CASE REPORT

A Rare Case of Bilateral Morgagni's Hernia with Intestinal Obstruction and Malrotation of the Gut in an Adult Patient with Severe Osteogenesis Imperfecta Presenting as Severe Respiratory Distress

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Abstract Osteogenesis imperfecta (OI) is associated with congenital inguinal hernia that can occur due to genetic mutation. Morgagni's hernia is a rare congenital diaphragmatic herniation and is usually diagnosed in childhood. We present a case of an 18-year-old male presenting with bilateral Morgagni's hernia with intestinal obstruction with osteogenesis imperfecta with malrotation of the gut.

Keywords Morgagni's hernia · Osteogenesis imperfecta · Malrotation of gut · Mesh repair

Introduction

Congenital diaphragmatic hernia of Morgagni's type is an uncommon birth defect that is associated with considerable morbidity and mortality. The average prevalence of congenital diaphragmatic hernia (CDH) was 1/4000 births. Osteogenesis imperfecta (OI) is a group of heterogenous conditions characterized by a varying degree of bone fragility. It is a heterogenous mutation in either type 1 collagen genes and it is inherited in an autosomal dominant manner. Mutations in the COLI

A1, COLI A2, CRTAP AND LEPRE 1 genes are known to cause OI [1, 2]. OI affects an estimated 7/100,000 people worldwide. It is rare for this condition to be present in adulthood. It has been reported that the incidence of malrotation in adults is approximately between 0.00001 % and 0.19 % [3–5] Figs. 1 and 2.

Case Report

An 18-year-old male, a known case of severe osteogenesis imperfecta with restricted mobility presented with sudden onset of respiratory distress and vomiting. He had vomiting and breathlessness for 3 days prior to reaching our hospital. Due to severe respiratory distress and clinical suggestion of the right lower lobe pneumonia, patient was intubated and ventilated. Chest X ray showed the right lower lobe replaced by solid and cystic areas suggestive of cystic adenomatous malformation (CAM) or diaphragmatic hernia. Due to diagnostic uncertainty, a CT scan was performed which confirmed Morgagni's hernia with intestinal obstruction. Patient was optimized and emergency laparotomy was performed. On opening of the abdomen, patient was found to have bilateral Morgagni's hernia—right larger than left side. Most of the small intestine and large intestine had herniated into the right side leading to obstruction but without evidence of strangulation. After reducing the hernia, he was noted to have partial malrotation of the gut with the caecum on the left side. Both Morgagni's herniae were repaired with No. 1 Prolene and reinforced with Prolene mesh. Prophylactic appendectomy was also carried out to avoid future diagnostic problem. Patient made rapid recovery after surgery and could be weaned off the ventilator and extubated on the 3rd post-op day. He was discharged on the 11th post op day.

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Fig. 1 Chest X ray pre-op with bony deformities of hand

Discussion

Morgagni's hernia is a rare diaphragmatic hernia and usually presents in childhood. Association of malrotation of the gut with Morgagni's hernia in an adult patient with severe osteogenesis imperfecta and presenting as pneumonia needing ventilator support is still a rarer entity. Patient with Morgagni's hernia may be diagnosed by a simple chest X ray, but in some other cases, a CT scan of the chest and abdomen is necessary [6–8].

Presence of severe osteogenesis imperfecta further complicates perioperative management including anaesthetic techniques and post operative recovery in these patients. In our case, we performed a laparotomy and the defect was closed with Prolene mesh after reduction of the contents. Presence of malrotation of the gut with the caecum lying on the left side warranted prophylactic appendicectomy. Fortunately, the patient made slow but uneventful recovery.

Conclusion

Osteogenesis imperfecta is a genetic disorder so along with the care of bones patient should be evaluated for the chances of occurrence of diaphragmatic hernia and diagnosed earlier to reduce the mortality and morbidity associated with it.

Fig. 2 Intra-op pic B/l Morgagni's hernia

Malrotation of the gut is most of the time asymptomatic; hence, when or if diagnosed, should be treated if possible or left alone.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interests.

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