



PEDIATRIC SURGERY Update ©

Vol. 39 No. 06 DECEMBER 2012

Ovarian Epithelial Tumors

Ovarian epithelial tumors are the second most common ovarian neoplasm in children after germ cell tumors. They arise from the epithelial surface of the ovary representing between 15-20% of all ovarian neoplasms in adolescent females. The histologic subtype of epithelial ovarian tumors in children includes serous and mucinous tumors. Adenocarcinoma of the ovary in children is an extremely rare entity. Epithelial tumors are further characterized as benign, malignant or of low malignant potential (borderline tumors). Mean age at diagnosis is 13 years. Most children present with history of abdominal pain or an asymptomatic pelvic mass. The most common type of epithelial neoplasm is the benign serous cystadenoma. Borderline epithelial ovarian tumors are defined as epithelial neoplasms of varying level of nuclear atypia that lacks stromal invasion of the ovary. They are more common in the pediatric age. Preoperative work-up includes imaging (Ultrasound, CT and MRI) and tumor markers (AFP, HCG, LDH, CA-125). Epithelial ovarian tumors frequently cause elevation of the serum level of the CA-125 tumor antigen. During laparotomy strict staging criteria must be met such as collection of ascites/washing, peritoneal surface examination, palpable lymph nodes removal, omentectomy if involved, suspicious contralateral ovarian biopsy and complete resection of the involved ovary intact with sparing of the fallopian tube if not involved (ovarian cystectomy vs. salpingoophorectomy). Fertility preserving surgery is recommended to preserve ovarian function. There is no benefit in removing clinically uninvolved tissue like uterus or contralateral ovary. With mucinous epithelial tumors the appendix should be removed to avoid synchronous lesions.

References:

- 1- Morowitz M, Huff D, von Allmen D: Epithelial ovarian tumors in children: a retrospective analysis. *J Pediatr Surg.* 38(3):331-5, 2003
- 2- Young JL Jr, Cheng Wu X, Roffers SD, Howe HL, Correa C, Weinstein R: Ovarian cancer in children and young adults in the United States, 1992-1997. *Cancer.* 2003 May 15;97(10 Suppl):2694-700, 2003
- 3- Borgfeldt C, et al: Fertility-sparing surgery and outcome in fertile women with ovarian borderline tumors and epithelial invasive ovarian cancer. *Eur J Obstet Gynecol Reprod Biol.* 134(1):110-4, 2007
- 4- Stankovic Z, Djuricic S, Djukic M, Jovanovic D, Vasiljevic M: Epithelial ovarian tumors and CA125 in premenarchal girls. *Eur J Gynaecol Oncol.* 2006;27(6):597-9, 2006
- 5- Aggarwal A, Lucco KL, Lacy J, Kives S, Gerstle JT, Allen L: Ovarian epithelial tumors of low malignant potential: a case series of 5 adolescent patients. *J Pediatr Surg.* ;44(10):2023-7, 2009
- 6- Song T, Choi CH, Lee YY, Kim TJ, Lee JW, Bae DS, Kim BG: Pediatric borderline ovarian tumors: a retrospective analysis. *J Pediatr Surg.* 45(10):1955-60, 2010
- 7- Grapsa D, Kairi-Vassilatou E, Kleantithis C, Dastamani C, Fillipidou A, Kondi-Pafiti A: Epithelial ovarian tumors in adolescents: a retrospective pathologic study and a critical review of the literature. *J Pediatr*

Ovarian Lymphoma

Primary ovarian non-Hodgkin's lymphomas are unusual and accounts for 0.5% of all non-Hodgkin's lymphomas and 1.5% of all ovarian malignancies. The diffuse, large B-cell lymphoma is the most common type of primary ovarian non-Hodgkin's lymphoma followed by non-endemic Burkitt's lymphoma. It has been suggested the tumor originates from lymphocytes in the ovaries, surrounding blood vessels at the hilum and related to the corpus luteum. Most patient with ovarian lymphoma present with symptoms of abdominal pain, pelvic mass, ascites and elevation of serum CA-125 antigen. This lymphoma has the fastest doubling time of any tumor and it can be as low as 24 hrs. Presence of positive staining for leukocyte common antigen (LCA) in the histological specimen distinguishes malignant lymphoma from nonlymphoid neoplasm. The MRI findings include solid mass with low signal intensity on T1 and mildly high signal intensity in T2. CT-Scan is utilized for staging the disease in the chest, abdomen and pelvis. Bone marrow biopsy is also mandatory for staging. B-cell lymphoma is positive for CD20 and BCL-6. Children with localized disease to one ovary are best managed with unilateral surgical resection (salpingo-oophorectomy) followed by systemic chemotherapy. Protocol of chemotherapy used in diffuse large B-cell histology is the standard CHOP regimen. Metastasis to the ovaries from another primary site lymphoma should also be considered and they can be solid, cystic and bilateral. Primary ovarian lymphoma has a good prognosis.

References:

- 1- Ambulkar I, Nair R: Primary ovarian lymphoma: report of cases and review of literature. *Leuk Lymphoma*. 44(5):825-7, 2003
- 2- Crawshaw J, Sohaib SA, Wotherspoon A, Shepherd JH: Primary non-Hodgkin's lymphoma of the ovaries: imaging findings. *Br J Radiol*. 80(956):e155-8, 2007
- 3- Vang R, Medeiros LJ, Warnke RA, Higgins JP, Deavers MT: Ovarian non-Hodgkin's lymphoma: a clinicopathologic study of eight primary cases. *Mod Pathol*. 14(11):1093-9, 2001
- 4- Elharroudi T, Ismaili N, Errihani H, Jalil A: Primary lymphoma of the ovary. *J Cancer Res Ther*. 4(4):195-6, 2008
- 5- Arnogiannaki N, Grigoriadis C, Zygouris D, Androutsopoulos G, Derdelis G, Terzakis E: Primary ovarian non-Hodgkin's lymphoma. *Eur J Gynaecol Oncol*. 32(4):441-2, 2011
- 6- Chakrabarti B, Bhaduri B, Barik S, Gupta D, Chakravorty S: Primary ovarian lymphoma in a child. *J Indian Med Assoc*. 109(9):679-80, 2011

Diverticulitis in Children

The most common diverticulum in children causing surgical problems is the Meckel's diverticulum. In very rare occasion the pediatric patient can develop diverticular disease of the colon similar to that occurring in the adult. Such diverticular disease can lead to colonic diverticulitis. Genetic disorders in pediatric patients can lead to weakening of the colonic wall and subsequent development of diverticular disease. Diseases with an early predilection for diverticulitis include cystic fibrosis, Ehlers-Danlos syndrome, Marfan syndrome and William-Beuren syndrome. Some of these syndromes have genetic

mutations that alter the collagen or elastin levels within tissue creating diverticula in early age. Children with the above syndromes and symptoms of constipation or relapsing or chronic abdominal pain should undergo colonoscopy or barium study to identify such diverticular disease. Stool softener and high-fiber diet can help in the prevention of early complicated diverticular disease.

References:

- 1- Benya EC, Nussbaum-Blask AR, Selby DM: Colonic diverticulitis causing partial bowel obstruction in a child with cystic fibrosis. *Pediatr Radiol.* 27(12):918-9, 1997
- 2- Shin JH, Son BH, Kim H: Clinically distinguishing between appendicitis and right-sided colonic diverticulitis at initial presentation. *Yonsei Med J.* 30;48(3):511-6, 2007
- 3- Bogue CO, Mann EH: Imaging findings in right-sided diverticulitis in a child. *Pediatr Radiol.* 38(10):1125-7, 2008
- 4- Garcia MA, Kling KM, Newbury RO, Huang JS: Complicated diverticular disease in a child with williams syndrome. *J Pediatr Gastroenterol Nutr.* 48(2):233-6, 2009
- 5- Santin BJ, Prasad V, Caniano DA: Colonic diverticulitis in adolescents: an index case and associated syndromes. *Pediatr Surg Int.* 25(10):901-5, 2009
- 6-Ignacio RC Jr, Klapheke WP, Stephen T, Bond S: Diverticulitis in a child with Williams syndrome: a case report and review of the literature. *J Pediatr Surg.* 47(9):e33-5, 2012

* Edited by: Humberto Lugo-Vicente, MD, FACS, FAAP

Professor of Pediatric Surgery, University of Puerto Rico - School of Medicine,
Rio Piedras, Puerto Rico. Director - Pediatric Surgery, San Jorge Childrens Hospital.
Address: P.O. Box 10426, Caparra Heights Station, San Juan, Puerto Rico USA 00922-0426.
Tel (787)-786-3495 Fax (787)-720-6103 E-mail: titolugo@coqui.net
Internet: <http://home.coqui.net/titolugo>

© PSU 1993-2012
ISSN 1089-7739