Mesothelial Inclusion Cyst In Patient With Beckwith-Weidemann Syndrome: A Rare Care Report

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BACKGROUND

Hepatic cysts in neonates are uncommon, with an incidence of 2.5% postnatally. The most common diagnosis is a congenital hepatic cyst which is a superficial cyst coated by a single layer of cuboidal or columnar epithelium. (5) Mesothelial inclusion cysts have been described in one other case study in neonates.(4) Mesothelial inclusion cysts represent benign cystic lesions delineated by a single layer of mesothelial cells and have been described in a variety of anatomical locations, typically described in adult females (1–3)

CASE

A 0 day-old female born via cesarean section at 34-weeks 6-days with omphalocele. Of note, other abnormalities included macrosomia and a sacral dimple. Primary operative repair was performed on first day of life.

Hospital course

•Findings included a 6-cm fascial defect, which was repaired. In addition, multiple hepatic cysts were discovered along a narrow tongue of liver protruding through the fascial defect that extended off the right hepatic lobe and included the gallbladder.

•Patient was managed in the NICU until tolerating PO intake. Genetic testing subsequently revealed a diagnosis of Beckwith-Weidemann syndrome.

•Pathology report of hepatic cysts showed mesothelial inclusion cyst with hepatic plate malformation.

•This the second case described mesothelial inclusion cyst of the liver in a clinical setting of Beckwith-Wiedemann syndrome in English literature

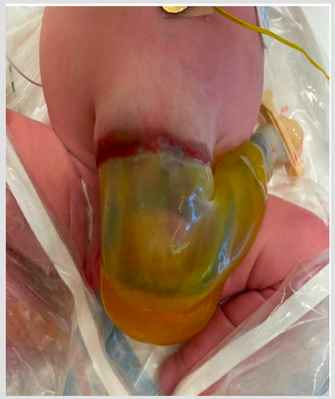

Figure 1. Patient's abdomen with omphalocele present

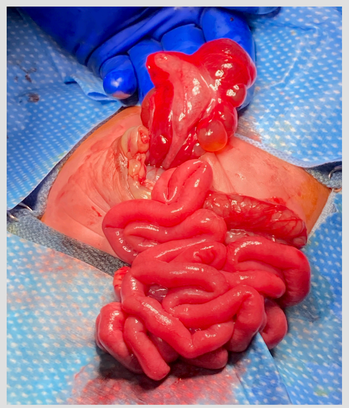

Figure 3. Intraoperative findings of cysts involving gallbladder

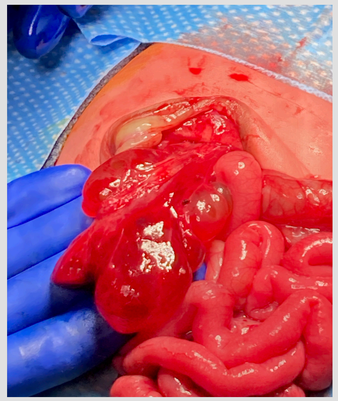

Figure 2. Intraoperative findings of multiple hepatic cysts

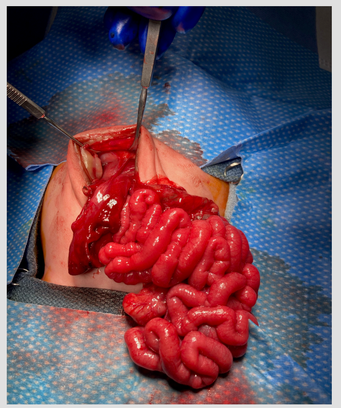

Figure 4. Post excision of cyst

DISCUSSION

Mesothelial inclusion cysts represent benign cystic lesions delineated by a single layer of mesothelial cells and have been described in a variety of anatomical locations, most-typically on the serosal surface of the visceral organs.(1) Cysts of mesothelial origin are lined by flat, cuboidal, or columnar cells and their wall is fibrous without any lymphatic structures. These cysts have no clear origin. (6) Beckwith-Weidemann Syndrome is a genetic disorder characterized by alterations in growth regulatory pathways leading to gigantism, macroglossia, and tumor predisposition. It is also known for increased frequency in abdominal wall defects such as omphalocele and diastasis recti. (7) While embryonal tumors are commonly described in this disorder in the literature, a mesothelial inclusion cyst has only been described in one other case report. In the original case report, the authors noted the patient had an omphalocele containing an accessory lobe of liver that contained cysts. Pathology revealed these cysts contained a thin layer of mesenchymal cells with immunohistochemical (IHC) staining reactive for calretinin, cytokeratin, and D2-40 thus confirming mesothelial origin. (4) Our patient had similar findings on IHC staining reactive for D2-40, Calretinin, CK5/6. Prior to the advent of IHC staining, cysts of this origin were difficult to discern endothelial vs mesothelial. (6)

REFERENCES

1.Soon DS, Shilton H, Andrabi A. Mesothelial inclusion cyst: a rare occurrence. J Surg Case Rep. 2016 Dec 19;2016(12):rjw213.
2.Urbańczyk K, Skotniczny K, Kuciński J, Friediger J. Mesothelial inclusion cysts (so-called benign cystic mesothelioma)--a clinicopathological analysis of six cases. Pol J Pathol Off J Pol Soc Pathol. 2005;56(2):81–7.
3.Elbouhaddouti H, Bouassria A, Mouaqit O, Benjelloun EB, Ousadden A, Mazaz K, et al. Benign cystic mesothelioma of the peritoneum: a case report and literature review. World J Emerg Surg WJES. 2013 Oct 13;8:43.
4.Rougemont AL, Sartelet H, Oligny LL, Bensoussan A, Yazbeck S, Fournet JC. Accessory Liver Lobe with Mesothelial Inclusion Cysts in an Omphalocele: A New Malformative Association. Pediatr Dev Pathol. 2007 May 1;10(3):224–8.
5.Recinos A, Zahouani T, Guillen J, Rajegowda B. Congenital Hepatic Cyst. Clin Med Insights Pediatr. 2017 Apr 10;11:1179556517702853.
6.Perrot M de, Bründler MA, Tötsch M, Mentha G, Morel P. Mesenteric Cysts. Dig Surg. 2000;17(4):323–8.
7.Weksberg R, Shuman C, Beckwith JB. Beckwith–Wiedemann syndrome. Eur J Hum Genet. 2010 Jan;18(1):8–14.