Case report

Congenital diaphragmatic hernia in an 81-year-old female presenting as painless jaundice: A case report

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ABSTRACT

Introduction and importance: Adult congenital diaphragmatic hernia (CDH) is rare and may present as an incidental finding in an asymptomatic individual.

Case presentation: We report the case of an 81-year-old female who presented to the ED with painless jaundice and bilirubin of 19.9 mg/dL. She was diagnosed with biliary obstruction secondary to a right-sided CDH. The patient underwent surgery, which resulted in the correction of her CDH and a downward trend in her bilirubin.

Clinical discussion: CDH usually presents in the newborn period with respiratory distress and can have a high mortality rate. Although CDH is most often associated with newborns, it can rarely present in adulthood, manifesting insidiously with a variety of signs and symptoms ranging from a mild cough to biliary occlusion.

Conclusion: CDH may be considered in the differential diagnosis when other investigations for respiratory or gastrointestinal complaints have been unrevealing.

1. Introduction

CDH occurs when the embryonic origins of the diaphragm fail to properly merge. There are four embryonic tissues that merge to complete the mature diaphragm: septum transversum, the pleuroperitoneal folds (PPFs), muscular ingrowths from the body wall, and the dorsal mesentery of the esophagus. The PPFs make up most of the diaphragm and are the most likely site of malformation, usually due to issues in cell migration, differentiation, and/or proliferation. The etiology of CDH is multifactorial and is not completely understood with a majority of cases being sporadic. Pleuroperitoneal folds should close during the 5th to 7th week of embryonic development, thus without proper development of the diaphragm, an orifice can remain, through which abdominal contents may herniate into the thoracic cavity [1]. The incidence of CDH is 1 in 3000 live births and can be classified based on the location of the defect in the diaphragm. Posterolateral congenital diaphragmatic hernias make up 98% of cases and are called Bochdalek Hernias. Morgagni Hernias make up the last 2%, and are located in the anteromedial diaphragm. Most Morgagni Hernias arise from the right side, with a small percentage being a defect on the left [2].

Ultrasound can be used to diagnose CDH perinatally with an accuracy of 59% and thus treatment can be initiated at delivery [3]. Newborns often present with respiratory distress secondary to lung hypoplasia due to compression from the abdominal viscera. Many cases of CDH have comorbidities that include pulmonary hypertension, hypoplastic left heart syndrome, coarctation, atrioventricular septal defects, and congenital abnormalities [4]. Mortality of infants with CDH is estimated to be 50% [5].

After birth, diagnosis can be suspected when bowel sounds are heard in the thoracic cavity on a physical exam. Diagnosis is confirmed by X-ray and CT, which would demonstrate ectopic viscera within the thoracic cavity [6]. An estimated 1% of CDH patients are asymptomatic and the hernias are often incidental findings in adulthood [5]. Published cases of adults with CDH are extremely rare and we report the case of an 81-year-old woman whose only presenting symptom was jaundice. Following diagnostic studies, an unusual right-sided Morgagni congenital diaphragmatic hernia was discovered.
2. Case report

An 81-year-old female without antecedent primary care presented to the emergency department with a chief complaint of yellowing of her eyes and dark "orange" colored urine over the past 2 weeks. She denied chest pain, shortness of breath, abdominal pain, nausea, vomiting, or any associated symptoms. Social history was negative for alcohol, drug use, smoking, and new medications. There was no family history, relevant genetic information, or psychosocial history to report. The patient's only medications were supplements she had been taking for years: daily vitamin B12 and D3. The patient reported a right-sided inguinal hernia of over 10 years. She had a remote history of hemorrhoidectomy and surgery for a femur fracture. She had no history of abdominal surgeries.

On physical exam, the patient demonstrated no acute distress and was awake, alert, and oriented with no focal deficits. Exam findings were significant for scleral icterus, cutaneous jaundice, mild bilateral lower extremity pitting edema, and a non-reducible right inguinal hernia. Initial labs were remarkable for: bilirubin 19.9 mg/dL, alkaline phosphatase 816 U/L, albumin 3.2 g/dL, AST 186 U/L, ALT 145 U/L, potassium of 3.3 mmol/L, hemoglobin 13.0 g/dL, WBC 7.9 × 1000/uL.

A CT of the abdomen and pelvis with IV contrast (Figs. 1, 2, and 3) demonstrated a right anteromedial diaphragmatic hernia, with the defect measuring approximately 6 cm at the level of the diaphragm. Contents of the hernia sac included the head and uncinate process of the pancreas, portions of the small and large bowel, with a leftward mediastinal shift. The gallbladder, intrahepatic, and extrahepatic biliary ducts were diffusely dilated. Finally, there was a right inguinal hernia containing a portion of the cecum without evidence of compromise.

The patient was admitted to the hospital that evening and gastroenterology and surgery were consulted. When endoscopy was performed, it became clear that an endoscopic retrograde cholangiopancreatography could not be completed because of the patient's altered anatomy secondary to her diaphragmatic hernia. Surgery recommended that magnetic resonance cholangiopancreatography (MRCP) be obtained for a higher resolution and delineation of anatomy. MRI abdomen without contrast MRCP (Fig. 4) showed stricture of the common bile duct distal to the origin of the cystic duct due to mechanical obstruction by the anatomy of the diaphragmatic hernia. There was no evidence of neoplastic change contributing to the biliary obstruction.

On hospital day 2, the patient was started on 40 mg of subcutaneous enoxaparin nightly as DVT prophylaxis, and a 10-day course of 1 g cefepime every 12 h.

On hospital day 6, surgical oncology performed surgical reduction and closure of the CDH followed by reinforcement of the peritoneum and insertion of a chest tube. Incarcerated bowel and contributing adhesions were seen and corrected. Malrotation deformity and Ladd's bands were noted and lysed. A cholecystectomy and an appendectomy were also performed. Surgery was assisted with intraoperative x-ray cholangiogram. The total surgical time was 3 h.

The patient's post-surgical hospital stay was complicated by bilateral pleural effusion, as demonstrated on chest films, transaminitis,
hyperbilirubinemia, and leukocytosis. The chest tube, placed for thoracic drainage intra-operatively, was removed on postoperative day (POD) 4. The patient reported her first bowel movement on POD 6. Final lab values were WBC $10 \times 10^3$/μL, Hgb 8.5 g/dL, bilirubin 6.0 mg/dL, AST 100 U/L, ALT 105 U/L, alkaline phosphatase 781 U/L, albumin 2.1 g/dL, potassium 3.9 mmol/L. The patient was deemed to have reached the maximum benefit of hospitalization with trending of her laboratory values towards reference ranges. She was scheduled for a post-hospitalization follow-up with a newly designated primary care physician to continue monitoring her health and ensure resolution of her abnormal lab values. The patient was discharged on POD 9, hospital day 15 on supplemental oxygen by nasal cannula and the following medications: furosemide, guaifenesin, folic acid, pantoprazole, and potassium chloride.

3. Discussion

Herniation of the abdominal contents through the diaphragm most often occurs in adults due to trauma and rarely occurs from a congenital defect since a majority of CDH cases present with respiratory distress in infancy. Our patient had no history of significant trauma and thus it can be inferred that she had a CDH that slowly progressed and eventually resulted in her presenting to the ED with jaundice. There were no respiratory symptoms and the only concerns were jaundice and dark urine in the preceding weeks. The patient was scheduled for a post-hospitalization follow-up with a newly designated primary care physician to continue monitoring her health and ensure resolution of her abnormal lab values. The patient was discharged on POD 9, hospital day 15 on supplemental oxygen by nasal cannula and the following medications: furosemide, guaifenesin, folic acid, pantoprazole, and potassium chloride.

4. Conclusion

This case report was meant to expand the literature on the adult presentation of congenital diaphragmatic hernias. CDH is usually discovered in the neonatal period and promptly treated due to the severity of symptoms. The incidence of CDH in adults is therefore extremely low and CDH is often an incidental finding or may present with vague symptoms as seen in this patient who presented with jaundice after being asymptomatic for 81 years. CDH is a differential to consider in adults with intermittent respiratory or gastrointestinal symptoms when an alternative source has not yet been identified.

5. Patient perspective

Patient stated that now that her intestines had returned to where they should be, she did not feel comfortable resting her hands on her abdomen, not because it was painful or uncomfortable, rather it was as if her intestines just needed some space and time to adjust. “It’s a hard feeling to explain”. She further states that she is a very active and positive person. “You always have to be positive. Being negative just drains you. And to feel good you have to be surrounded by people who come from God”. That’s why she was very calm and at peace with her life, and did not have any concerns with the procedure because she knew she would do very well.

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Ethical approval

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Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Research registration

N/A.

Guarantor

Shalini Paliwal, M.S.
Credit authorship contribution statement

Shalini S. Paliwal: Conceptualizing, research of literature, writing, and editing.
Bradley M. Lewis: Research of literature, writing, and editing.
Henry Kaufman IV: Editing.

Declaration of competing interest

No conflicts of interest or outside contributions to disclose for all authors.

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