pelvic mass or during diagnostic imaging being performed for other conditions. The presence of the ovarian lesion requires evaluation of whether it more likely to be physiologic, infectious, or neoplastic.

History

To begin the evaluation, a history should be obtained from the parents and the patient. A focused gestational history should include questions about maternal hypothyroidism, gestational diabetes, and whether an ovarian cyst was noted on antenatal US. For prepubertal patients, questions should include recent signs of puberty, including thelarche, growth spurt, vaginal discharge, or uterine bleeding, which may indicate physiologic ovarian cysts or an estradiol-secreting granulosa cell tumor. Symptoms of virilization including hirsutism, acne, deepening of the voice, and clitoromegaly may indicate testosterone excess from a Sertoli-Leydig tumor. In adolescents, a menstrual history including the timeline of pubertal development (thelarche and menarche), last menstrual period, and the presence of dysmenorrhea or irregular menses are helpful to assess the likelihood of physiologic cysts and endometriosis. The patient should be asked about the quality, duration, and severity of pain. An acute onset of pain or intermittent severe pain, especially associated with vomiting, is suspicious for ovarian torsion. Severe pain may also indicate a ruptured hemorrhagic cyst or neoplasm. Adolescent patients should be asked about contraceptive use and sexual activity privately and confidentially to assess for the likelihood of a pregnancy-associated cyst or tubo-ovarian abscess.

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Table 1
Common Benign and Malignant Ovarian Masses in Children and Adolescents

Benign tumors
<ul style="list-style-type: none"> • Mature teratoma • Epithelial tumors: serous and mucinous cystadenoma • Functional corpus luteum cyst • Endometrioma • Thecoma • Fibroma • Gonadoblastoma
Malignant tumors
<ul style="list-style-type: none"> • Immature teratoma • Dysgerminomas • Sex cord stromal tumor • Endodermal sinus tumor • Juvenile granulosa cell tumor • Embryonal carcinoma • Sertoli-Leydig cell tumor • Polyembryoma • Epithelial carcinoma • Choriocarcinoma

Physical Exam

Large ovarian masses may be palpable abdominally. Signs of peritoneal inflammation, including guarding, rigidity, and extreme tenderness may indicate ovarian torsion, rupture of a hemorrhagic cyst, ruptured teratoma, tuboovarian abscess, or ectopic pregnancy. The presence of breast budding prior to age 7 or 8 may reflect prematurely elevated estradiol levels, either from central stimulation from gonadotropins or from the presence of an ovarian tumor or a physiologic cyst. Growth charts can be used to indicate whether there has been a recent growth spurt indicating puberty. The presence of clitoromegaly may reflect elevated testosterone from a Sertoli-Leydig tumor. In prepubertal patients, vaginal examination can be extremely painful due to the small hymenal opening. Rectal examination in prepubertal and young adolescents is typically much less painful than a vaginal examination and should be considered an alternative to digital and speculum vaginal examination. Malignant tumors are more likely than benign tumors to be bilateral, solid, fixed, irregular, and associated with ascites. In sexually active and older adolescents, speculum examination can be performed to assess for signs of infection including cervical mucopus. Bimanual examination using a single digit can assess for cervical motion tenderness and the size and mobility of the adnexa.

Laboratory Tests

The most important laboratory test which should be performed in any adolescent patient with an ovarian lesion is a urine β -human chorionic gonadotropin test (β -hCG) to rule out pregnancy. In addition, testing for sexually transmitted infection is especially important in a sexually active adolescent who may have pelvic inflammatory disease, hydrosalpinx, or a tuboovarian abscess. When the ovarian lesion appears to be large and neoplastic, the decision must be made whether to pursue laboratory testing for tumor markers. Though not validated, tumor markers commonly assessed in the pediatric population include alpha fetoprotein (AFP), β -hCG, lactate dehydrogenase (LDH), and

inhibin A and B.² Tumor markers may not be elevated with early staged ovarian neoplasms; however, they can be helpful in ovarian tumors where suspicion for malignancy is elevated, to facilitate preoperative planning for a staging procedure and to follow the patient for evidence of remission postoperatively. Germ cell tumors, such as dysgerminoma, choriocarcinoma, embryonal, yolk sac tumor, immature teratomas, and mixed germ cell tumors may have elevated levels of AFP, β -hCG, and LDH. Granulosa cell tumors are associated with elevated estradiol and inhibin levels. Thecomas are associated with elevated estradiol and testosterone levels. Sertoli-Leydig cell tumors are associated with elevated testosterone and inhibin levels. CA 125 levels can be elevated with epithelial ovarian carcinoma and immature teratomas; however, caution should be used with this tumor marker as it can be elevated with a variety of benign reasons in postpubertal patients, including endometriosis, torsion, and menses.^{3,4} Given the possibility of a false positive elevated tumor markers and the cost associated with testing, we recommend tumor markers when imaging modalities indicate a higher risk of malignancy. This includes imaging with a solid component greater than 2 cm, ascites, thick septations, and evidence of precocious puberty or virilization. If tumor marker levels have not been obtained preoperatively and a malignancy is identified intraoperatively or postoperatively, levels may be drawn at the time of diagnosis and then followed to permit postoperative surveillance.

Imaging

Ultrasonography

Ultrasonography is the imaging modality of choice to assess the ovarian volume and presence of an ovarian lesion. Typically, in prepubertal and young adolescent patients, transabdominal US is recommended over transvaginal US. Ultrasonography can assess if the mass is smooth walled, unilocular or multi-loculated, unilateral or bilateral, and assess whether it contains solid components. Indicators of malignancy include solid components greater than 2 cm, thick septations, and papillary projections.⁵ Hemorrhagic cysts typically will have echogenic material without Doppler flow to solid components. There may be a reticular or lacy appearance. Granulosa cell tumors appear sponge-like and there may be endometrial thickening in response to estradiol production. Cystadenomas are cystic masses without enhancing septations or solid components. Mucinous tumors may appear to have different densities of fluid within the cyst due to layering of mucin. Mature teratomas are cystic and may have solid components with calcifications, shadowing echogenic material of sebaceous material, and fat-fluid levels.⁶

Computerized Tomography (CT) and Magnetic Resonance Imaging (MRI)

When children and adolescents present with right lower quadrant pain or back pain, they may undergo an abdominal pelvic CT which can identify an ovarian lesion. In

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