Daughter cyst sign

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ABSTRACT

The daughter cyst sign is a specific indicator of an uncomplicated ovarian cyst and pathologically represents a stimulated ovarian follicle. This finding must be differentiated from an ectopic pregnancy in a patient who has the potential to become pregnant. We report an uncomplicated ovarian cyst in a 3-year-old female with McCune-Albright syndrome and precocious puberty mimicking an ectopic pregnancy.

CASE REPORT

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A 3-year-old female presented for a screening ultrasound of the kidneys and a right-sided cystic ovarian lesion was demonstrated with an internal cystic component. The patient is an ex-35 week premature female, with a prenatal diagnosis of a dysplastic left kidney, born to a mother with pre-eclampsia with a past medical history also significant for precocious puberty, café-au-lait spots, and fibrous dysplasia of McCune-Albright syndrome as well as a congenital nevus over the left side of her face and neck. The patient's bone age was approximately 11 months older than her chronologic age.

The patient was without abdominal symptoms at the time of our current screening ultrasound but had a history of vaginal bleeding, gynecomastia and body odor, which began one month following her second birthday. The left dysplastic kidney was not visualized. A right sided cystic ovarian lesion was demonstrated that measured 3.3 cm x 3.5 cm x 3.4 cm with internal peripheral cystic component measuring 7.0 mm x 6.5 mm x 7.0 mm and without internal color Doppler flow (Figures 1, 2, 3). This lesion resembled an ectopic pregnancy and as such, a beta human chorionic gonadotropin level was drawn. It was reported to be less than 5 mIU/dL, which was negative for pregnancy and the patient was followed conservatively.

DISCUSSION

The differential diagnosis for a lower abdominal mass in a female child includes benign, malignant, and infectious etiologies of the ovaries, fallopian tubes, and uterus. Other differential considerations consist of an enteric duplication cyst, meconium pseudocyst, lymphangioma, anterior meningocele, mesenteric cyst, urachal cyst, and other renal cystic lesions [1,2]. Of note, the most common abdominal mass in the female newborn is ovarian in etiology [3] with malignant neoplasm reported in up to 3.5% of neonatal ovarian masses [4]. Obtaining the correct diagnosis is important as the clinical management varies widely depending on the etiology of the mass.

Cystic lesions of the ovary are not uncommon in the prepubescent population. In utero, there is normal stimulation of ovarian follicles by maternal hormones, resulting in ovarian cysts, which then involute when maternal hormonal stimulation decreases [5,6]. An ovarian follicle is a physiological maturing cyst of less than 20mm in diameter in the postnatal age group [7]. In one series, approximately 10% of healthy girls without signs of thelarche demonstrated ovarian follicles less than or equal to 10 mm in size [8].

Cystic ovarian lesions are commonly found in children with precocious puberty, resembling normal findings in post pubescent ovaries. There are two types of precocious puberty: central and peripheral. Central precocious puberty, known as gonadotropin-dependent, is idiopathic in two-thirds of cases while being caused by central nervous system conditions in the other cases. Peripheral precocious puberty is gonadotropinindependent. Autonomous ovarian follicles are the most common cause of peripheral precocious puberty with estrogensecreting neoplasms much less common [9,10].

Ovarian cysts in precocious puberty have been reported to range in size from 25 mm to 65 mm with normal contralateral ovarian size for the patient's chronological age. In addition, these patients demonstrated an enlarged, stimulated uterus measuring more than 35 mm in length [9]. Ovarian volumes in patients with precocious puberty have been reported as 4.1 to 4.6 cm3, while less than 1 cm3 in other patients [11]. In the same series, ovarian cysts greater than 9 mm were found in peripheral precocious puberty. As such, cysts larger than 10 mm may be associated with hormone production and precocious puberty [12] with cysts greater than 10 mm not observed in one series of healthy girls between the ages of 1 and 13 [8].

Peripheral precocious puberty may be associated with cafe-au-lait spots and fibrous dysplasia, which is termed McCune-Albright syndrome. It is caused by a mutation in the GNAS1 gene resulting in activation of adenyl cyclase [12]. Ovarian cysts are common in McCune-Albright syndrome [13] with ultrasound typically demonstrating a unilateral follicular ovarian cyst [9,14,15].

To aid in diagnosing cystic masses in neonates, infants, and young children, the daughter cyst sign has been described as a specific indicator of an uncomplicated ovarian cyst. The reported sensitivity is 82%, specificity 100%, and positive predictive value 100% for predicting an ovarian etiology when the daughter cyst sign was present [2].

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The daughter cyst sign describes a peripherally based cyst within a cyst. In one study, the size of the daughter cysts ranged from 2 mm to 8 mm with the number of daughter cysts ranging from 1 to 3 [2]. Quarello et al. identified the daughter cyst sign prenatally with daughter cysts measuring up to 18 mm in diameter [16]. Pathologically, the daughter cysts corresponded to ovarian follicles [2].

To the authors' knowledge, a daughter cyst sign has yet to be reported in a case of precocious puberty with only two cases reported in a young child [2]. Unique to precocious puberty is the real possibility of pregnancy and excluding an ectopic pregnancy is an important clinical consideration in a patient with an adnexal cystic mass.

Similar in appearance to a daughter cyst is a yolk sac within a gestational sac of pregnancy, which is also a peripheral cyst within a cyst. Pregnancy in children with precocious puberty has been reported in the literature [17]. Typically, ectopic pregnancies are tubal in location with ovarian-based ectopic pregnancy much less common [18]. Unlike an ectopic pregnancy, an uncomplicated ovarian cyst, as denoted by the daughter cyst sign, does not demonstrate a "ring of fire" sign, which is intense peripheral color Doppler flow around the gestational sac. Additionally, a fetal pole or fetal heart tones may be noted in an ectopic pregnancy.

As opposed to ectopic pregnancy, an uncomplicated ovarian cyst may be followed conservatively with an excellent prognosis. However, ovarian cystic masses increase the incidence of ovarian torsion. The daughter cyst sign should also not be confused with an ureterocele, which has a similar sonographic appearance but is present in the posterior lateral wall of the bladder.

TEACHING POINT

It is important to recognize the daughter cyst sign as a specific indicator of an uncomplicated ovarian cyst, as opposed to an ectopic pregnancy. It may be found in normal neonates and infants or in children with precocious puberty. However, in a patient with the potential to become pregnant, excluding an ectopic pregnancy is an important clinical consideration.

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Figure 1: 3-year-old female with precocious puberty and McCune-Albright syndrome. Sonogram of the pelvis performed with multi-frequency probe (GE sector 4-10) in transverse view demonstrates a right adnexal cyst (C) measuring 3.3 cm x 3.5 cm x 3.4 cm. A daughter cyst is seen (arrow) inside the large cyst measuring 7.0 mm x 6.5 mm x 7.0 mm.



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Figure 3: 3-year-old female with precocious puberty and McCune-Albright syndrome. Sonogram of the pelvis performed with multi-frequency probe (GE sector 4-10) in longitudinal view with color Doppler analysis demonstrates a right ovarian cyst (C) measuring 3.3 cm x 3.5 cm x 3.4 cm without peripheral or internal color Doppler flow. A small amount of color flow is noted in the right ovarian parenchyma. A daughter cyst is seen (arrow) inside the large cyst measuring 7.0 mm x 6.5 mm x 7.0 mm. B - Bladder.

Etiology	Ovarian stimulation or precocious puberty resulting in ovarian follicle formation		
Incidence	Small studies report daughter cyst sign in 82% of ovarian cystic lesions in neonates, infants, and		
	young children [7] and 25% of prenatal ovarian cysts [15]		
Gender ratio	Exclusively female		
Age predilection	Reported prenatally up to 36 months of age [7,15]		
Risk factors	Stimulation in utero from maternal hormones, central precocious puberty, peripheral precocious		
	puberty		
Treatment	Conservative management with clinical follow up, must exclude ectopic pregnancy and other cystic		
	abdominal masses		
Prognosis	Excellent, however, increases likelihood of ovarian torsion		
Imaging features	Peripherally based simple cyst within a larger simple cyst on US		

Table 1: Summary table for daughter cyst sign

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	US	СТ	MRI	
Uncomplicated	-Peripherally based simple cyst within	-Cystic ovarian lesion	-Cystic ovarian lesion	
ovarian cyst	a larger simple cyst associated with the	-Daughter cyst likely below	-High T2 signal intensity	
	ovary	resolution on CT	-Daughter cyst below MRI	
	-No increase in color Doppler flow		resolution	
Ectopic	-Peripheral cyst (yolk sac) within a	-Ring enhancing cystic	-Cystic sac-like structure	
pregnancy	larger echogenic cyst (gestational sac)	structure on contrast	-Low T2 intensity surrounding	
	-"Ring of fire" on color Doppler flow	enhanced CT [19]	hematoma [20]	
	-Possible fetal pole associated with	-Possible hemoperitoneum	-Enhancing tubal structure post	
	yolk sac	or intraperitoneal free fluid	contrast with tubal ectopic [20]	
	-Fetal heart tones may be present		-Hematosalpinx: low signal intensity	
	-Possible free fluid in the abdomen		on T2, intermediate on T1 [21]	
	and pelvis			
Ureterocele	- Peripherally based simple cyst within	Posterior lateral cystic	Posterolateral cystic lesion within	
	the bladder	mass within the bladder	the bladder following fluid signal	
	- Associated ureteral jet on Doppler		intensity	
	imaging		-Bright T2	

Table 2: Differential diagnosis of the daughter cyst sign as demonstrated by various imaging characteristics

ABBREVIATIONS

CT = Computed tomography MRI = Magnetic resonance imaging US = Ultrasound

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KEYWORDS

Daughter cyst; ovarian cyst; ovarian follicle; ectopic pregnancy