

Death due to Congenital Transmesenteric Hernia in an Adult: A Rare Autopsy Case Report

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ABSTRACT

Death due to congenital transmesenteric hernias are uncommon in adults. We report a rare autopsy case, of an unmarried 25 years old female who presented with congenital transmesenteric hernia with no previous history of surgery or trauma. Autopsy examination revealed 180 cm of strangulated small intestine herniated through a defect in the small bowel mesentery. This case highlights the importance of considering rare conditions like congenital transmesenteric hernia in the differential diagnosis of acute abdomen and the awareness along with high level of suspicion with prompt advanced imaging and surgical intervention.

Keywords

Autopsy; congenital hernia; death; transmesenteric hernia

INTRODUCTION

According to The World Health Organization (WHO), International classification of diseases version 10 (ICD-10), sudden death is defined as a death which is non-violent and not otherwise explained occurring less than 24 hours from the onset of symptoms¹. The most common cause of sudden death is due to disease of the cardiovascular system whereas, gastrointestinal disease as a cause of sudden death is uncommon². Hernia is the protrusion of viscus from a defect through a wall. Internal hernia is the protrusion of viscus through a defect or aperture, which can be either acquired or congenital, but remains in the peritoneal cavity. Transmesenteric hernia is a form of internal hernia which can be either acquired or congenital. The acquired transmesenteric hernias are common and may present after intra-abdominal surgery or abdominal trauma. On the other hand, congenital transmesenteric hernias are rare and usually seen in pediatric age group.³ Congenital transmesenteric hernia in an adult is even rarer.

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CASE PRESENTATION

A 25-year-old unmarried female, with no history of previous surgery, trauma and abdominal inflammatory diseases, presented to a tertiary level hospital with sudden onset abdominal pain, nausea and vomiting of 1 day. On examination her BP was 90/60 mm of Hg, pulse rate was 96 beats per minute and abdominal examination revealed generalized tenderness, with no signs of peritonism. Her blood count showed elevated leukocyte of 14,000/cu mm with neutrophilia. Abdominal ultrasonography revealed minimal free fluid in the peritoneal cavity and no signs of ischemic bowel. Computed tomography (CT) scan was not available.

She was advised for admission but went home against medical advice. Next morning she had similar symptoms for which she was taken to the hospital and was declared brought dead on arrival. Following which the deceased was brought for autopsy examination in Department of Forensic Medicine, Maharajgunj Medical Campus, Maharajgunj, Tribhuvan University, Nepal.

In Nepal, medico-legal autopsy examination is mandatory in all unnatural deaths and brought dead cases in the hospital. Hence, medico-legal autopsy examination was carried out as per law. External examination was unremarkable. Internal examination revealed 500ml of blood mixed fluid in peritoneal cavity. Loops of small intestine measuring 180 cm comprising part of jejunum and ileum were found herniating through a mesenteric defect of 4 cm x 3 cm. The herniated loops of intestine were strangulated and appeared dark red in color and were filled with dark reddish blood, as shown in Figure 1, 2 and 3. The cause of death was given as strangulated congenital transmesenteric hernia.

DISCUSSION

The present case of sudden death due to congenital transmesenteric hernia is probably the only documented autopsy case till date in Nepal. In developed world such cases rarely land up in mortuary due to their advancement in diagnostic facility and prompt surgical intervention in the hospital. In this particular case, limited access to advanced diagnostic facilities and insufficient counseling regarding the potential severity of the condition may have contributed to the unfortunate outcome. Autopsy studies around the world report a 0.2 - 0.9 % incidence of internal hernias causing bowel obstruction in 4.1 % of all cases of internal hernias³.

Internal hernias are the protrusion of viscus through a defect or aperture present in the mesentery or peritoneum which could be either congenital or acquired.

Acquired hernias constitute majority of the hernias



Figure 1. Strangulated loops of intestine

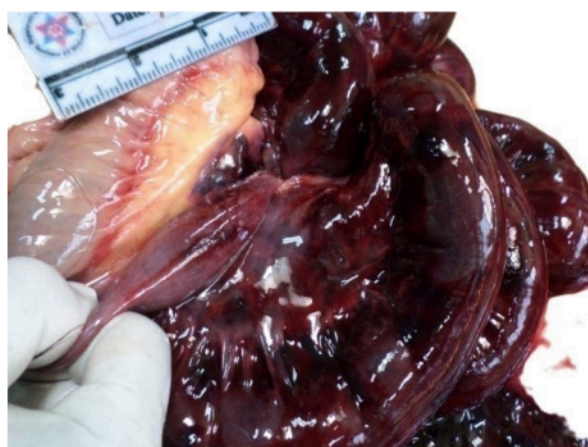


Figure 2. Hernia with narrow neck

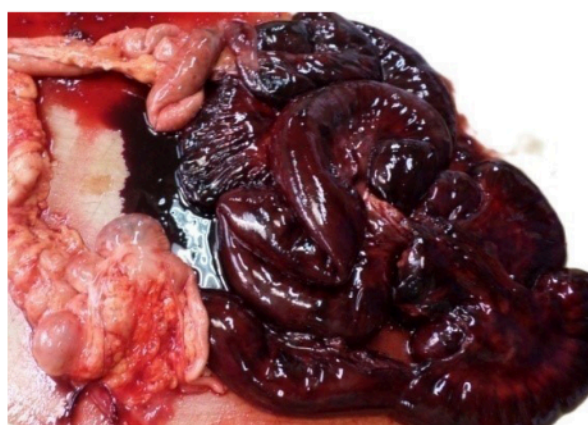


Figure 3. Strangulated and non-strangulated loops of intestine

which occur due to intra-abdominal surgeries.^{2,3} Congenital intra-abdominal hernias (CIAH) can be either retroperitoneal or formed from congenital

anomalous openings. Retroperitoneal hernias can be subdivided into para-duodenal (30–53% of CIAH), Winslow's foramen (8% of CIAH), para-cecal (6% of CIAH) and inter-sigmoid hernias (5% of CIAH), whereas hernias formed from congenital anomalous openings can be categorized as transmesenteric (5–10% of CIAH), broad ligament (4–7%) or trans-omental hernias (1–4%). Transmesenteric hernias (TMHs) are the most common internal hernias in children, and are mainly caused by openings in the mesentery of the small bowel (71% of TMH) and less by meso-colic defects (26% of TMH).^{1,4} The literature on CIAH is sparse with most cases being reported in children whereas cases in adults are even rarer.^{5,6}

Based on above findings this is a case of strangulated congenital transmesenteric small bowel hernia. The morbidity and mortality largely depend on factors such as the time of onset, clinical severity, the patient's co-morbidities, and vascular compromise. Congenital transmesenteric hernia is indeed a serious condition with high mortality rate of up to 50 %, if left untreated.^{7,9} Internal hernias can be either congenital or acquired. Congenital transmesenteric intra-abdominal hernia in adults is a very rare entity and sudden deaths from such hernia in adult are even rarer.^{7,8}

Internal hernia is difficult to diagnose because of its non-specific clinical presentations and lack of specific radiological and laboratory findings to confirm. Internal hernia presents with colicky pain abdomen (epigastric, or generalized), nausea, vomiting, and other signs and symptoms of intestinal obstruction (not passing stool and flatus, abdominal distension). It can even be asymptomatic initially and become symptomatic only when the intestine gets strangled leading to ischemia of herniated intestine. Rarely, it can present with sudden death.^{7,8}

Ideally, internal hernia should be diagnosed pre-operatively, but due to lack of specific radiological and laboratory investigation the diagnosis is difficult leading to misdiagnosis. On the other hand, the morbidity and mortality increase with delay in diagnosis, as it leads to bowel ischemia and gangrene. One of the best methods of diagnosing internal abdominal hernia is CT scan of the abdomen.

CONCLUSION

Death within two days of presentation due to a rare gastro-intestinal condition like congenital transmesenteric small bowel hernia is rare event. These findings underscore the importance of considering even the most unlikely diagnosis.

Unexplained abdominal pain definitely warrants thorough investigation and advanced imaging techniques. Timely and advanced imaging techniques along with high level of suspicion can make a huge difference in the patient care.

CONSENT

Written informed consent was taken from the patient's guardian for the case report publication.

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CONFLICT OF INTEREST

The author(s) declare that they do not have any conflicts of interest with respect to the research, authorship, and/or publication of this article.

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