Mini-Review

The Auto-Amputated Adnexa: A Review of Findings in a Pediatric Population

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A B S T R A C T

Study Objective: To quantify our experience and that of the literature with diagnosis and management of the auto-amputated adnexa in a pediatric population.
Design: Case series and literature review.
Setting: Tertiary care medical center.
Participants: Case series of pediatric patients (<18 years of age) with surgically documented adnexal auto-amputation collected from our medical center and the literature.
Interventions: None.
Main Outcome Measure: Auto-amputated adnexa.
Results: In addition to the 3 cases discussed from our institution, 91 cases of auto-amputated adnexa were identified in the literature dating back to 1943, for a total of 94 cases. Forty-nine percent (46/94) of the cases involved girls in a pediatric population (<18 years of age). Of these, the majority ($n = 26$) were identified in a subgroup of girls who were diagnosed with an adnexal cyst by antenatal ultrasound. Most of these neonates were asymptomatic at birth or had a palpable abdominal mass ($n = 6$) and at the time of surgical exploration were found to have an auto-amputated adnexa. 34 out of 46 cases were analyzed in detail. The right adnexa were involved in 56% of the cases. The most common presenting complaint verbalized by the older girls was pain; however, 8 cases were identified in asymptomatic girls undergoing unrelated diagnostic testing.
Conclusion: The auto-amputated adnexa is a rare finding in the pediatric population, but it must be considered as a possible explanation for the incidental finding of absence of the fallopian tube or ovary in the subgroup of patients who undergo surgery for any reason. Patients with an antecedent history of pelvic pain either chronic or intermittent in nature may be diagnosed with torsion or less frequently auto-amputation of the adnexa. A fetal "pelvic mass" or "ovarian cyst" may predispose the adnexa to torsion and subsequent auto-amputation either in-utero or post-delivery. Many of these antenatally diagnosed cysts and even subsequent auto-amputations are completely asymptomatic, however, and do not compromise fertility assuming the contralateral adnexa are normal. Thus expectant management is appropriate for small (less than 4 cm), asymptomatic simple cysts and even suspected auto-amputated adnexa in an asymptomatic patient.
Key Words: Auto-amputation ovary, Adnexal torsion, Ovarian torsion, Ovarian remnant

Introduction

Auto-amputation of the adnexa and/or the finding of an absent fallopian tube or ovary has been a unique finding reported in the literature with one report suggesting an incidence of 1 in 11,421.[1] The etiology is thought to be acquired and not congenital as these 2 structures have different embryologic origins.[2] The most accepted theory to explain the etiology of auto-amputation is chronic adnexal torsion and subsequent devascularization. The auto-amputated structure then may reattach to another surface or often times, become a free-floating, possibly calcified intraperitoneal object. While chronic torsion of an adnexal mass, presenting as long standing intermittent or chronic pelvic and lower abdominal pain, has been reported as a cause of acquired auto-amputation in children and adults,[3] there are several reports of auto-amputation that occur in utero, in neonates, and in asymptomatic young women.[2–4]

Fetal cysts are increasingly being reported, most likely due to ease of detection with increasing use of today's high resolution prenatal ultrasounds. The fetal ovary contains follicles which appear as "cysts" in high-resolution sonographic studies.[5,6] These follicles, which can range in size from 1 mm to 10 mm, are normal findings and are associated with stimulation from the maternal hormones.[7] These asymptomatic follicles undergo resolution by atresia and should be considered normal findings in infants and children. The management of fetal or persistent neonatal ovarian cysts is controversial. Infants with simple cysts less than 4 cm in diameter should be initially managed conservatively and followed expectantly by post-natal sonography, as the majority will resolve spontaneously.[8] Larger cysts may require surgical intervention, especially if the lesions convert from simple to complex on ultrasound imaging, suggesting hemorrhage or torsion; or if the neonate is symptomatic (i.e., poor feeding, emesis).[8]

Torsion and the ultimate loss of fallopian tube and/or ovarian viability both in-utero and peri-natally have been

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well documented.[9] Twenty-four cases of auto-amputation of the ovary and/or fallopian tube in the neonate, associated with prenatally diagnosed ovarian cysts, have been reported.[10,11] Auto-amputation refers to a situation in which a tubo-ovarian remnant is found free- floating in the pelvis[2,5,12,13]; or in which torsed adnexa detach from their normal anatomic position and become adherent to the pelvic wall or other pelvic viscera.[3,14] If there is a remnant present, it often appears calcified on imaging prior to surgical exploration. Many times the remnant is not identified, in which case it is thought to be resorbed.

In this review, 3 cases from our institution will be presented along with a series of other young girls with documented auto-amputation collected from the medical literature, in order to determine the most common presenting symptoms and discuss if any preventative measures could be taken.

Materials and Methods

A PubMed search was performed using the search terms “auto-amputation,” “torsion,” “ovarian remnant,” and a search of collected literature was performed to identify additional cases in order to provide a comprehensive review. Cases were limited to the English language and to those affecting women under the age of 18. In order for the case to be included, the following clinical information was required: (1) age of diagnosis, (2) surgical intervention and intraoperative findings, and (3) pathologic findings. Two authors reported cases and summarized the findings in aggregate.[15,16] These aggregate cases (12 patients in total) were included in the total number of cases of auto-amputated adnexa but were omitted when examining each case individually because not all clinical information was presented in the case series. Three cases from our practice were identified and included in the evaluation. All private health information for the patients was collected and compiled according to our Institutional Review Board Protocol for Human Studies.

Case Presentations

Case 1: Autoamputation of Left Adnexa with Free-floating Mass in Cul-de-sac

A pre-menarchal 9-year-old female presented to her primary provider complaining of increased lower abdominal pain over the course of 3 months. Her abdomen was soft and diffusely tender but there were no peritoneal signs. An abdominal-pelvic computed tomography (CT) scan with contrast was performed which was reported as normal, with the exception of a 2.0 cm $\times$ 1.0 cm calcified pelvic mass suspicious for a dermoid that was noted adjacent to the uterus and right ovary. The left ovary was not seen in this study. A trans-abdominal ultrasound showed similar findings.

The patient was then referred to our center for evaluation. Ovarian cancer tumor markers (alpha feto-protein, human chorionic gonadotropin, lactate dehydrogenase, and inhibin A and B levels) were normal. A follow-up trans-abdominal sonogram was performed which again showed a normal right ovary and an absent left ovary. The previously described 2 $\times$ 1 cm calcified pelvic mass was not visualized. The patient's abdominal and pelvic pain remained intermittent throughout this time and after consultation with a pediatric surgeon, the patient was scheduled for a diagnostic laparoscopy. Findings included a prepubertal and midline uterus. The right ovary was normal and the right fallopian tube and fimbria were normal. The left tube truncated about 2-3 cm distal to the cornua; no fimbria were seen. A remnant utero-ovarian ligament was identified but no ovarian tissue was grossly visualized. The infundibulopelvic ligament was visualized and terminated blindly as well. The appendix and bowel were normal. After reflecting the small bowel, a 2-cm free floating pelvic mass was noted in the posterior cul-de-sac. It appeared consistent with the mass described on CT scan and was noted to be neither inflamed nor necrotic (Fig. 1). The mass was brown, hard, and smooth with purple speckles. Pathologic assessment of the mass was consistent with a calcified nodule within an ovarian remnant. There was no evidence of malignancy. The patient's intraoperative and postoperative course was unremarkable and she went home on postoperative day 1. The patient's pain subsequently resolved.

Case 2: Autoamputation of Left Adnexa with Adherence and Implantation at Transverse Colon

A 17-month-old female infant presented to the emergency room after a 1-week history of constipation and non-bloody, non-bilious emesis. The infant's neonatal, medical and surgical history had been unremarkable up to this time. On abdominal CT, a 6.4 $\times$ 9.3 $\times$ 10.2 cm septated, cystic mass was noted just inferior to the liver in the right upper quadrant. The mass appeared separate from the biliary system and both kidneys appeared normal without renal pelvic dilation. The ovaries were not identified, and the rest of the pelvis was unremarkable. The mass was palpable on physical examination.

The patient was taken to the operating room for an exploratory laparotomy. A right upper quadrant abdominal mass measuring 10 cm in size was encountered upon entering the peritoneal cavity. Initially it was thought to be an omental cyst given its location proximal to the transverse colon (Fig. 2). Once the structure was de-torsed, it was

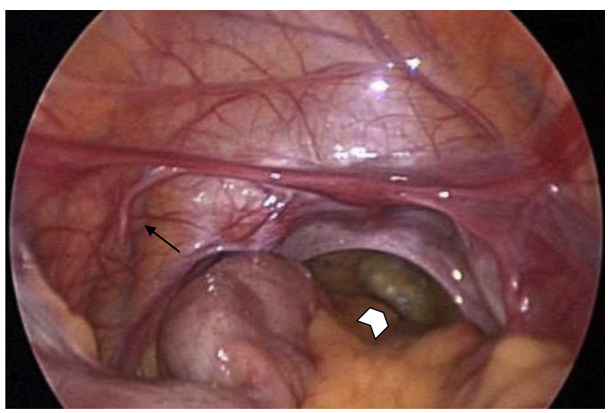

Fig. 1. Panoramic view of pelvis with ovarian remnant floating in posterior cul-de-sac (arrow = remnant of left tube, arrowhead = ovarian remnant).