Central Congenital Diaphragmatic Hernia Presenting As Intestinal Obstruction In A 73 Year Old

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Abstract

Presentation of Congenital Diaphragmatic Hernia (CDH) in adulthood is rare. Further, most cases are attributable to Bochdalek and Morgagni hernias with central tendon hernias being particularly uncommon. This is a case of central Congenital Diaphragmatic Hernia in a 73 year old woman who presented with large intestinal obstruction and respiratory failure. Open repair of the hernia was performed which brought about resolution of her symptoms. This case report highlights the possibility for Congenital Diaphragmatic Hernia to present in adulthood and the importance of early diagnosis and treatment in these situations.

Case Presentation

A 73 year old woman presented to the Emergency Department with a 3 day history of abdominal pain, nausea, feculent vomiting, constipation and dyspnoea. She had a past medical history of hypertension, uterine fibroids and osteoarthritis. On presentation, she was hypoxic with a peripheral SpO2 of 88%. Her respiratory rate was 20 breaths per minute, the heart rate was 106 beats per minute, blood pressure 111/82 mmHg and temperature 37.3°C. The respiratory examination revealed a central trachea with reduced breath sounds in the right hemithorax. The abdominal examination revealed distension but otherwise no tenderness, guarding or palpable masses.

An antero-posterior radiograph of the chest (Figure 1) showed haustrations in the right hemithorax suggestive of large intestine in the chest. A subsequent CT (Figure 2) confirmed herniation of the hepatic flexure of the colon as well as the right lobe of the liver into the right hemithorax through a defect in the central tendon of the diaphragm measuring 5.5cm × 6cm. The intra-thoracic colon was grossly dilated with radiographic features of large intestinal obstruction, and there was compressive atelectasis of the right lung with minimal deviation of the mediastinal structures to the contralateral side.

The patient underwent laparotomy with reduction of the herniated structures into the abdomen and primary closure of the diaphragmatic defect (Figure 3).

On follow up, 6 weeks post-operatively, the patient had recovered well with no complaints.

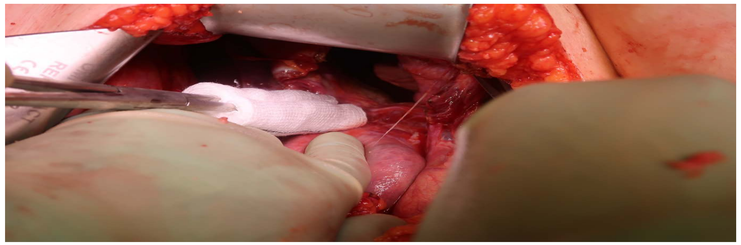


Figure 3a: Abdominal (inferior) view of the defect. The right lobe of the liver can be seen in the thoracic cavity



Figure 1: Antero-posterior radiograph of the chest on admission. Haustrations are apparent in the right hemithorax and there is compressive atelectasis of the right lung

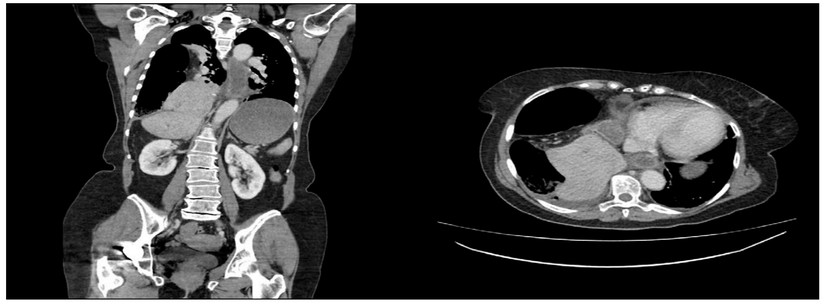


Figure 2: Coronal and transverse CT showing a large defect in the central tendon of the diaphragm through which the right lobe of the liver and the hepatic flexure of the colon have herniated into the right hemithorax. The herniated colon is grossly dilated and there is shifting of the mediastinal structures towards the left

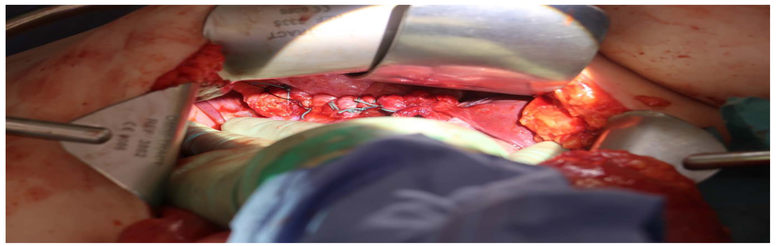


Figure 3b: Abdominal (inferior) view of the diaphragm following closure of the defect with sutures

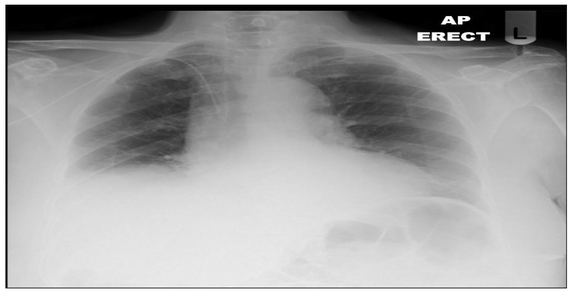


Figure 4: Post-operative antero-posterior radiograph of the chest. The previous haustrations in the chest as seen in the pre-operative radiograph are no longer apparent and the right lung has re-expanded

Discussion

Congenital Diaphragmatic Hernias occurs in 1/5000 – 1/3000 live births and results from abnormal embryonic development of the diaphragm; specifically in abnormalities in development of the pleuroperitoneal membranes which is normally completed by the 12th week of gestation (1). CDH is broadly classified according to three sub-types namely posterolateral (Bochdalek), anterior (Morgagni) and central. Posterolateral CDH is by far the most common accounting for 70%-75% of all CDH followed by anterior CDH accounting for 20%-25% and lastly central CDH represents just 2%-5% of all CDH cases (1). Central CDH refers to a defect in the central tendon of the diaphragm.

Presentation of CDH is usually in the neonatal period as it may result in pulmonary hypoplasia and persistent pulmonary hypertension of the newborn. Presentation in adulthood is rare but when it does occur most often causes abdominal and/or chest pain, vomiting, dysphagia or dyspnoea. Central CDH is particularly uncommon and there have been only two such cases presenting in adulthood reported to date (3, 4).

Conclusion

Congenital Diaphragmatic Hernia is rarely diagnosed in adulthood as most cases present in infancy. Most of these cases are attributed to Bochdalek and Morgagni hernias; central diaphragmatic hernia is particularly uncommon. We present a case of central Congenital Diaphragmatic Hernia with consequent herniation of intra-abdominal viscera into the right hemithorax presenting as large intestinal obstruction and respiratory failure in a previously asymptomatic 73 year old.

This case highlights that, whilst uncommon, Congenital Diaphragmatic Hernia can present in adulthood and the differential diagnosis should be considered for patients with gastrointestinal and respiratory symptoms.

Prompt diagnosis and treatment is essential to prevent sequelae such as gastrointestinal strangulation.

References

1. Dumpa V, Chandrasekharan P. Congenital Diaphragmatic Hernia. [Updated 2021 Aug 11]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK556076/
2. Chandrasekharan, P.K., Rawat, M., Madappa, R. et al. Congenital Diaphragmatic hernia – a review. Maternal Health, Neonatology and Perinatology 3, 6 (2017). https://doi.org/10.1186/s40748-017-0045-1
3. Jagad RB, Kamani P. Central diaphragmatic hernia in an adult: a rare presentation. Hernia. 2012 Oct;16(5):607-9
4. Paci M, De Franco S, Della Valle E, Ferrari G, Annessi V, Ricchetti T, Sgarbi G. Septum transversum diaphragmatic hernia in an adult. The Journal of Thoracic and Cardiovascular Surgery. 2005 Feb 1;129(2):444-5