Published in final edited form as:

J Pediatr Adolesc Gynecol. 2010 February ; 23(1): e47. doi:10.1016/j.jpag.2009.04.002.

Premenarchal Ovarian Torsion and Elevated CA-125

Jenifer D. McCarthy, MD¹, Kimberly M Erickson, MD², Yolanda R. Smith, MD¹, and Elisabeth H. Quint, MD¹

¹Department of Obstetrics and Gynecology, University of Michigan, Ann Arbor, MI 48109

²Department of Pediatric Surgery, University of Michigan, Ann Arbor, MI 48109

Abstract

Background—Ovarian tumors are the most common gynecologic malignancy occurring in childhood, with germ cell tumors being most frequent. This contrasts with adults where epithelial tumors account for most ovarian neoplasms. Tumor markers are an integral part of the work up and may guide management.

Case—A 6 year old girl with a persistent adnexal mass was found to have a highly elevated CA-125. Other tumor markers were normal. Laparoscopy revealed an enlarged, adherent ovary. A minilaparotomy revealed an ovary filled with necrotic material. This necrotic material was excised and the ovary was spared. The pathology was consistent with necrosis. Follow up ultrasound and CA-125 were normal.

Summary and Conclusions—This case demonstrates for the first time the association of an elevated CA-125 and ovarian torsion in a pediatric patient. This benign finding allowed attempting a conservative ovarian-sparing approach during the surgery even in the presence of a highly elevated CA-125. However, in general, for children CA-125 is of limited utility, as it will not affect the indication for surgical exploration of persistent masses and elevations in CA-125 may discourage ovarian conservation.

Introduction

Ovarian cancer is the most common gynecologic cancer occurring in childhood.¹ Malignant ovarian tumors are relatively rare in children, accounting for only 0.9% of malignancies in this age group.² However, in premenarchal girls, up to 40% of ovarian neoplasms are malignant.³ Therefore, malignancy should be excluded in any adnexal mass found in this age group. In evaluating a pre-menarchal adnexal mass, numerous tumor markers are available for clinical use. Because germ cell tumors are the most common tumors seen in childhood, tumor markers for those masses are always indicated. Other tumor markers may also be helpful in narrowing the differential diagnosis. One of these is CA-125, which is an indicator for ovarian epithelial malignancies but in adults can also be associated with other intra-abdominal pathology such as adenomyosis, uterine fibroids, pelvic inflammatory disease, pregnancy, menstruation, bowel

© 2009 North American Society for Pediatric and Adolescent Gynecology. Published by Elsevier Inc. All rights reserved.

Correspondence: Jenifer D. McCarthy, M.D., 1500 East Medical Center Drive, L4000 Women's Hospital Box 0276, Ann Arbor, MI 48109, Phone: 734-678-5685, Fax: 734-936-8617, e-mail: jmccarth@umich.edu.

Publisher's Disclaimer: This is a PDF file of an unedited manuscript that has been accepted for publication. As a service to our customers we are providing this early version of the manuscript. The manuscript will undergo copyediting, typesetting, and review of the resulting proof before it is published in its final citable form. Please note that during the production process errors may be discovered which could affect the content, and all legal disclaimers that apply to the journal pertain.

Disclosure: None of the authors have any actual or potential conflict of interest including any financial, personal or other relationships with other people or organizations within three (3) years of beginning the work submitted that could inappropriately influence (bias) their work.

disorders, endometriosis and ovarian torsion.⁴ Here we report the first pediatric case of a highly elevated CA-125 associated with an ovarian torsion.

Case

A previously healthy six year old girl presented to her pediatrician with complaints of episodic left lower quadrant abdominal pain for one week with radiation to the left hip. She denied fever and her bowel movements were regular. On exam, her abdomen was soft, non distended and mildly tender without rebound or guarding. Rapid streptococcus test was positive and she was treated with azithromycin. Her abdominal pain was attributed to her streptococcus pharyngitis infection and her symptoms improved. One week later, she developed a recurrence of her abdominal pain and evaluation with a pelvic ultrasound demonstrated a $5 \times 5 \times 4$ cm mass suspected to be a dermoid cyst. Her pain resolved completely during the evaluation.

The patient presented without any symptoms to pediatric gynecology and a repeat transabdominal ultrasound was performed, which demonstrated a right ovary measuring $2.0 \times 1.2 \times 2.2$ cm with normal flow. The left ovary measured $1.2 \times 1.5 \times 0.7$ cm, no flow was mentioned. The uterus measured 3.5×0.6 cm. There was a heterogeneous, hypoechoic, well-circumscribed solid mass posterior to the uterus and abutting both ovaries, measuring $4.7 \times 3.2 \times 2.4$ cm. There was no internal blood flow or surrounding free fluid identified. The origin of the mass was unclear. MRI demonstrated a $4.5 \times 4.0 \times 3.0$ cm ovoid, heterogeneous mass in the lower pelvis with minimal peripheral enhancement. This mass was questionably attached to the left ovary and signal characteristics suggested either a torsed ovary or a hemorrhagic cyst. Dermoid was felt to be less likely given the absence of fat signal. No pelvic lymphadenopathy was evident. HCG, LDH, AFP, and CEA were within normal limits. CA-125 was elevated at 501.5 units/mL (normal <35 in adults).

The combination of the persistence of the mass and the elevated CA-125 level was concerning for possible malignancy and the patient was taken to the operating room for diagnostic laparoscopy with the gynecology and pediatric surgery services. A normal-appearing right ovary was noted [Figure 1]. The left ovary and tube were enlarged and closely adherent to the rectum and posterior surface of the uterus, limiting visibility of the vascular pedicle [Figure 2]. The procedure was converted to a mini-laparotomy and the ovary was delivered into the field, allowing a frozen section to be performed without the risk of peritoneal contamination. Examination of the ovary during laparotomy did not reveal a currently torsed pedicle. Since no tissue plane could be identified to accommodate a cystectomy, the ovary was incised. Necrotic material was identified within the ovary [Figure 3]. A biopsy of the ovary as well as this necrotic material was sent to pathology and necrotic tissue with no evidence of malignancy was diagnosed on frozen section. The necrotic tissue was excised and the normal appearing cortex closed with suture. Final pathologic diagnosis demonstrated ovarian stroma and necrotic cellular debris. Due to the small size of the biopsy submitted, the pathologist was unable to comment on the potential for cortex viability.

While we were unable to confirm ovarian torsion during laparoscopy due to the extensive adhesions present or after externalizing the ovary, the pathologic results were consistent with some necrosis after ovarian torsion. The resolution of the patient's pain during her initial ultrasonographic evaluation suggest either a torsion/detorsion or a torsion with subsequent necrosis of the ovary.

Follow up transabdominal ultrasound three months later demonstrated a normal prepubertal uterus and ovaries. Follow up CA-125 was normal at 14.0 and 13.0 units/mL at 3 and 9 months post-procedure, respectively. Function of this ovary remains unknown, as puberty has not initiated in this patient.

Summary and Conclusion

Here we present a case of a premenarchal girl in whom an elevated CA-125 reflected ovarian torsion. This case demonstrates several interesting issues in the management of pediatric ovarian masses, including conservative surgery, appropriate tumor markers and elevation of CA-125 in the presence of pediatric ovarian torsion.

In a pediatric patient with a pelvic mass, gynecologic malignancy must first be ruled out. In children, oophorectomy is the most common ovarian surgery, even for benign conditions such as ovarian cysts and torsion.³ Since removal of the entire ovary has significant implications for future fertility, every effort should be made to avoid unnecessary oophorectomy. In this case, rather than removing the entire ovary, a biopsy was performed in an open fashion to prevent intra-abdominal spillage and obtain a frozen section. Once malignancy had been ruled out, the necrotic component within the ovary was excised and the ovarian cortex was preserved. Long term follow up is necessary to assess ovarian function.

Multiple serum markers for ovarian tumors are available including HCG, LDH, AFP, CEA and CA-125. Elevated AFP, LDH and HCG are associated with germ cell tumors, while CA-125 can indicate an ovarian tumor of epithelial origin. CA-125 is perhaps the most widely studied marker for ovarian cancer in adults and has sensitivities for stage I and II disease of 50 and 90%, respectively.⁵ However, the specificity of CA-125 is limited. In addition to ovarian malignancy, other gynecologic states such as adenomyosis, uterine fibroids, pelvic inflammatory disease, pregnancy, menstruation, endometriosis and ovarian torsion can cause elevations in serum CA-125.⁴ In a premenarchal girl, many of the common benign etiologies of elevated CA-125 are absent and therefore, an elevated CA-125 and a persistent adnexal mass raises concern for malignancy. Two thirds of ovarian malignancies in girls under 18 years of age are of germ cell origin.¹ Epithelial tumors, which account for most ovarian neoplasms in adults, are rare in children.⁶ In general, for children with adnexal masses, CA-125 should not be routinely obtained, as it will not affect the indication for surgical exploration of persistent masses and elevations in CA-125 may discourage ovarian conservation even in benign conditions such as ovarian torsion.

Ovarian torsion is most commonly seen in menstruating women, but cases have been described in neonates and premenarchal girls.⁷ In a prior study, torsion was the most common diagnosis leading to ovarian surgery in premenarchal girls. Misdiagnosis of this condition can result in unnecessary oophorectomy. The predominant cause of ovarian torsion in children is thought to be functional cysts and benign neoplasms. Only two cases of malignancy have been described associated with ovarian torsion.⁷ Detorsion, with or without concomitant oophoropexy has been used successfully to preserve ovarian function in both adults and children, even when the ovary appears necrotic on gross examination.⁸ CA-125 can be highly elevated with ovarian torsion. In one report of two adult torsion patients with endometriosis the CA-125 value rose from the 40's to the 500–600 range.⁴ Expected CA-125 levels in children with torsion have not been reported.

In conclusion this case represents the first reported association of an elevated CA-125 and ovarian torsion in a pediatric patient. In addition, this benign finding allowed attempting a conservative ovarian-sparing approach during the surgery even in the presence of a highly elevated CA-125.

References

1. You W, Dainty LA, Rose GS, et al. Gynecologic malignancies in women aged less than 25 years. *Obstet Gynecol* 2005;105(6):1405–1409. [PubMed: 15932836]

2. Templeman C, Fallat ME, Blinchevsky A, et al. Noninflammatory ovarian masses in girls and young women. *Obstet Gynecol* 2000;96(2):229–233. [PubMed: 10908768]
3. Quint EH, Smith YR. Ovarian Surgery in Premenarchal Girls. *J Pediatr Adolesc Gynecol* 1999;12:27. [PubMed: 9929837]
4. Ghaemmaghami F, Karimi Zarchi M, Hamed B. High levels of CA125 [over 1,000 IU/ml] in patients with gynecologic disease and no malignant conditions: three cases and literature review. *Arch Gynecol Obstet* 2007;276(5):559–561. [PubMed: 17497163]
5. Carlson KJ, Skates SJ, Singer DE. Screening for ovarian cancer. *Ann Intern Med* 1994;121(2):124–132. [PubMed: 8017726]
6. Brown MF, Hebra A, McGeehin K, et al. Ovarian masses in children: a review of 91 cases of malignant and benign masses. *J Pediatr Surg* 1993;28(7):930–933. [PubMed: 8229571]
7. Cass DL. Ovarian torsion. *Seminars in Pediatric Surgery* 2005;14(2):86–92. [PubMed: 15846564]
8. Eckler K, Laufer MR, Perlman SE. Conservative management of bilateral asynchronous adnexal torsion with necrosis in a prepubescent girl. *J Pediatr Surg* 2000;35(8):1248–1251.

Figure 1.
Laparoscopic overview.

Figure 2.
Ovaries after lysis of adhesions.

Figure 3.
Laparotomy: initial view (left) and open ovary (right).