Fetal ovarian cyst mimicking a CSF pseudocyst in the setting of shunt failure

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Fetal ovarian cysts are common congenital lesions encountered in the neonatal population. These cysts are typically benign and rarely require any invasive intervention. Abdominal pseudocyst formation as a result of a ventriculoperitoneal shunt is a relatively infrequent occurrence and is similarly an uncommon cause of shunt failure. The authors present the case of a 4-month-old girl with shunted hydrocephalus who presented with shunt failure from a suspected abdominal pseudocyst that was found to be a fetal ovarian cyst.

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KEY WORDS fetal ovarian cyst; abdominal pseudocyst; ventriculoperitoneal shunt; hydrocephalus

The formation of an intraabdominal fluid collection at the distal end of a functioning ventriculoperitoneal (VP) shunt is somewhat rare and can be found in the setting of shunt failure. These collections are typically referred to as pseudocysts, and are usually associated with shunt infection. Such pseudocysts are frequently associated with shunt malfunction. In this paper we present a case of an infant who presented with a potential shunt malfunction, and was found to have a fetal ovarian cyst mimicking a pseudocyst.

Case Report

History and Examination

The patient was born preterm as a twin at 25 weeks gestational age. Her early neonatal course was complicated by Escherichia coli sepsis and meningitis, with the subsequent development of postinfectious hydrocephalus requiring a right occipital VP shunt at 11 weeks of age. Placement of the peritoneal catheter was performed laparoscopically, and no intraabdominal issues were reported at that time. She then presented at 4 months of age with a 5-day history of lethargy and emesis. On examination, there was a slight upward gaze preference (which was her baseline), an increased head circumference, and a bulging fontanelle. There was slight dullness to percussion on abdominal examination, but the abdomen was otherwise soft and not tender, with no peritoneal signs. The patient exhibited breast budding and growth of pubic hair.

An MR image of the brain revealed acute hydrocephalus with an encysted left temporal horn (Fig. 1). The temporal horn cyst was not observed on previous imaging and was believed to have developed because of her previously treated meningitis. An MR image of the abdomen was obtained that demonstrated a large, well-defined cyst measuring $5.3 \times 3.7 \times 4.4$ cm$^3$ in the right lower quadrant adjacent to the right ovary. There was another $3.4 \times 2.8 \times 2.2$ cm$^3$ well-defined daughter cyst found within the larger cyst. Multiple loops of the peritoneal catheter were noted adjacent to the larger cyst, while the tip of the catheter was located in the left lower quadrant with minimal free fluid surrounding it (Fig. 2). The radiological diagnosis was that of an ovarian cyst given the presence of a daughter cyst; a CSF pseudocyst was believed to be less likely (Fig. 3). Subsequent workup included CSF cultures, which were held for 5 days, from both the VP shunt and ultrasonography-guided aspiration of the abdominal cyst. These cultures were found to be negative for infection or malignancy. Benign mesothelial cells were identified on the abdominal cyst aspirate, which helped distinguish this cyst from a CSF pseudocyst, and along with the radiological characteristics, confirmed the diagnosis of an ovarian cyst.

Operation and Postoperative Course

From a neurosurgical perspective, the patient clinically had signs of shunt malfunction; distal obstruction related

ABBREVIATION VP = ventriculoperitoneal.
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to the ovarian cyst was believed to be possible, but unlikely. Given the dilated ventricular system and encysted temporal horn, it was felt that she would benefit from an endoscopic fenestration of the cyst within the left temporal horn, as well as an endoscopic third ventriculostomy; these were performed on Day 3 after admission. Because the CSF cultures from the VP shunt were negative for infection, the shunt hardware was not removed. CSF sent for microbiology assessment at the time of ventricular endoscopy was cultured for 5 days and was negative for infection. Unfortunately, the volume of fluid drawn from the abdominal cyst was inadequate for hormone analysis. Serum alpha-fetoprotein and estradiol were highly consistent with a functioning ovarian cyst. The gynecology service was consulted and a conservative approach was suggested. The patient recovered well postoperatively and she was eventually discharged, clinically improved, on postoperative Day 3. A postoperative MR image and CT scan showed adequately decompressed ventricles.

**Discussion**

Fetal ovarian cysts are the most common, prenatally diagnosed intraabdominal cysts, comprising approximately 5% of all intraabdominal masses diagnosed in females shortly after birth. The exact etiology of these cysts is not known, but their pathogenesis is likely endocrinological in origin. As reviewed by Akın et al., there is an increased incidence of fetal ovarian cysts among pregnancies complicated by diabetes, preeclampsia, and Rhesus factor incompatibility, attributed to the increased levels of placental chorionic gonadotropin levels that accompany these conditions. Furthermore, fetal hypothyroidism, congenital adrenal hyperplasia, and ovarian hyperstimulation observed in premature infants have been implicated in ovarian cyst formation.

Fetal ovarian cysts are simple, benign cysts, which typically resolve during the perinatal period in more than...
half of the cases. Histologically, these cysts are lined by benign mesothelial cells, which in our case helped to differentiate it from a CSF pseudocyst, which lacks mesothelial cells. Once diagnosed, these cysts are monitored with serial ultrasonography examinations for changes in size, structure, or intracystic hemorrhage, which may lead to or be a sign of impending torsion, the most common complication associated with these cysts. Other potential complications include hydramnios and ascites. There are no specific features for simple cystic lesions except for the presence of “daughter cysts” or a “cyst within a cyst” being specific for an ovarian cyst. Another possibility for the cyst-within-a-cyst appearance is a hydatid cyst, which is extremely uncommon in infants.

A CSF pseudocyst is a rare but well-described complication of VP shunts, with an incidence of 0.7%–4.5%. Pseudocysts typically arise between 3 weeks and 5 years after placement of the VP shunt, but have been reported to develop as long as 10 years after VP shunt implantation. Patients typically present with abdominal pain, nausea, vomiting, fever, decreased appetite, and symptoms and signs of shunt failure. The etiology of pseudocyst formation is believed to be either a low-grade infection or an inflammatory reaction involving the peritoneum and intestinal serosa. This notion is supported by histopathological findings of nonpapillary fibrous tissue and granulomatous tissue lined within the pseudocyst and inflammation in the surrounding serosal surfaces. When a CSF pseudocyst is associated with an infection, Staphylococcus epidermidis, Staphylococcus aureus, and Propionibacterium acnes are the most commonly isolated pathogens. A review of the literature by Yuh and Vassilyadi demonstrated that the reported incidence of infection with a pseudocyst ranged between 8% and 100%. Treatment of a documented pseudocyst in the context of a VP shunt typically consists of externalization of the peritoneal catheter for a period of time while treating the VP shunt infection with antibiotics, followed by repositioning of the distal catheter within the abdominal cavity. On occasion, image-guided percutaneous drainage of the pseudocyst is indicated, especially if the cysts are large.

In most instances, the finding of a well-organized intraabdominal fluid collection around a distal VP shunt catheter in the setting of clinical shunt failure has a relatively short differential diagnosis, including intraabdominal abscess or a sterile or infected pseudocyst. To our knowledge, this is the first reported description of a fetal ovarian cyst occurring in the context of a patient with an indwelling VP shunt presenting with shunt malfunction. The presence of a fetal ovarian cyst in such patients has implications for therapy that are different than those for patients with documented and infected pseudocysts.

References

Author Contributions
Acquisition of data: Calayag, Cantillano Malone, Drake, Chavhan. Analysis and interpretation of data: Rutka, Chavhan. Drafting the article: Calayag, Cantillano Malone. Critically revising the article: Rutka, Calayag, Cantillano Malone, Drake. Reviewed submitted version of manuscript: Rutka, Calayag, Cantillano Malone, Drake. Approved the final version of the manuscript on behalf of all authors: Rutka.

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