A rare case of adult Bochdalek hernia in Tanzania: Presentation and review of the functional anatomy and embryological basis

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INTRODUCTION

Abdominal hernias are generally common and may be congenital or acquired. They occur into two directions: inferiorly via the pelvis, anteriorly via the abdominal wall and superiorly into the thorax. While the pelvic and abdominal wall hernias are easily detected and managed, thoracic hernias tend to be insidious and become noticeable when associated with overt symptoms. Anatomically the intrathoracic hernias are classified as hiatal (via esophageal hiatus), anterior parasternal (via Morgagni openings) and Bochdalek hernia [1]. Due to the location of vital organs of the thoracic cavity, massive intrathoracic hernias tend to be associated with severe complications and neonatal death. Small hernias may affect the lung, heart and vital vascular functions in the thoracic cavity. Majority of these cases present signs of severe pulmonary distress, cardiac and upper abdominal dysfunction [2]. In Tanzania, East Africa and indeed Africa at large, there is scant literature regarding the occurrence of Bochdalek hernias or any other congenital diaphragmatic hernias [3,4]. Globally, there is an enormous amount of clinical literature on thoracic hernias but the gross anatomical literature based on cadaveric or postmortem studies are sketchy—which curtails the understanding of the structural and developmental aspects of the anomaly. In the present paper we report a rare occurrence of a massive Bochdalek hernia in an adult male cadaver otherwise died of unrelated disease condition. We also discuss possible basis of embryological development and functional implications of the hernia to the patient.

CASE REPORT

During the dissection sessions of the medical students at the Muhimbili University of Health and Allied Sciences (MUHAS) we observed a massive case of stomach hernia permanently herniated and bulging into the thoracic cavity. The herniation occurred on the left posterolateral part of the diaphragm at the junction of the muscular part and the central tendon (Figure 1). Careful inspection noted that there were no other abdominal viscera such as intestinal loops, spleen or mesentery insinuating into the thoracic cavity (Figure 2). The defect opening was circular and circumscribed indicating a permanent structure from a congenital process. The pear shaped hernia measured about 8.5 cm in length and 4.6 cm in width and breadth indicating a permanent structure from a congenital process. The pear shaped hernia measured about 8.5 cm in length and 4.6 cm in width and breadth indicating a permanent structure from a congenital process.

The present work reports a case of massive Bochdalek hernia observed during the cadaveric dissection classes of the medical students at the Muhimbili University of Health and Allied Sciences (MUHAS) in Dar es Salaam, Tanzania. The work examines the anatomical, developmental and clinical importance a black Tanzanian male cadaver presenting with a massive Bochdalek hernia case. We conclude that CDH may go undiagnosed while causing morbidity to the patients. We also review the literature on the causation, development and functional anatomy.

Key Words: Bochdalek; Hernia; Congenital; Adult

Figure 1) Diaphragm (broad arrows) superior view; (A) descending aorta; (B) Bochdalek hernia (stomach); (C) caval hiatus; (E) esophagus.

Figure 2) Superior surface of the diaphragm (D) showing the massive Bochdalek hernia of the stomach (broad arrows) emerging from a well formed and circumscribed hernia defect (H).
each. Internally the herniated stomach contained chime remnants (Figure 3) indicating a normal digestion functionality during life. No other congenital anomaly was noted in the diaphragm, thoracic cavity, thoracic cavity or abdomen.

Figure 3) Herniated stomach with decomposed chyme (broad arrows); (S) stalk of the herniated stomach; (W) cut wall of the herniated stomach.

DISCUSSION

Bochdalek hernia is a relatively common form of congenital diaphragmatic hernia (CDH) first described by Czech anatomist Vincent Alexander Bochdalek in 1848 [5]. Despite their relatively common occurrence there is scant literature in Africa. Whether the scarcity in the African literature is due to fewer incidences or simply the lack of detection and reporting, it remains to be elucidated. The present case occurred on the left side of the diaphragm which is in agreement with literature in which over 70% of all Bochdalek hernia cases occur on the left side part of the diaphragm [4,6] although a few studies have reported a right-sided predominance [7]. The defect in the present case did not affect the anatomy of the thoracic cavity as heart, lungs and other mediastinal organs all of which depicted a normal disposition. This could be explained by the fact that while the defect was formed early in life, the herniation of the stomach occurred much later after all the thoracic organs had been formed. The multiplicity of embryonic sources of the diaphragm may accelerate the increased occurrence of Bochdalek hernias. The diaphragm develops from four sources namely: the septum transversum ventrally, the pleuroperitoneal membranes laterally, the esophageal mesentery dorsally and the body wall roundabot [8,9]. These four components fuse to form the adult diaphragm and any error in the morphogenesis and fusion thereof may produce a potential defect for herniation. The Bochdalek hernia may also form as a result of the intestines becoming permanently locked into its anatomical location, the Bochdalek hernia case reported in the present work seems to be a result of a failure in the fusion of the pleuroperitoneal membrane and the septum transversum.

Some studies have shown familial trends of cases [6] but a clear genetic predilection is still highly debatable—entailing that there could be some other factors such as environmental which can add to the occurrence of the condition. Studies have shown that Bochdalek hernias and other congenital diaphragmatic hernias are associated with some chromosomal aberrations such as aneuploidy, translocations, deletions, and marker chromosomes in about 30-34% of the cases with the abnormal chromosomal processes possibly affecting the mesenchymal structural development. It is understood that familial inheritance patterns of Bochdalek and other CDH are generally rare and occur at around 2% of detectable cases with autosomal recessive or dominant genes involved [12,13]. Studies employing hybridization methods for example have identified specific candidate genes NR2F2 and CHD2 on chromosome 15q26 [14,16]. Notably there are generally few cases of CDH in the African literature—a situation which can be attributed to low detection and reporting or less occurrence. For example, about 59 cases were reported in South Africa in a period of 13 years studied from 1984-1997 [17] and 8 other cases from Nigeria [18] which indicate that the frequency of occurrence may also be higher in other countries where no reports exist. Environmental and maternal life style factors such as alcohol use during pregnancy and pregestational diabetes have been identified as strong risk factors for CDH occurrence [19] which could otherwise explain a generally low occurrence of the anomaly in Africa especially in the Sub-Saharan countries where women for example are less likely to develop lifestyle-related diseases due to cultural norms. Empirical data from large studies, though, are needed to elucidate whether the low occurrence of the Bochdalek hernia in Africa is due to low reporting, poor diagnosis or whether the anomaly is less common among the black race for example.

CONCLUSION

Bochdalek hernia, like most other types of intrathoracic hernias is relatively common. Unlike other abdominal hernias, these hernias tend to be insidious in nature due to their deep body location and their presentations are usually diverse and present with significant morbidities and mortalities. Clinicians and particularly thoracic surgeons, gastroenterologist and radiologist should take into consideration probable occurrences of these hernias when considering the deferential diagnosis of chronic abdominal, respiratory and cardiac conditions.

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CONFLICT OF INTEREST

None

REFERENCES


