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Congenital Diaphragmatic Hernia, Finding In Dissection Class (Case Report)

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1. Abstract

During the dissection practice, anatomical variants occur, pathological findings or alterations, and structural lesions may be found. This report deals with the discovery of a diaphragmatic hernia on the right side that was found during a dissection practice, in the corpse of a 71-year-old man, whose reported cause of death was severe respiratory failure. However, the anatomopathological findings demonstrate that it is a congenital hernia, that is, the patient was able to survive his entire life with this alteration thanks to compensatory mechanisms, which leaves a great opportunity for discussion and learning in the classroom.

2. Keywords:
Dissection, anatomy, diaphragm, learning,

3. Introduction

You cannot talk about human anatomy without imagining a corpse on a dissection table; For medical teachers and students, dissection is the main tool for the teaching-learning process of anatomy, touch and a direct view of the corpse are essential to appreciate the complexity of the human body, as well as to recognize the amplitude of anatomical variations, since the structures as described in the literature are not always found. Anatomical variants are found in all organisms, but are considered normal at an anatomical level; During the dissection practice, pathological findings or alterations may be found, such as agenesis, structural lesions, and tumors; All of the above offers the opportunity for learning, since it becomes a forum for students to engage in dynamic discussions, therefore we consider it important to share the findings of anatomical variants or alterations to generate awareness of the existence of the same, as corresponds to the following case.

4. Case Presentation

This is the discovery of a diaphragmatic hernia on the right side that was found during dissection practice, in a 71-year-old male corpse, whose reported cause of death was acute respiratory failure and community-acquired pneumonia, as stated in file No. 134-IF17 located in the Department of Innovation in Human Biological Material of the Faculty of Medicine of the National Autonomous University of Mexico. The material used in the present biological report was obtained from the Institute of Forensic Sciences of Mexico City (INCIFO), through an agreement celebrated in 2006 between this institution and the Faculty of Medicine of the National Autonomous University of Mexico (UNAM). The institutional agreement was approved by the Jurisdiction of Mexico City (Of. No. 1908-2006). All the procedures and manipulation of the biological material performed were aligned with the established Regulations and the Federal Health Law in terms of the sanitary control and disposal of human tissues and human corpses, chapter V (DOF 03-26-2014). During the dissection of the thoracoabdominal cavity, it was identified that the lower two-thirds of the right hemithorax was occupied by abdominal viscera, which entered through a 17 cm diameter hole made in the right hemidiaphragm (Figure 1).

Figure 1:

Figure 1: shows the upper surface of the diaphragm (1), and the opening of the hernial ring (yellow dotted line), through which the abdominal viscera, such as the small intestine, cross towards the thorax (3).

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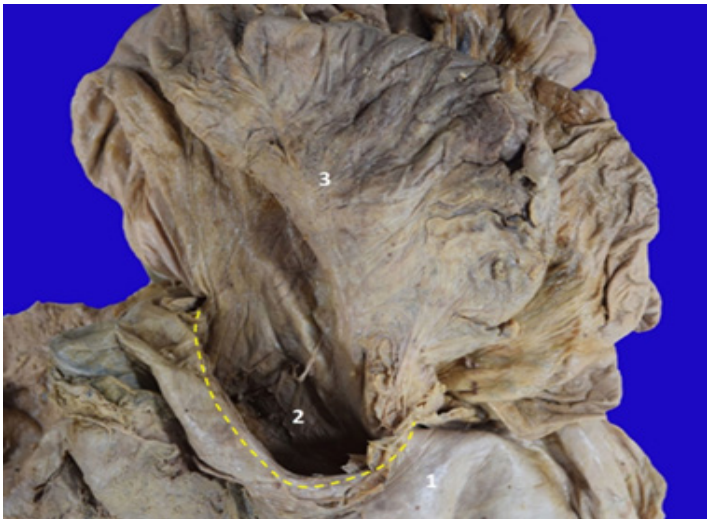

After performing the dissection, the mediastinum compromised with the hernia, as well as the right lung, was reduced in size and partially collapsed, with hypoplasia of the middle lobe, the presence of a fibrous ligament that joins the lung to the diaphragm, the thoracic organs such as the trachea and heart were displaced to the left, by the abdominal viscera, among which were identified: loops of the small intestine, transverse colon, part of the omentum, pancreas, gastric antrum, as well as a portion of the left lobe of the liver (Figure 2). In the right hemidiaphragm, an area of thinning was located next to the hernial ring of fibrous consistency.

Figure 2:

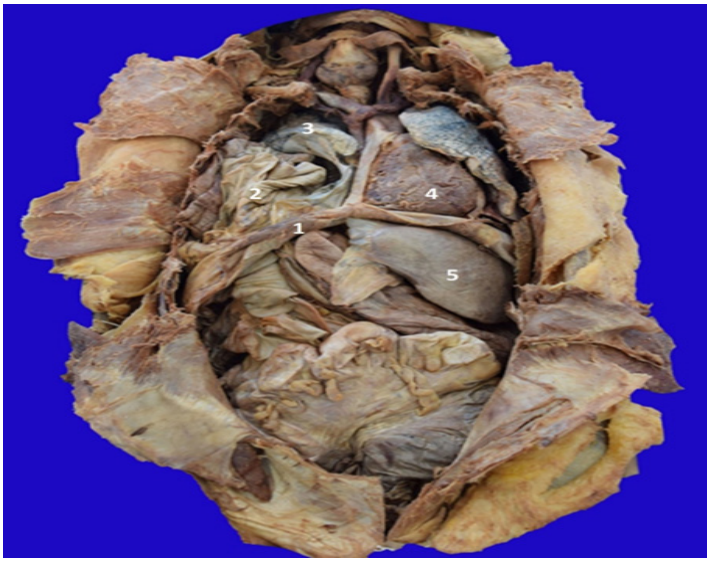

Figure 2: The opening of the thoracic cavity is observed with the organs in situ, we can see the diaphragm (1) crossed by the abdominal viscera (2), which collapses into the right lung (3), which generates in the right hemithorax the displacement of the mediastinum and heart (4). In the abdominal cavity, the liver (5) is located on the left side.

In an inferior view of the diaphragm, part of the gastric antrum, the tail of the pancreas, and the right colon can be seen ascending towards the right hemithorax through the hernial ring. On the other hand, in the left hypochondrium, the liver is observed, as well as the inverted direction of the falciform ligament and the stomach (Figure 3).

Figure 3:

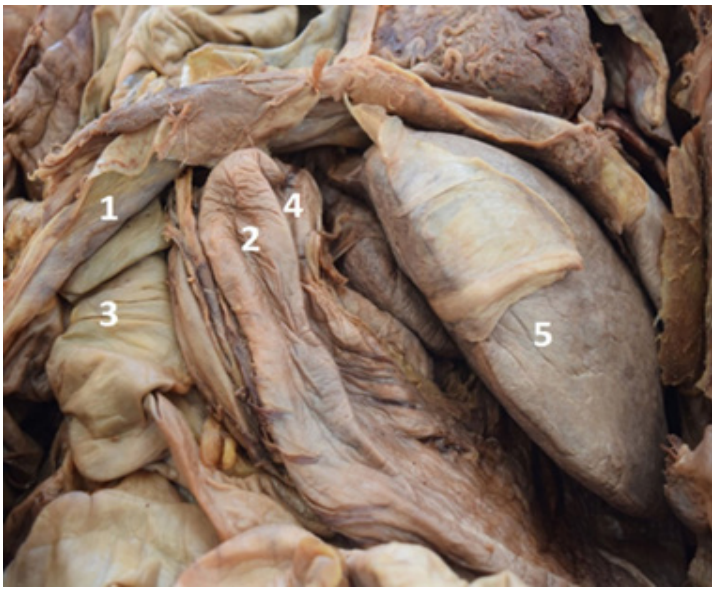

Figure 3: Inferior view of the diaphragm (1), where it can be seen crossed by the abdominal viscera such as the stomach (2), the transverse colon (3), the tail of the pancreas (4), and the inverted situation of the liver and the ligament sickle cell (5).

The histological study of the diaphragm, as well as the hernial ring, showed the following findings: in the left hemidiaphragm not involved with the hernia, it was observed that the thoracic aspect is associated with the parietal portion of the diaphragmatic pleura, while the abdominal aspect is It is associated with the peritoneum, on both sides, underlying these covers, sheets of dense connective tissue with a tendinous appearance are observed where the collagen fiber fascicles maintain a regular arrangement. In the center of the diaphragmatic wall, different fascicles can be seen of large, striated muscle fibers and the presence of some nerve fascicles (Figure 4b). It was also evident that near the annulus fibrosus, the density of striated skeletal muscle fibers decreases.

Figure 4:

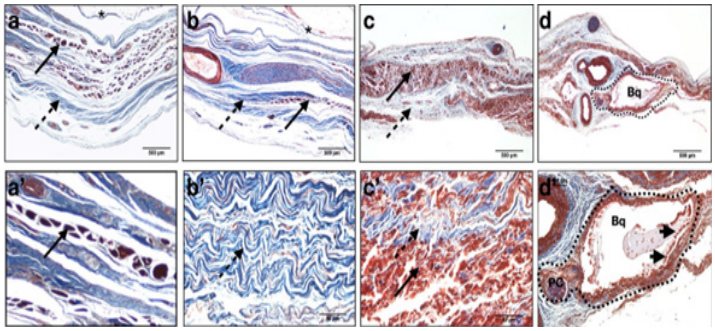

Figure 4: Histological analysis of diaphragmatic wall. Slides were processed with Masson's trichromic stain. (a-a') left hemidiaphragm; (b-b') fibrous ring; (c-c') right hemidiaphragm; d-d') right hemidiaphragm with bronchial penetration. In a-d) Low maginfication images; a'-d') high magnification images. (a-a') The left hemidiaphragm shows on the thoracic aspect the parietal pleura (*), a layer of thick striated muscle fibers (solid arrows), and on both sides numerous collagen fibers of regular dense connective tissue with a tendinous appearance (dotted arrows).

(b-b') In the annulus fibrosus, a thinning of the skeletal muscle fiber layer and a predominance of collagen fibers are observed. (c-c') In the right hemidiaphragm, a loss of regularity is seen in the collagen fibers and although the muscle layer is thick, the muscle cells are very small, reminiscent of a state of atrophy. (d-d') In the right hemidiaphragm, in greatest thinning, it is observed that the wall has been invaded by a small-caliber bronchus.

In the case of the right hemidiaphragm, an absence of peritoneum on the abdominal surface, a decrease in tendon fibers, and a muscular layer with atrophied muscle fibers (figure 4b-c) were found. In the region with the greatest thinning of the muscular layer, the incursion of a bronchial-like tubular structure lined with ciliated pseudostratified cylindrical epithelium associated with a small irregular plate of hyaline cartilage is observed (figure 5d). Due to the presentation of the case, it is considered that it is probably a congenital diaphragmatic hernia.

5. Literature Review

Congenital diaphragmatic hernia (CDH) is a failure of the diaphragm to close during development that allows the abdominal viscera to herniate into the thorax (Hedrick & Adzick, 2022; Chatterjee et al., 2020). The global prevalence of this disorder is 2-4 per 10,000 births (Hedrick & Adzick, 2022; Chatterjee et al., 2020; Alghamdi & Raboe, 2021) and represents 8% of all major congenital anomalies (Aihole, 2021). An incidence of CDH between men and women 1.7:1 has been reported (Katsaros et al.,2022). The diaphragm develops by the fusion of four structures during the fourth to tenth week of gestation, the transverse septum (tendinous component), the pleuroperitoneal folds (muscular component), the mesenchyme of the thoracic wall of the body and the esophageal mesentery (crura or diaphragmatic pillars). Incomplete fusion of these structures causes a diaphragmatic defect that results in herniation of the abdominal viscera into the thorax (Hedrick & Adzick, 2022; Chung et al., 2022). The location of the diaphragmatic defect can be on the dorsolateral aspect of the diaphragm, known as Bochdalek's hernia, on the ventral aspect of the diaphragm, known as Morgagni's hernia (Chung et al., 2022; Katsaros, et al 2022) or central (Aihole, 2021), can occur congenitally or secondary to trauma (Ramsport et al., 2021). Approximately 95% of diaphragmatic defects are posterolateral and the rest ventral, and can be located retrosternal or peristernal (Chung et al., 2022). In 80 to 85% of cases, the hernia is located on the left side, and between 10 to 15% on the right side and occurs bilaterally in <2% of cases (Chatterjee et al., 2020; Chung et al. al., 2022; Katsaros2022; Okamoto, et al., 2021; Spiridakis et al., 2021). Left-sided diaphragmatic hernias with mediastinal shift can decrease left ventricular mass, which can lead to hypoplastic left heart syndrome (Hedrick & Adzick, 2022; Chatterjee et al., 2020; Chung et al., 2022).

The type and effect of CDH on the fetus depend on the gestational age when the viscera herniate (Hedrick & Adzick, 2022), in addition to the extent and duration of the herniated organs in the thoracic cavity that inhibit the normal growth of the lungs and result in structural and functional changes in the heart, pulmonary circulation, and lung parenchyma (Chatterjee et al., 2020). CDH can be an isolated abnormality, part of a syndrome, or nonsyndromic but associated with other abnormalities (Hedrick & Adzick, 2022; Chatterjee et al., 2020). Cardiovascular malformations such as tetralogy of Fallot, and atrial and ventricular septal defects, occur in 11%-15% of cases of isolated CDH and 25%-40% of all cases. Lung changes include acinar hypoplasia, decreased terminal capacity with fewer alveoli, thickened walls, increased interstitial tissue, persistent pulmonary hypertension, and pulmonary vascular dysfunction (Chatterjee et al., 2020; Gupta & Harting, 2020). Isolated CDH occurs in 30 to 70% of cases, presenting pulmonary hypoplasia, intestinal malrotation, and dextrocardia. On the other hand, complex, non-isolated, or syndromic CDH has an incidence of 30 to 50% of cases, they are called this way because they are associated with important structural malformations (Hedrick & Adzick, 2022), skeletal, neural, and gastrointestinal alterations. and other genitourinary defects, they can also be part of some syndromes such as Pallister-Killian, Fryns, Ghersoni-Baruch, WAGR, Denys-Drash, Brachman-De Lange, Donnai-Barrow or Wolf-Hirschhorn syndrome (Franco et al., 2015). The size of the diaphragmatic defect can be used to stratify the risk in patients with isolated CDH, but this evaluation can only be performed during surgical repair; these intraoperative findings have been described as defects "A" to "D" (Chung et al., 2022). Where "A", is the smallest defect, usually intramuscular, and is present in 90% of cases and only affects less than 10% of the diaphragm, the defect "B", is present in 50 to 75% of cases and affects less than 50% of the diaphragm; the "C" defect is present in the hemidiaphragm in less than 50% and affects more than 50% of the chest wall and the "D" defect is the largest, previously known as diaphragmatic agenesis, there is a complete or almost complete absence of the diaphragm; It is present in less than 10% of cases and affects more than 90% of the chest wall. All "D" defects require a patch (or muscle flap) for repair (Chung et al., 2022).

Most CDH (60% of cases) is diagnosed prenatally within routine ultrasounds; on average at 24.2 weeks of gestation (Chatterjee et al., 2020; Chung et al., 2022), or as part of screening for polyhydramnios and hydrops fetalis (Hedrick & Adzick, 2022; Chatterjee et al., 2020). At birth, it is diagnosed immediately when there is a respiratory difficulty, a scaphoid or pan-shaped abdomen, intestinal sounds in the chest, and mediastinal changes on the chest x-ray (Aihole, 2021). Bochdalek hernias in adults occur much less frequently, with an incidence of 0.17% (Ramspott et al., 2021; Chen et al., 2019); In most cases, they are asymptomatic (Spiridakis et al., 2021), women are affected more frequently than men, the average age of presentation in them is 58 years. A sudden or prolonged increase in

intra-abdominal pressure, in cases that include chronic cough, constipation, pregnancy, and trauma in the adult population, can develop Bochdalek hernias (Katsaros et al., 2022). In 5 to 10% of affected individuals, signs and symptoms of diaphragmatic hernia may include intermittent, acute, or chronic respiratory or gastrointestinal problems (Ramspott et al., 2021; Chen et al., 2019; Spiridakis et al., 2021).

6. Discussion

It is necessary to make the distinction between CDH and diaphragmatic eventration (DSE), defined as an abnormal elevation, either partial or total, of the diaphragm without a break in its continuity and with a displacement of the viscera from the abdominal cavity towards the thorax (Guzmán-Valderrábano et al.,2017). Only a few cases of CDS require urgent surgical treatment in the neonatal period, while CDH is usually managed by perinatal surgery. Classic CDH without a sac is an easy diagnosis due to the nonencapsulated abdominal contents within the chest and displaced mediastinum. It is crucial to check if any part of the diaphragm does not have muscle fibers or extracellular matrix; we would be talking about CDH; To evaluate this, immunohistochemistry techniques for desmin, or Elastic van Gieson for collagen, are especially useful and thus make a differential diagnosis between EDC and HDC; since in CDH with a sac, muscle fibers and collagen fibers will not be present (Inoue et al., 2020). In the present case, there was a clear orifice that allowed the continuity between the abdomen and the thoracic cavity, without any membrane that would make us think of diaphragmatic eventration. The abdominal contents described in the right hemithorax were without the presence of any membranous capsule.

This case of amphitheater that we report was classified as a congenital Bochdalek diaphragmatic hernia on the right side, its incidence in adults is estimated between 0.17% and 6% (Katsaros et al., 2022); The size of the hernial orifice for this anatomopathological case is considerably larger than that reported by other authors, where the average defect is 7.01+ 3.18cm (Katsaros et al., 2022), the findings lead us to consider that it refers to a Classic CDH, with deviation of the mediastinum to the left side, without causing hypoplasia of the left ventricle, although it did cause a decrease in wall thickness; The atrial septal defect such as fossa ovale type ASD that we observed is a minor malformation (Chung et al., 2022), associated with CDH. Other pathologies associated with CDH described in this case are hypoplasia of the middle lobe of the right lung. , considered a Type III pulmonary agenesis, in the classification of pulmonary agenesis modified by Boyden, described as a variable amount of lung parenchyma, bronchial tree and vasculature, the diagnosis of pulmonary agenesis in the fifth decade of life is unusually rare (Tanrivermis- Sayit & Elmali); the histological findings related to the tubular structure with a bronchial appearance in the area of greatest thinning of the diaphragm, was related to the thinned fibrous tissue that joined the lung to the diaphragm and which corresponded to the hypoplasia of the middle lobe; The mechanical compression of the lung in the fetal stage by the abdominal viscera represents the classic paradigm for the etiology of hypoplasia related to CDH (Chung et al., 2022) as was the case we present; The other findings coincide with what was previously reported, transverse colon and stomach are the most frequently herniated abdominal viscera, with 42.7% and 37.1% of cases respectively, followed by small intestine (18.0%), greater omentum (16.9%); In our case, we also found within the hernial contents the tail of the pancreas and a part of the left lobe of the liver (Katsaros et al., 2022); Some authors have used the presence of the latter as a predictive factor at birth for the use of extracorporeal life support, increasing morbidity and mortality (Chung et al., 2022). This report shows that, even in basic areas, anatomy and especially dissection, the teacher and especially the students find a great opportunity for teaching-learning, where it has always been considered an arid and static study, leaving evident that each organism is unique and can offer us countless learning opportunities.

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