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Bochdalek Hernia in Adulthood: A Review and an Autopsy Case Report

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ABSTRACT: Bochdalek hernia is rare in adulthood with fewer than 100 cases having been reported in the world literature. We present a delayed case of Bochdalek hernia in an adult leading to sudden death, first diagnosed at medicolegal autopsy.

KEYWORDS: pathology and biology, Bochdalek hernia, posterolateral diaphragmatic hernia, sudden unexpected death, medicolegal autopsy

Congenital posterolateral diaphragmatic hernia was described in 1848 by Bochdalek, a professor of anatomy, and thus named as Bochdalek hernia [1].

A congenital diaphragmatic hernia results from the failure of the fusion of pleuroperitoneal folds. During the third week of gestation, the diaphragm originates from the septum transversum. The septum transversum migrates posteriorly, it fuses with the dorsal mesentery of the foregut, leaving a defect referred to as a pleuroperitoneal hiatus on each side of the spine. For a short time, the abdominal and thoracic contents may freely communicate through this defect until the pleuroperitoneal folds are fully developed. This process is usually completed by the ninth week of gestation, with the left side being the last to close. If these folds fail to develop completely, a Bochdalek hernia may occur. No sac will be present if failure of fusion occurs early during gestation. If development stops sometime later, a sac may be present, made up of the peritoneum and parietal pleura [2-4]. Anatomy of the diaphragm showing the position of the foramen of Bochdalek is illustrated in Fig. 1.

Bochdalek hernia may present in childhood or adulthood with acute or chronic symptoms [2,5,6]. The incidence of Bochdalek hernia in neonates has been reported as ranging from one in 3600 to one in 7000. In contrast, it is rare in adulthood with fewer than 100 cases reported in the literature [2,6-10]. Males predominate by a 2:1 ratio in the neonatal period and 3:1 in the delayed presentations. The left side predominance of the diaphragmatic defect has been noticed in 70 to 90% of the cases [2,6]. The incidence of hernias with a sac varies from 10 to 38%. Although most hernias are sporadic, some authors have reported

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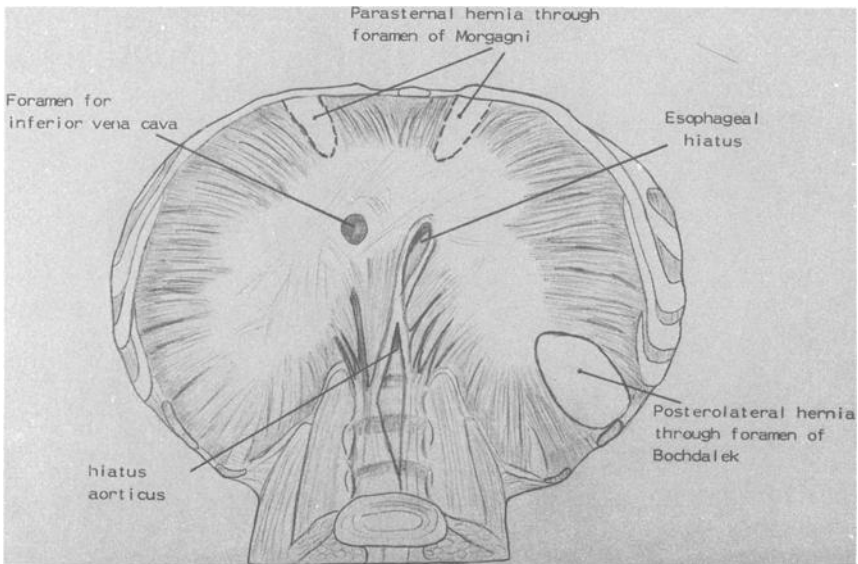


FIG. 1—Anatomy of the diaphragm, showing position of the foramen of Bochdalek.

the occurrence in siblings, twins and two generations of the same family [4,11]. Although most delayed presentations of this defect occur in the first or second decades of life, there have been several reports of patients presenting in the third to eighth decades [6]. In adult 34% of cases, Bochdalek hernia is only diagnosed when the acute complications occur. The others are only detected during health survey programs [2,11].

The clinical presentation in adulthood differs greatly from that seen in the neonatal period. It has been reported that most of the delayed cases have gastrointestinal tract symptoms and chest pain with no evidence of pulmonary disorders. Because of this, the diagnosis of delayed cases is still a problem and the cases reviewed in the literature have highlighted the misleading symptomatology and also the lethal consequences of a missed diagnosis [1-12].

Some of the delayed cases have been reported after trauma, gunshot wound, sexual intercourse, physical exertion, ingestion of a large meal, during pregnancy, labor and childbirth, or as a result of sneezing or coughing [2,5,9].

The reason of the late manifestations of lethal defects in the adult is not known. Some authors believe that a confining sac may allow the patients to survive and a later rupture may trigger symptoms [4,5]. In others, it has been suggested that occlusion of the diaphragmatic defect by the liver or spleen prevents herniation in many cases, and acute herniation occurs with increased intra-abdominal pressure [2,6,9].

In the Bochdalek hernia the contents of the chest vary a great deal; on the left side the cavity is usually filled with small intestine, the spleen, stomach, left lobe of the liver and most of the colon. Hernia on the right side usually contains liver and variable amounts of small and large intestine. Uncommon hernial contents include gallbladder and kidney on the right side, pancreas and adrenal gland on the left side. The lung on the side of hernia is atelectatic and compressed, the weight of the both lungs are markedly decreased [4].

Congenital hernia has a high mortality rate in neonates even after treatment, however surgical treatment of hernia in adults is beneficial when it is diagnosed before the occurrence of complications [13]. Incarceration is the most potentially life-threatening situation, with serious complications such as small and large bowel ischemic necrosis, perforation, gastric

gangrene, splenic infarction, colopleural fistula, obstructive jaundice, and acute respiratory failure with hypoxemia and shock [7,9,10,12].

Case Report

A 23-year-old man was in good health until the morning of his death. The day before his death, he started to complain of chest and abdominal pain after his regular daily sports activities. He went to a private clinic and appendicitis was diagnosed. He was sent to the emergency service of a local hospital and received classical critical care. After a period, the symptoms disappeared and he was sent home for bed rest. Appendicitis was considered to be a misdiagnosis. Early on the following day, he was unconscious when he was found in the bathroom.

He was taken to the local hospital but died on the way to the hospital. His death was considered sudden and unexpected. He was referred to the prosecutor as medicolegal case and his body was sent to The Adana Division of Council of Forensic Medicine of Turkey for medicolegal autopsy. He did not have a history of drug or alcohol abuse. He had no medical history except a surgical repair of the left inguinal hernia three years previously.

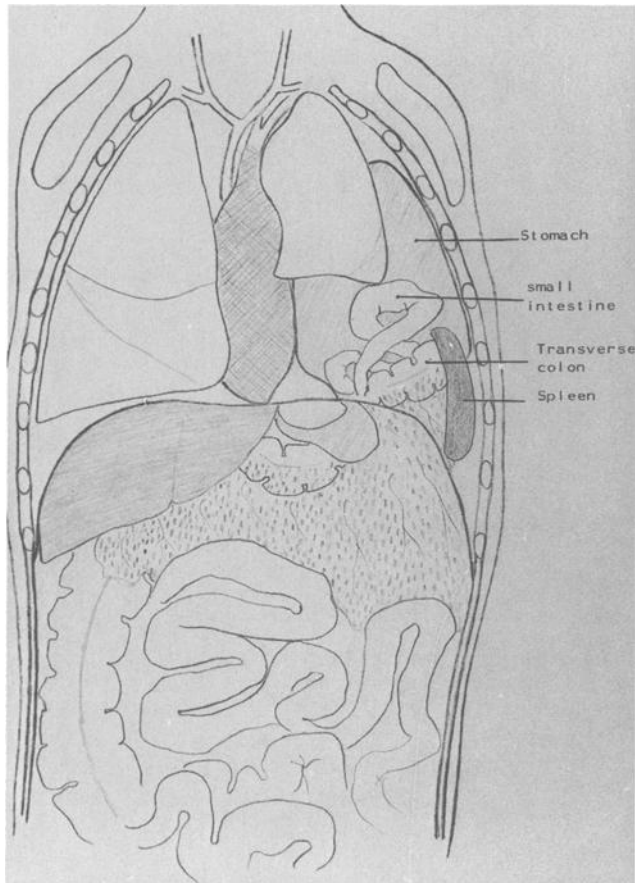


FIG. 2—Schematic representation of the chest and the abdominal cavity showing herniated viscera.

Autopsy Findings

The autopsy examination revealed a generally well developed, well nourished man. He had no skin lesions or evidence of trauma. He had a surgical scar on the left inguinal region, 5 cm in length. Significant findings at autopsy were limited to the chest and the abdominal cavity. When the thoracic cavity was opened, it was seen that the left lung was atelectatic and compressed at the apical region by the abdominal viscera (stomach, small intestines, spleen, transverse colon, and omentum) herniated through the 6 cm posterolateral defect of left hemidiaphragm. There was no sac. The mediastinum and the heart were shifted to the right, and the left side of the heart had become smooth by compression. These findings are illustrated in Fig. 2. The left lung weighed 250 g, and the right, 500 g. The heart was normal size (320 g) and there was no atherosclerosis. But, the chordae tendineae were shortened and the papillary muscles were hypertrophic. After the abdominal cavity was opened and the herniated viscera were repositioned, no remarkable changes were noticed except some small hemorrhages within the mesentery. No chemical and toxic agents or alcohol were revealed by postmortem toxicological analysis.

The cause and manner of death of this man were reported as sudden death due to respiratory failure due to left lung atelectasis and mediastinal shift due to Bochdalek hernia.

Conclusion

We present a delayed case of Bochdalek hernia leading to sudden death in an adult. This case is of particular interest to forensic pathologists since the phenomenon is likely to produce cases of sudden or unexpected death in persons who do not have a significant medical history. In sudden deaths preceded by chest and abdominal pain, among several causes Bochdalek hernia should also be considered. It is obvious that this case would have been misdiagnosed without autopsy.

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