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Case Report

Rare Type 4 Hiatal Hernia: An Incidental Cadaveric Discovery

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ABSTRACT

A hiatal hernia (HH) occurs when a portion of the stomach slides up into the thorax through the esophageal hiatus of the diaphragm. Most of these hernias are acquired and result from factors which increase the intra-abdominal pressure resulting in laxity of phreno-esophageal ligament and an increase in size of the esophageal hiatus. HHs are classified into 4 types. Type-1 HH is the commonest and frequently associated with gastroesophageal reflux disease (GERD). Type-4 HH is rare and is prone to develop mechanical complications such as gastric volvulus and obstruction. Awareness and early diagnosis and treatment of these complications can reduce morbidity and mortality and ensure a positive outcome.

Keywords: Hiatal Hernia, Esophageal Hiatus, Phreno-esophageal Ligament, Gastric Volvulus

INTRODUCTION

The diaphragm is a large, dome-shaped muscle that separates the thoracic cavity from the abdomen and assists in respiration. The esophagus passes through an opening in the diaphragm called the esophageal hiatus, located at the level of tenth thoracic vertebra. This hiatal orifice is elliptical in shape with a slightly oblique long axis and is bounded by muscle fibres that originate from the right crus of the diaphragm[1]. The esophagus and both Vagus nerves pass through it to gain access to the abdominal cavity. A hiatal hernia (HH) happens when the stomach partially or completely pushes into the thoracic cavity through this hiatus. HHs are incidental findings during routine imaging or endoscopy and are typically associated with only minor symptoms such as gastroesophageal reflux or discomfort in the epigastrium[2]. Four types of HH have been described: sliding HH (Type-1) where gastroesophageal junction lies above the diaphragm, paraoesophageal hernia (Type-2) where gastric fundus slides into the thorax but the gastroesophageal junction lies in the abdomen, mixed HH (Type-3) which has components of both type 1 & type 2 hernia and giant HH (Type-4) which involves herniation of major part of stomach into the thorax along with other abdominal structures like omentum, colon, small intestine, spleen or liver[3]. Of these, sliding HH (Type-1) have been found to be the commonest type (95%)[4] and are intimately associated with gastroesophageal reflux disease (GERD)[5]. On the contrary, type 4 HHs are rare, accounting for less than 5% of all cases[6]. In this report, we present a case of rare type-4 HH which was detected during routine cadaver dissection in the department of Anatomy.

CASE REPORT

During routine dissection of the thoracic region in a male cadaver, a large mass was observed in the posterior mediastinum at the lower end of esophagus, just above the diaphragm (Figure-1). The swelling occupied the central part of the posterior mediastinum and did not show any deviation to either side. The swelling was soft in consistency and was covered by a serous membrane. On further dissection, the swelling

was found to contain the fundus and cardia of the stomach along with a part of greater omentum (Figure-2) and was covered by a sac derived from parietal peritoneum (Figure-3). The position of gastroesophageal junction was altered and was found in the thorax within the hernial sac. It was also found that the herniated upper portion of the stomach was twisted along its long axis which led to distortion of the shape and size of the stomach. The twisting of the stomach led to posterior shifting of the greater curvature of the stomach. The greater omentum had partially herniated into the thorax while the rest was still in the abdominal cavity. The position of lesser omentum was unchanged.

The esophagus was found to be slightly tortuous and dilated at its lower end. No constriction or stricture was noted at the gastro-esophageal junction. The esophageal hiatus was located in the muscular part of the diaphragm and was transversely oval in shape and markedly enlarged in size (Figure-4). The hiatus measured 5.8cm transversely and 2.5 cm antero-posteriorly. A band of muscle fibres measuring about 1.5cm in thickness separated the anterior margin of the esophageal hiatus from the central tendon of the diaphragm. There was no evidence of shortening of the esophagus as the herniated portion of the stomach could be readily pushed back into the abdominal cavity.

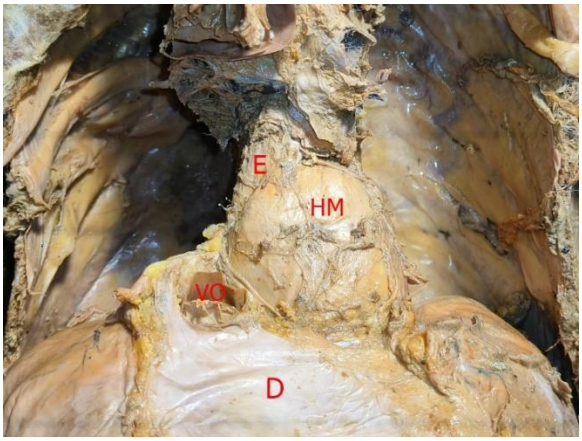

Figure-1: Large hernial mass visible in posterior mediastinum

E: Esophagus; HM: Hernial mass; VO: Vena caval opening of diaphragm; D: Diaphragm

Apart from stomach and greater omentum, there was no herniation of any other associated organs in the thorax. Based on the protrusion of stomach along with greater omentum in the thoracic cavity we would consider this as a type-4 HH. Additionally, two small duodenal diverticula were observed in the first part of duodenum just distal to the pylorus of stomach.

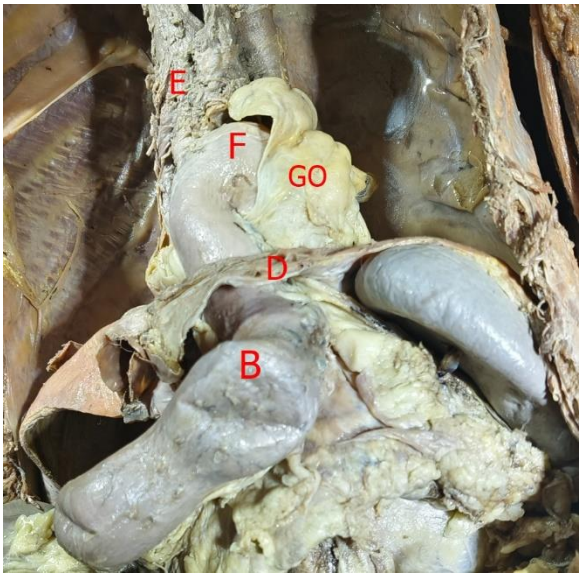

Figure-2: Hernial mass comprising of parts of stomach and greater omentum

E: Esophagus; F: Fundus of stomach; GO: Greater omentum; D: Diaphragm; B: Body of stomach

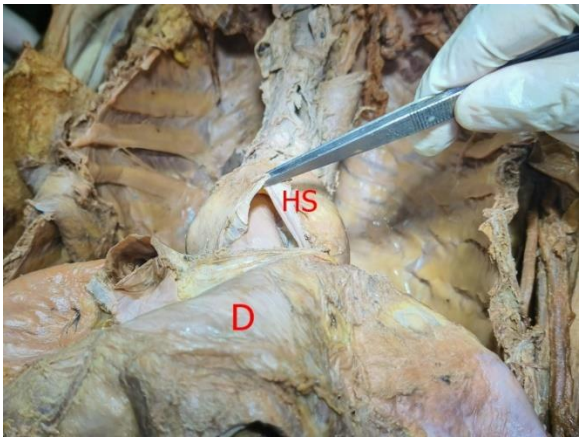

Figure- 3: Hernial contents visible through incised hernial sac

HS: Hernial sac; D: Diaphragm;

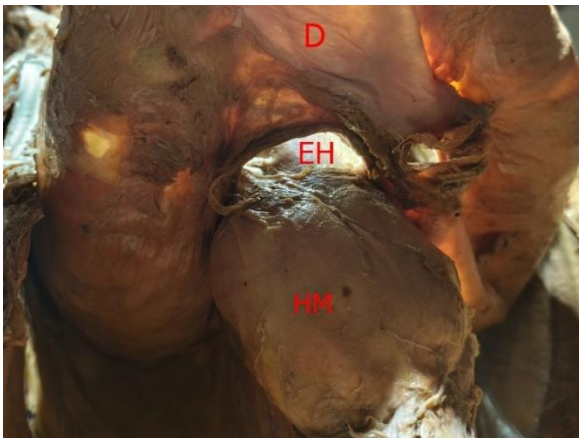

Figure- 4: Thoracic view of hernial mass showing large esophageal hiatus

EH: Esophageal hiatus; HM: Hernial mass; D: Diaphragm

DISCUSSION

HH is the most common type of gastric hernia and its prevalence increases with advancing age. The condition is relatively common in western population with a high prevalence rate (15-20%)[3]. It is usually an acquired condition but rare congenital cases have also been reported[7]. The acquired HH can be further classified as traumatic and non-traumatic. For the non-traumatic HH, the usual etiology is gradual weakening of the phreno-esophageal ligament and enlargement of the esophageal opening in the diaphragm[8]. The phreno-esophageal ligament is derived from fascia on the inferior surface of diaphragm and to a lesser extent from endothoracic fascia and is rich in elastic fibres[1].

This ligament blends with the adventitia of esophagus 2-3 cm above the gastro-esophageal junction and allows some freedom of movement to the esophagus while preventing its upward displacement. With age, the quantum of elastic fibres in the phreno-esophageal ligament diminishes, making the ligament lax and therefore increases the risk of developing HH[9]. In fact, the phreno-esophageal ligament which can be easily identified in young people, becomes attenuated and difficult to detect in old age and in presence of HH[10]. In the present case, a definite phreno-esophageal ligament could not be identified.

The esophageal hiatus in the diaphragm is described as elliptical or tear drop in shape and normally has its long axis in the sagittal plane. However, in presence of hernia, there is an increase in the transverse diameter of the hiatus so that the opening appears more rounded in appearance[10]. Mous A et al[11] reported that in case of HH the esophageal opening may be abnormally widened to 3-4 cm though the upper limit of its normal dimension is 1.5 cm. In the present case, the transverse dimension of the esophageal opening was found to be remarkably wide, measuring 5.8 cm and was more than double of its sagittal length.

The factors that have been hypothesized as predisposing for HH are increased abdominal pressure stressing the phreno-esophageal ligament and esophageal shortening. Increased intra-abdominal pressure may be due to obesity, constipation, urinary obstruction, COPD and heavy exertion at workplace[4]. Central obesity with high intra-abdominal fat, an increasingly common pathology, can lead to hiatal hernia by shifting gastric contents cranially leading to dysfunction of the lower esophageal sphincter. Excess intra-abdominal fat can also cause separation of the all-important diaphragmatic crura due to excessive pressure or stretching[12]. Occasionally, chronic esophagitis can cause fibrosis of the longitudinal muscle coat of the esophagus leading to shortening of the esophagus and development of hiatal hernia[13]. Cases of familial inheritance of HH have also been reported indicating a possible genetic basis[14].

The hernia in the present case revealed involvement of the fundus and cardia of the stomach, esophago-gastric junction as well as a part of the greater omentum, all of which were encased in a peritoneal sac. Type-1 sliding hernia does not have a hernial sac[15]. Only type-2, 3 and 4 HHs are considered as true hernias possessing a hernial sac[16]. The findings thus point towards type 3 or type 4 HH. However, due to the inclusion of part of greater omentum in the herniated contents, we are inclined to consider it as a type 4 hernia. A somewhat identical case of HH involving lesser omentum have been reported in literature where the authors similarly classified it as type-4 hernia[12]. To circumvent this classification conundrum, some scholars in fact have combined type 3 and 4 hernias into a single type, collectively called as mixed HH[17].

Patients with HH tend to develop gastroesophageal reflux disease. This is mainly due to shifting of lower

esophageal sphincter from high pressure zone of abdominal cavity to low pressure zone of thorax which in turn impairs the activity of the sphincter[12]. Bohmer AC et al[18] noted that HH is associated with more frequent occurrence of transient lower esophageal sphincter relaxation, impaired peristalsis and increased acid exposure of the esophagus. Long-standing and erosive GERD can pave the way for further complications like Barrett's esophagus and esophageal adenocarcinoma. Type 2, 3 and 4 HH are more prone to develop mechanical complications[9]. The herniated stomach can undergo rotation around its longitudinal axis as observed in this case resulting in gastric volvulus which in turn may lead to acute gastric obstruction, incarceration and perforation[19].

The HH in this case was found associated with diverticula in the proximal portion of duodenum. Duodenal diverticula are usually asymptomatic. Since the prevalence of both hiatal hernia and duodenal diverticula increases in the later decades of life, they are frequently encountered in association to each other in older individuals[20].

CONCLUSIONS

This report highlights the case of a rare type-4 HH involving upper part of the stomach and a portion of greater omentum. It occurs mainly due to the enlargement of esophageal hiatus of the diaphragm as well as laxity of the phreno-esophageal ligament. Patients with HH usually present with abdominal discomfort or GERD and are routinely managed with medication. But in type-4 HH, the frequent twisting of the stomach as observed in this case increases the probability of developing mechanical complications like volvulus and gastric obstruction and perforation. Thus, awareness and early recognition of HH in patients with GERD and its correct classification as well as appropriate medical/surgical management can reduce morbidity and mortality and significantly improve patient outcome.

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