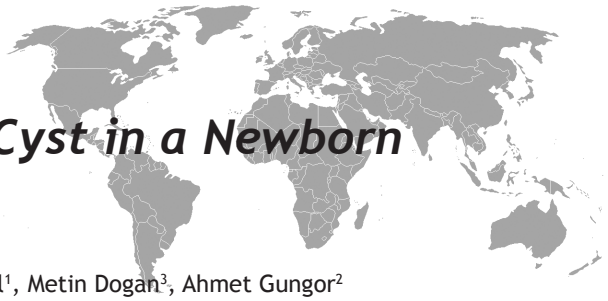

Congenital True Pancreatic Cyst in a Newborn

Emine Turkmen Samdanci[1], Sema Uguralp[2], Zeynep Nurkabul[1], Metin Dogan[3], Ahmet Gungor[2]

ABSTRACT

Congenital true pancreatic cysts are quite rare in the children. Fetus, which shows an intraabdominal cystic mass in ultrasound (US) performed during the antenatal examination, was delivered at a gestational age of 37 weeks. As a result of the histopathological examination, pancreas-originated cystic mass with the dimensions of 8x5x1cm, which has been totally excised during the operation was reported to be a congenital true pancreatic cyst. This rarely seen entity, which has been diagnosed during the neonatal period before the occurrence of any clinical signs and which has been successfully treated, was presented along with the literature.

Key words: *Pancreas, congenital cyst, newborn, true cyst, antenatal diagnosis*

Yenidoğanda Konjenital Gerçek Pankreatik Kist

ÖZET

Konjenital gerçek pankreatik kistler, çocuklarda oldukça nadirdir. Antenatal muayenesinde ultrasonografi ile intraabdominal kistik kitle tespit edilen fetus, 37 haftalık iken doğurtuluyor. Ameliyatta total eksize edilen pankreas kaynaklı 8x5x1cm'lik kistik kitlenin histopatolojik inceleme sonucu konjenital gerçek pankreas kisti olarak rapor edildi. Klinik bulgu vermeden yenidoğan döneminde tanı alan ve başarılı şekilde tedavi edilen bu nadir antite literatür eşliğinde sunulmuştur.

Anahtar kelimeler: *Pankreas, konjenital kist, yenidoğan, gerçek kist, antenatal tanı*

INTRODUCTION

Non-traumatic congenital true pancreatic cysts are very rarely seen during the childhood. True pancreatic cysts account for less than 1% of all pancreatic cysts. Generally, they are symptomatic below the age of 2 years. In the literature, there are about 26 case reports (1, 2). We presented this case because it was seen in a newborn, as a rare entity.

CASE

Female newborn with a birth weight of 3 kg is the 2nd live birth of the 4th pregnancy of the 35-years-old mother. Fetus, which shows an intraabdominal cystic mass in antenatal ultrasound (US) performed, was delivered with cesarean section at a gestational age of 37 weeks. The case was consulted to the department of pediatric surgery. The patient, who showed a good general status and normal vital signs, in the physical examination, had a mobile, palpable mass with a diameter of approximately 5 cm in the epigastric area. Examinations of other organ and system were normal. Blood and urine biochemistry and tumor markers were within normal range. Abdominal US revealed a cystic mass with the dimensions of 7x5 cm with septa, laid across the midline, for which the relation with splenic hilus cannot be clearly observed. In the direct lateral abdominal radiograph obtained by giving oral

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contrast, the mass was uniformly bounded and was pushing the intestine towards the anterior part (Figure 1). As, in US, the mass was mostly considered to be consistent with mesenteric cyst, the patient did not have computerized tomography (CT) or magnetic resonance imaging (MRI). Patient was taken into the operation with the prediagnosis of mesenteric cyst or duplication cyst. In the exploration, in addition to two adjacent cysts with the sizes of 7 cm, and 5 cm which originated from the tail of the pancreas and did not show adhesion to adjacent tissues, a mass that contains numerous milimetric cysts was observed. Cystic mass that have been totally excised after the aspiration of large cysts was sent to pathology laboratory in 10% formalin.

Macroscopically, the mass with a multicystic appearance had the dimension of 8x5x1 cm. In addition to the largest cyst with a diameter of 7 cm and a wall thickness of 0.2cm, several similar cystic structures with variable sizes were observed. Samples, which have been stained with hematoxylene-eosine (H-E) after the routine tissue monitorization, and evaluated using light microscope. Microscopically, cystic spaces of different dimensions, lined with cuboidal epithelium without atypia, were nested within pancreatic tissue (Figure 2). While epithelium was positively stained with cytokeratine 7 antibody (CK7) (Figure 3), it was not stained with CD34, calretinin and mesothelin antibodies. The case had the diagnosis of congenital true pancreatic cyst, with histomorphologic, immunohistochemical and clinical findings.

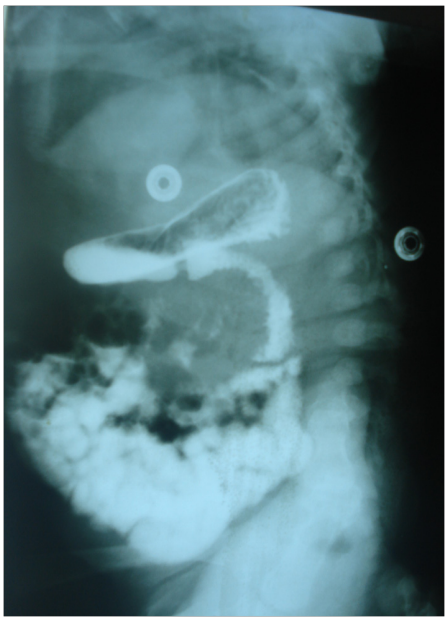

***Figure 1**. Increase of uniformly bounded density consistent with homogenous mass, that cause a compression on intestinal segments filling the upper abdomen in the epigastrium (Barium swallow)*

DISCUSSION

Congenital pancreatic cysts are rarely symptomatic and, in general, they are incidentally detected (3, 4). These cysts are subgrouped as congenital, retention, duplication, pseudocysts, parasitic and neoplastic cysts (4, 5). True cysts (congenital, retention and duplication) are very rarely seen and have a true epithelium on their wall (6). They may be associated with the syndromes that lead to large malformations or cystic diseases localized in other organs (7). There are some publications that reported the presence of pancreatic cysts in the patients with trisomy, tuberculosis, von Hippel-Lindau or cystic fibrosis (7, 8). Our case was sporadic and had no any syndrome. Wall of the most commonly encountered post-traumatic pancreatic pseudocysts had no epithelium (1, 4, 5, 9)

Congenital pancreatic cysts generally show its presenting clinical signs below the age of 2 years. Majority of approximately 26 cases described in the literature were below the age of 2 years (10). Our case is a newborn with a mass detected during the fetal period. Although the patients are generally asymptomatic, they may exhibit abdominal distension, vomiting, jaundice or pancreatitis (9). Our case was not diagnosed based on a symptoms related to existing cyst, but incidentally in US performed during the antenatal examination. True cysts are most commonly localized in the tail or body of the pancreas (62%) and less commonly seen in the head of pancreas (33%) (9). In our study, true cyst was localized in the tail of pancreas, consistently with the literature. It is thought that true pancreatic cysts result from a developmental abnormality related to the secretion of primitive pancreatic ductus (1, 11). In the case presented herein, great number of cysts associated with the tail of pancreas detected during the operation and the similarity between the epithelium lining the cysts and pancreatic ductal epithelium were considered as abnormality of ductal development. Cysts that develop secondarily to chronic obstruction of the ductal system are called retention cysts. It is difficult to histomorphologically differentiate the retention cysts from congenital true cysts. Late occurrence

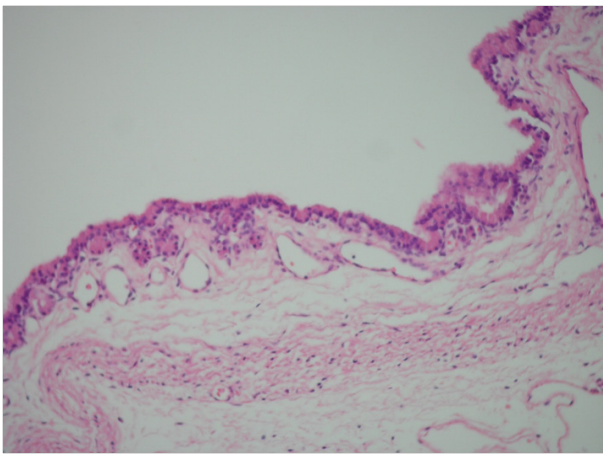

***Figure 2**. The cyst which consist of fibrovascular wall, is lined by duct type cuboidal epithelium (H-E X200)*

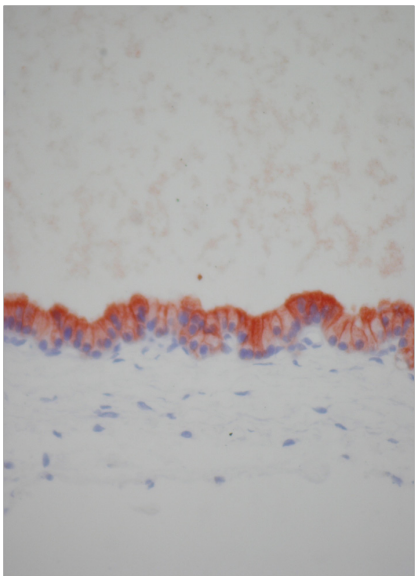

***Figure 3**. Positive staining of the cyst epithelium with CK7 (X400)*

of the signs of retention cysts and the biochemical analysis of the cystic fluid (such as the level of amylase) may be useful in the differential diagnosis (1, 5). Our case had the diagnosis of congenital true cyst based on the fact that the condition has been noted in the late period and on the pathological examination of the excision material. In the treatment of cysts, total excision should be used. If it is not possible, the use of internal drainage methods is recommended (12). In our case, total excision was successfully performed. Long-term (11 months) controls of the patient, who have been discharged owing to the lack of any problem during post-operative three days, were normal.

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