Prenatally Detected Giant Solitary Nonparasitic Hepatic Cyst in a Newborn: Report of a Case

Yenidoğanda Prenatal Saptanan Dev Soliter Nonparazitik Hepatik Kist: Olgu Sunumu

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Abstract

Giant solitary nonparasitic hepatic cysts are rarely encountered in children, and few antenatally diagnosed cases have been reported in the newborn period. We report a 1- day -old girl with large solitary hepatic cyst, that was detected antenatally as a cystic mass in the abdomen and presented with abdominal distention. Although guite large the cyst could be removed totally without any complication.

Solitary nonparasitic hepatic cysts should be considered in the differential diagnosis of intraabdominal cystic lesions. If total excision is possible, it should be preferred because of the risk of recurrence and malignant transformation. *Pam Med J 2010;3(1):45-7.*

Key words: Neonatal abdominal cyst, hepatic cyst, solitary

Özet

Dev soliter nonparazitik hepatik kistler ile çocuklarda nadiren karşılaşılır ve antenatal tanılı çok az yenidoğan bildirilmiştir. Antenatal dönemde karında kistik kitle saptanan ve karın distansiyonu ile gelen dev soliter hepatik kistli bir günlük kız bebek rapor edilmiştir. Kist oldukça büyük olmasına rağmen herhangi bir komplikasyon olmadan total olarak çıkarılabilmiştir. Soliter nonparazitik hepatik kistler intraabdominal kistik lezyonların ayırıcı tanısı içinde değerlendirilmelidir. Rekürrens ve malign transformasyon riskleri nedeniyle mümkünse total olarak çıkarılmalıdırlar. *Pam Tıp Derg 2010;3(1):45-7*.

Anahtar sözcükler: Neonatal abdominal kist, hepatik kist, soliter

Introduction

Solitary nonparasitic hepatic cysts (SNPHC) are uncommon in children, and few cases have been reported in the newborn period. There is a female predominance of 4:1 in comparison to males. The most common location is the right lobe of the liver [1,2]. The cysts are usually unilocular but may be multilocular in 5% of patients [3]. We report a female newborn with a giant nonparasitic hepatic cyst which was antenatally diagnosed as a cystic lesion in the abdomen and presented with abdominal distention soon after birth.

Case

A 3320 g, 1-day-old full-term female with an antenatally diagnosed cystic lesion in the

abdomen was admitted to our clinic with abdominal distension. Physical examination except abdominal was unremarkable distension, marked on the right side. The results of routine blood investigations were normal. The α -fetoprotein level was within normal limit. Abdominal radiography showed a large mass occupying the entire right side of the abdomen, displacing the bowel to the left. Ultrasonography and magnetic resonance imaging (MRI) revealed a huge, unilocular cystic mass, extending from the inferior surface of the liver to the pelvis (Fig. 1). The mass extended posteriorly and leaned on inferior vena cava at the infrahepatic level. But no connection was detected with main vessels. No communication was detected between the cyst and the biliary

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system. The laparotomy was performed on the tenth day of life. There was a giant cystic mass measuring 14x10.5x5 cm arising from the antero-inferior edge of the right lobe of the liver, and the gallbladder was stretched by it (Fig. 2).



Figure 1: Magnetic Resonance Imaging reveals a huge, unilocular cystic mass, extending from the inferior surface of the liver to the pelvis.



Figure 2: Intraoperative appearance: The cyst is thin-walled, unilocular, and filled with serous fluid stretching the gallbladder by it

There was no connection with the main

vessels and no adherence with adjacent organs. Macroscopically, the cyst was thinwalled, unilocular and filled with serous fluid. A colourless cystic fluid that indicated the absence of communication between the cyst and bile ducts was aspirated. The mass was resected totally with minimal hepatic tissue. 20 ml/kg blood transfusion was required on the peroperative (intraoperative and early postoperative) period and 20 ml/kg for the following 3 days. Histopathological examination of the specimen revealed a simple solitary cyst, lined by cuboidal epithelium. The postoperative period was uneventful. The patient was discharged on the seventh postoperative day.

Discussion

Cystic lesions in the fetus and newborn period are mainly renal, choledochal, ovarian, mesenteric or duplication cysts [1,2]. Although uncommon, our case and previously reported cases indicate that hepatic cysts should be added among the differential diagnosis of intraabdominal cystic lesions.

Congenital nonparasitic hepatic cysts, polycystic liver disease, hepatic cysts of Caroli's disease, cystadenomas. mesenchymal mucinous hamartomas, teratomas, and foregut cysts are mainly included in benign noninfectious hepatic cystic lesions of childhood [4]. It is reported that congenital cysts of the liver can be divided histologically (mesenchymal hamartoma or simple parenchymal), or on the basis of content (bile containing or not). It is also stated that those that contain bile should be regarded as intrahepatic choledochal malformations. Histologically, congenital simple hepatic cysts have a largely coboidal epithelial lining and an underlying loose connective tissue [5]. In the presented case the cyst was lined with cuboidal epithelium with colourless cystic fluid.

Since the first description of SNPHC by Michel in 1856, a number of cases have been reported. SNPHC occur far less frequently in children than in adults. A review article showed that only 12.7% of patients with SNPHC were younger than 15 years [3]. Furthermore, they are extremely rare in the prenatal and neonatal period and few cases have been reported [2,6,7,8]. Our patient is one of these rare cases having prenatally detected giant solitary hepatic cyst.

Solitary nonparasitic hepatic cysts are usually asymptomatic, and they are detected when patients become symptomatic, or incidentally on radiological imaging performed for another reason. Symptoms are generally due to

compression of the adjacent structures or hemorrhage, torsion or rupture of the cyst [1,2]. Other complications are infection and neoplastic degeneration [9]. The presenting symptoms are usually vomiting and respiratory distress in neonates with SNPHC. Our case presented with abdominal distension in the immediate postnatal period. Although the other two prenatally detected giant solitary hepatic cyst reported in the literature were symptomatic, no symptom due to compression was present in our case. This may probably be due to a difference in growth rates of these tumors thus allowing adaptation in our case.

Antenatally detected hepatic cysts should be investigated with ultrasonography first to demonstrate unilocularity vs multilocularity and other hepatobiliary pathologies. Detailed computed tomography or MRI are also required. Some unilocular cysts contain bile and represent intrahepatic choledochal malformations with a connection to the biliary tree. For such cases, functional hepatic scintigraphy can be highly suggestive [5]. Magnetic resonance cholangiography can be used to evaluate anatomic abnormalities, if required. Intraoperative cholangiography is suggested if connection with biliary tree is suspected or a bile stained cyst was detected [2].

Various therapeutic methods are used in the treatment of solitary non-parasitic hepatic cysts. The simplest is percutaneous obliteration with some agent, for example ethyl alcohol, minocycline chloride or tetracycline hvdrochloride [10.11]. Fenestration excision of the cyst wall is increasingly performed laparoscopically. Total excision of the cyst with or without liver resection is the choice of treatment for nonparasitic cysts of the liver. Deroofing and partial excision may be indicated if major vascular or biliary ductal structures are present in close proximity to the cyst [10]. The possibility of malignant transformation into adenocarcinoma or squamous cell carcinoma justifies the need for complete excision of the cyst [9,10,12,13]. In addition, recurrences are less likely to occur with total excision compared with other methods [1]. For this reason, we performed total excision of the cyst with minimal hepatic tissue.

In conclusion, a solitary non-parasitic hepatic cyst should be considered in the differential diagnosis of intraabdominal cystic lesions in the perinatal period and if total excision is possible, it should be preferred because of the risk of recurrence and malignant transformation.

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