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Rare Congenital Left Ovarian Cyst with Elevated Tumor Markers in a Newborn

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Dear *MJSCC* Editor-in-Chief,

Please find below our manuscript entitled *Rare Congenital Left Ovarian Cyst with Elevated Tumor Markers in a Newborn* for consideration for publication in *MJSCC*. This case report describes a female neonate with a prenatally identified left ovarian cyst that was found postnatally to be a large complex left adnexal mass with cystic and solid components, internal septations, absence of identifiable normal left ovarian tissue, and markedly elevated tumor markers.

This case is clinically noteworthy because complex ovarian masses in newborns are rare and can present diagnostic uncertainty. The lesion's size, complex morphology, absent normal ovarian tissue, and elevated tumor markers raised concern for future torsion, hemorrhage, infarction, or current neoplasm, highlighting the importance of early imaging, laboratory evaluation, and multidisciplinary referral.

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Thank you for your time and consideration. We appreciate the opportunity to submit this case report to *MJSCC*.

Sincerely,

Rachel Battersby MSIII

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Abstract:

A female neonate was born at 38 weeks and 1 day of gestation via normal spontaneous vaginal delivery to a 26-year-old G3P1 now P2 mother. mother. Prenatal ultrasound imaging identified a 4 × 4 cm left ovarian cyst. On day one of life, a postnatal pelvic ultrasound revealed a complex left adnexal mass measuring 5.9 x 4.8 x 4.6 cm with both cystic and solid components, internal septations, and absence of normal left ovarian tissue, raising concern for ovarian torsion or neoplasm. The right ovary was mildly enlarged, and the endometrium appeared thickened for age. Laboratory evaluation revealed markedly elevated alpha-fetoprotein (AFP >20,000 ng/mL) and lactate dehydrogenase (LDH 1,235 U/L), further increasing suspicion for a malignant germ cell tumor, although physiologic neonatal elevations were considered. The combination of the lesion's size, complex morphology, and elevated tumor markers prompted patient transfer to Valley Children's Hospital for multidisciplinary evaluation and management. This case illustrates a rare presentation of a complex ovarian mass in a newborn and highlights the importance of early imaging and laboratory assessment.

Key Words: Neonatal ovarian cyst, Congenital adnexal mass, Alpha Fetoprotein (AFP), Pediatric gynecology

Introduction:

Ovarian cysts are the most frequently identified abdominal masses in female fetuses and newborns, with an estimated incidence of about 1 in 2,500 live births [3]. A cyst is generally considered pathological when it measures larger than 2 by 2 centimeters. Most neonatal ovarian cysts are small follicular cysts that appear simple and unilocular on prenatal ultrasound and often resolve on their own within the first few months of life. However, when cysts become large, the most serious complication is torsion, which can threaten ovarian function if not recognized promptly [1].

In contrast, complex cysts, which contain internal septations, solid components, or echogenic debris, carry a higher risk of complications, such as torsion, hemorrhage, or infarction. These lesions often develop because of a larger pre-existing simple cyst. Ovarian vascular dysgenesis is another risk associated with complex cysts that is not typically seen with simple cysts, further emphasizing the seriousness of these findings [1].

Interpreting tumor markers in newborns adds further diagnostic difficulty. Alpha fetoprotein (AFP), a glycoprotein produced by the fetal yolk sac and liver, is normally elevated in the neonatal period but can also increase significantly in the presence of germ cell tumors [3]. Distinguishing between a normal physiologic elevation and a pathologic rise requires close correlation between laboratory findings, imaging results, and clinical presentation.

We present the case of a newborn with a complex left ovarian mass, markedly elevated AFP and LDH levels, and absence of normal ovarian tissue, first identified before birth and evaluated after delivery. This case, managed at Valley Children's Hospital in Madera, California, highlights the

diagnostic challenges in distinguishing benign neonatal ovarian cysts from neoplastic lesions and underscores the importance of early multidisciplinary assessment.

Case Presentation:

35 weeks and 3 days of gestation, a maternal fetal medicine specialist identified a 4 × 4cm left ovarian cyst on the routine anatomy ultrasound of a G3P1 26-year-old. At that time, no medical treatment was indicated, and the mother and baby continued to be monitored.

A female neonate was born at 38 weeks and 1 day of gestation via normal spontaneous vaginal delivery. Maternal history was notable for multiple ovarian cysts, gestational thrombocytopenia, and rubella non-immunity. The newborn's Apgar scores were 8 at one minute and 9 at five minutes. The newborn's initial physical examination was unremarkable.

On the second day of life, a pelvic ultrasound of the newborn revealed a complex left adnexal mass measuring approximately 5.9 × 4.8 × 4.6 cm, characterized by both cystic and solid components with internal septations. A nonvascular solid component measuring about 3 × 2.7 × 2.4 cm was also identified. The left ovary was not visualized separately, while the right ovary appeared mildly enlarged for age (volume 2 cc) and contained a small simple cyst measuring 9 × 8 × 4 mm. No free intraperitoneal fluid was observed.

Additional ultrasound findings included a heterogeneous and thickened endometrium measuring 5 mm, which was considered prominent for age, and mild urinary bladder wall thickening, likely related to underdistention or possible cystitis.

On the fourth day of life, pediatric surgery at Valley Children's Hospital in Madera County, along with Pediatric Gynecology at Stanford Hospital in Palo Alto, was contacted for further guidance.

On the fifth day of life, an MRI of the abdomen and pelvis WO contrast was performed at Valley Children's Hospital. Imaging showed mild left hydronephrosis and a cystic mass with a large mural nodule anterior to the left kidney near the colon measuring 3.9 x 3.8 cm and showing no enhancement. A rounded, solid appearing structure inside the cyst was 2.7 cm in the long axis.

Laboratory studies were obtained based on pediatric gynecology recommendations, including CA-125, Inhibin A and B, alpha-fetoprotein (AFP), lactate dehydrogenase (LDH), and human chorionic gonadotropin (hCG). Results demonstrated a markedly elevated AFP at >20,000 ng/mL (reference range: < 8ng/mL), elevated LDH at 1,235 U/L (reference range: 84-246 U/L), elevated quantitative B-hCG at 14 (reference range: 0-5), normal CA-125 (4 U/mL (reference range: < 35 U/mL), and normal Inhibin B < 10pg/mL (reference range: < 10pg/mL),. Inhibin A was not performed.

Given the size and complex morphology of the adnexal mass along with the marked elevation of tumor markers, the decision was made to transfer the patient to Valley Children's Hospital for further multidisciplinary evaluation and management.

Discussion:

While neonatal ovarian cysts are the most common fetal/neonatal abdominal masses in female infants, *complex neonatal ovarian cysts* (with solid elements, septations, or signs of torsion)

remain relatively uncommon. For example, in a 20-case series, only 30% were “complex” cysts [1]. In a 77-case retrospective study, spontaneous regression of simple cysts occurred in 41% vs only 12% in complex cysts [5]. Thus, while simple cysts occur often, the subtype with complexity or potential for neoplasm/torsion is notably less common.

This case is particularly noteworthy because of the presence of a complex neonatal ovarian mass with both solid and cystic components, markedly elevated tumor markers, and no identifiable normal ovarian tissue. Together, these findings raise concern for ovarian torsion, hemorrhage, or infarction, while also increasing suspicion for an underlying neoplasm.

Management of such cases remains challenging, as there is limited consensus regarding the optimal approach to neonatal ovarian cysts, especially those with complex features [1].

Differentiating benign cysts from malignant or pre-malignant lesions in neonates is difficult because of overlapping imaging features and physiologic elevation of tumor markers. However, features such as larger size (>40 mm or >4-5 cm), complex morphology (solid parts, septations, debris, absence of flow), or associated symptoms increase the risk of complications like torsion, hemorrhage, or ovarian loss [5]. Surgical intervention remains controversial; initial observation spared a significant number of neonates from surgery without worsening ovarian salvage [4].

Conclusion:

This is a rare instance of an ovarian mass with complex characteristics and significantly high levels of serum tumor markers that fall into a diagnostic gray zone. In this particular case, the inability to identify any ovarian tissue along with an AFP level greater than 20,000 ng/mL cannot be attributed

to physiological variations seen in newborns alone, making expectant management an unreasonable option at this point. The complexity of the issue warranted a multidisciplinary approach involving maternal-fetal medicine, neonatology, pediatric surgery, and gynecology, all of which started even before birth.

The important takeaway from this case is the common dilemma of making timely clinical decisions with little evidence to guide intervention. The presence of solid elements in addition to no ovarian tissue led to the prompt consultation of a surgeon; however, there is still no prospective literature available to establish any clinical guidelines. Preservation of ovarian tissue in neonates holds significant future reproductive implications that cannot simply be overlooked for a mere consideration of cancer risks.

For now, multicenter cooperation and prospective database registries are the only means of establishing evidence-based clinical guidelines, which is why cases such as these provide the basis for such future work.

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