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Case report
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A rare cause of cholestasis: Congenital right diaphragmatic hernia

Kolestazın nadir bir nedeni: Konjenital sağ diafragma hernisi

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Abstract

An 82-day-old infant, investigated for sudden onset jaundice and acholic feces, was referred to our department, as no gallbladder was observed by abdominal ultrasonography. Her chest x-ray revealed an elevated right diaphragm, and the breath sounds were diminished on the right. Suspicious arteriovenous malformation in the fifth segment of the right lobe accompanied by a rotation anomaly of liver was reported. Tomography scans showed right diaphragmatic hernia where the left lobe of liver and gallbladder herniated to the thoracic space. In this case with right Bochdalek hernia, the bile ducts had kinked in the thoracic cavity, causing obstructive jaundice. If the kinking continues despite relocation of organs into the abdominal cavity, cholecystectomy-hepaticojejunostomy is an effective and reliable surgical method. Myriad pathologies may cause cholestasis, and surgical reasons should be investigated and diagnosed without delay to improve prognosis. For closure of the diaphragmatic defect, primary repair should be the preferred method. With this case, we wanted to emphasize a highly different presentation of Bochdalek hernia and remind that diaphragmatic hernia should be kept in mind in patients presenting with cholestasis and acholic stools.

Keywords: Obstructive jaundice, Acholic feces, Cholestasis, Diaphragmatic Hernia

Öz

82 günlük kız hasta ani gelişen sarılık, akolik dışkılama olması nedeniyle yapılan Abdominal Ultrasonografisinde safra kesesinin görülmemesi üzerine kliniğimize yönlendirildi. Akciğer grafisinde sağ diyafram elevasyonu mevcut idi. Hastanın muayenesinde aynı tarafta solunum sesleri azalmış olarak alınıyor idi. Çekilen Tomografisinde sağ diyafragmatik herni mevcut ve karaciğer ile safra kesesi toraks boşluğuna herniye idi. Olgumuzda sağ Bochdalek hernisi mevcut idi ve safrayollarının torasik kavitede sıkışmasına bağlı obstrüktif sarılık gelişmiş idi. Operasyon sırasında alınan karaciğer biyopsisinde fibroz bulguları oluşmaya başlamış idi. Operasyona alınan hastanın abdominal organları yerine alınarak, kolesistektomi ve hepatikojejunostomi operasyonu yapıldı. Erken tanı ve tedavi ile hastanın takibinde yapılan karaciğer biyopsisinde karaciğerde gelişen bulguların düzelmeye başladığı görüldü. Kolestaz, çok çeşitli etyolojik nedenlere bağlı olabilmekle birlikte cerrahi nedenlerin ivedilikle tanı ve tedavisinin yapılması prognoz açısıdan önem arz etmektedir. Bu olgu ile Bochdalek hernisinin çok farklı bir prezantasyon şeklini sunmak istedik. Kolestaz ile gelen akolik dışkısı mevcut olan hastaların ayırıcı tanısında diafragma hernisi de akılda tutulmalıdır.

Anahtar kelimeler: Obstrüktif sarılık, Akolik gaita, Kolestaz, Diyafragmatik hernia

Introduction

Congenital diaphragmatic hernia occurs in about one in every 2000-5000 live births [1]. Bochdalek hernias are left-sided in 78-84% of cases, right-sided in 14-20% and even more rarely bilateral [2,3]. Although congenital right diaphragmatic hernia (CRDH) is known as a condition that may cause obstructive jaundice, it is quite rare fortunately [4]. These hernias typically occur before or shortly after birth and only % 5- 25 develop after 2 months of age [5]. An 82-day-old female infant was diagnosed with CRDH when examined due to obstructive jaundice. With this case, we wanted to emphasize a highly different presentation of Bochdalek hernia and remind that diaphragmatic hernia should be kept in mind in patients presenting with sudden onset jaundice, cholestasis and acholic stools.

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Case presentation

Our 82-days-old term female patient who had no abnormalities in her prenatal follow-up was investigated in another institution for sudden onset jaundice and acholic feces developed within the last week. The patient was referred to our department, as no gallbladder was observed in the abdominal ultrasonography. She had no history of perinatal trauma or distress and she was icteric on physical examination. The liver was 3/2.5 cm palpable subcostally. Her cardiovascular system examination was normal. Laboratory test results were as follows: AST: 91 IU/L (2.2x upper limit of normal (ULN)), ALT: 69 IU/L (1.7x ULN), GGT: 1044 IU/L (41.7x ULN), LDH: 302 IU/L (normal), ALP: 589 IU/L (3.8x ULN), total bilirubin: 8.9 mg/dL (7.4x ULN), direct bilirubin: 8.07 mg/dl (40x ULN). Her chest x-ray revealed an elevated right diaphragm (Figure 1).

After the x-ray findings we found that the breath sounds were diminished on the right side on auscultation. In abdominal ultrasonography (USG), no gallbladder was observed and a suspicious arteriovenous malformation appearance in the fifth segment of right lobe accompanied by a rotation anomaly of liver was reported. Thoracic and abdominal computed tomography (CT) scans revealed a right diaphragmatic hernia. CT scans showed that the left lobe of liver and gallbladder were herniated into the thoracic space (Figure 2).

Venous stasis in the left lobe of liver was noted and it was concluded that the image imitating an arteriovenous malformation was the gallbladder shifted to the paravertebral area and that extrahepatic biliary ducts may be kinked. Magnetic resonance (MR) cholangiographic assessment was planned to confirm the biliary pathology. MR cholangiography showed that the left lobe of liver was in the thorax and that intrahepatic biliary tracts were dilated due to kinked external biliary ducts (Figure 3).

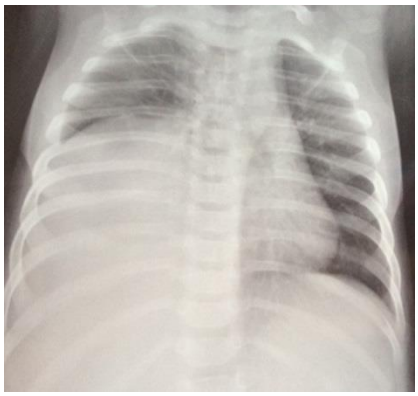

Figure 1: Posteroanterior Chest X-ray; Right diaphragm eventration

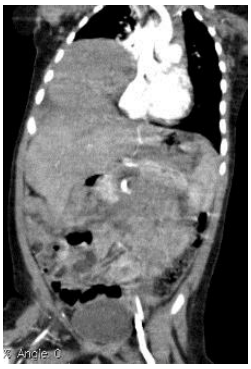

Figure 2: CT scan with intravenous contrast agent showing the placement of CRDH, liver and bile ducts in thoracic cavity

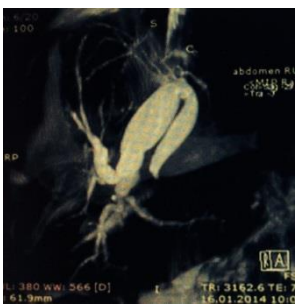

Figure 3: MR cholangiography showing intrahepatic biliary tract dilatation because of kinked up extrahepatic bile ducts

In surgical exploration performed when she was 100 days old, a right sided Bochdalek hernia was found. An ileal segment, right lobe of liver and gallbladder were noted in thoracic cavity. The right lobe had a shapeless structure as it had developed in thoracic cavity. The gallbladder was enlarged with thickened walls. Extrahepatic biliary ducts were kinked up on themselves and hepatic channel along with bile ducts were dilated, 1.1 cm in diameter. Perfusion and hemodynamics were stable as the liver and gallbladder were relocated into the abdominal cavity.

Cholecystectomy, hepaticojejunostomy, 1 cm distal to the right and left hepatic channels, and end-to-side jejunojejunostomy were performed. The congenital defect on diaphragm was repaired. Histopathologic evaluation of liver biopsy taken during surgery revealed severe bile stasis, parenchymal degeneration, mild inflammatory activity, and marked fibrosis (Figures 4, 5).

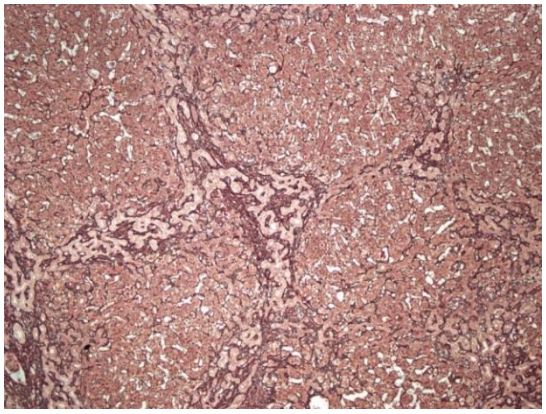

Figure 4: A reticulin stain reveals the parenchyma framework of the lobule in the formerly biopsy (x40)

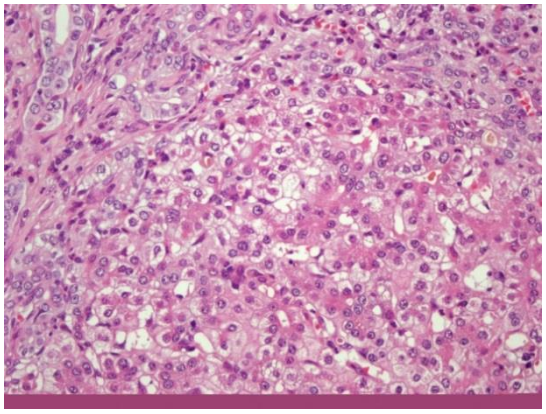

Figure 5: In the same biopsy material, mild inflammation, and mild bile duct proliferation were observed in the parenchyma. Hematoxylin and eosin. x100

She passed a cholic stool on the 4th postoperative (PO) day, oral nutrition was initiated on 5th PO day and laboratory values at discharge on the 8th PO day were as follows: AST: 44IU/L, ALT: 11 IU/L, GGT: 27I U/L, ALP: 103 IU/L, total bilirubin: 1.4 mg/dL, direct bilirubin: 1.02 mg/dL. Laboratory test results of the first visit to our outpatient clinic were within normal ranges. Liver biopsy was performed at the 8th month after the operation due to evidence of severe fibrosis in the first liver biopsy. The last biopsy revealed no cirrhosis and significant reduction of fibrosis from severe to mild in comparison with the first one (Figures 6, 7). The patient is still followed uneventfully by our outpatient clinic. Written consents were obtained from the patient's caregivers for this case presentation.

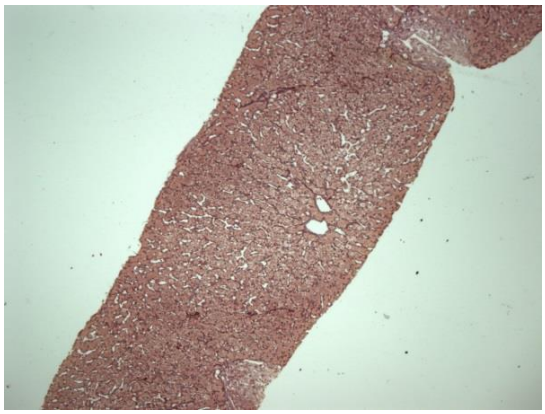

Figure 6: New biopsy material showed no fibrosis. Reticulin stain. x40

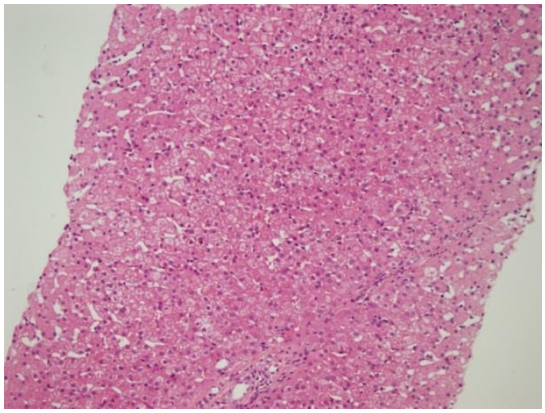

Figure 7: Parenchymal and portal tracts are regular in the new biopsy sample. Hematoxylin and eosin (x100)

Discussion

While most congenital diaphragmatic hernias manifest through dyspnea in the first 24 hours, 10-20% of cases are diagnosed at a future date [6,7]. BH in adults is mostly asymptomatic, hence, it is usually discovered incidentally [8]. Between 40-90% of cases are diagnosed prenatally by MR imaging and USG [9, 10]. In cases with postnatal dyspnea, it is diagnosed easily by physical examination and chest x-ray. In cases without dyspnea, on the other hand, diagnosis comes later due to concomitant pathologies and recurrent lung infections. In jaundiced patients, the reasons of cholestasis, whether it be medical or surgical, should be investigated without delay.

Diagnosis of CRDH is more difficult than that of congenital left diaphragmatic hernia. Chest x-ray is often inadequate in this group, and patients are diagnosed by thoracic CT scan [11]. Correspondingly, chest x-ray and USG were inadequate for diagnosis in our case and USG yielded a false diagnosis of arteriovenous malformation.

Three main problems may be encountered during the surgical treatment of CRDH. First, venous stasis and hemodynamic impairment in liver and portal areas may occur because of kinked hepatic veins when the liver is placed in the abdominal cavity. Second, the hepatic veins may be injured during the repair of diaphragm, and third, inadequate intraabdominal volume for the liver and other organs after the repositioning may cause difficulties in abdominal wall closure. In our case, none of the above occurred and the diaphragm was successfully repaired. The abdomen was closed surgically without any complications.

When the literature is reviewed, right sided Bochdalek hernias causing obstructive jaundice in neonates and infants are quite rare. In two reported cases, jaundice developed due to stasis in herniated and kinked extrahepatic bile ducts. As no problem was experienced in bile passage after the relocation of organs, no interventions were performed on the bile duct and gallbladder.

On the contrary, bile ducts and gallbladder were highly dilated and thickened in our case. Despite the transfer of organs into the abdominal cavity, folding of extrahepatic bile duct on itself continued to take place and thus, cholecystectomy, hepaticojejunostomy and an end-to-side jejunojejunostomy were performed. Cholic stool were observed on the 4th postoperative day and her total bilirubin levels decreased down to 2 mg/dl on the 8th PO day.

Conclusions

CRDH cases without dyspnea may present with digestive system-related and hepatobiliary findings in later phases. Chest X-rays are a useful diagnostic tool in addition to thorax CT, especially in unclear cases [12]. CT with intravenous contrast agent is a reliable and effective modality for diagnosis. In right Bochdalek hernia cases presenting with obstructive jaundice, kinking of bile ducts should be resolved, immediate diagnosis should be made and fibrosis or cirrhosis due to hepatic bile stasis should thereby be prevented. If the kinking of extrahepatic bile ducts continues despite transfer of organs into the abdominal cavity, cholecystectomy-hepaticojejunostomy is an effective and reliable surgical method. For the closure of diaphragmatic opening, primary repair should be preferred as first-line approach. Reconstruction with patch is chosen only in case of increased abdominal compartment syndrome risk or for cases when a primary repair is technically not feasible.

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