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Case Report

Laparoscopic correction of midgut volvulus due to malrotation in a patient with Marfans syndrome

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ABSTRACT

This case describes 13-year-old female with Marfans syndrome who presented recurrent small intestinal obstruction which was later diagnosed as acute midgut volvulus due to malrotation after extensive workup. Laparoscopic Ladd's procedure was performed. Patient had uneventful recovery and discharged on post-operative day five. To best of our knowledge the index case is the first case of laparoscopic ladds procedure in known case of Marfans syndrome.

Keywords: Marfans syndrome, Laparoscopic Ladd's procedure, Intestinal malrotation, Midgut volvulus

INTRODUCTION

Marfan syndrome was initially classified by a triad of: skeletal changes- includes long thin extremities, frequently associated with loose joints, decreased vision-result of dislocation of ectopia lentis, cardiovascular abnormalities-aortic aneurysms etc.[1] But there is uncommon association with gastrointestinal manifestations.[2] Midgut volvulus is a condition in which the intestine becomes twisted as a result of malrotation of the intestine during embryonic development. Malrotation of the intestine occurs when the normal embryological sequence of bowel development and fixation is interrupted.[3] Primary midgut volvulus has no known cause while in our case, intestinal malrotation, secondary midgut volvulus has known etiology.[2] No exact data on gastrointestinal complication in marfans syndrome are available. Albeit the etiology of volvulus and clinical features will remain same like vomiting, abdominal distension and pain.[2] Described below is the clinical presentation, workup and management of our patient with midgut volvulus.

CASE REPORT

This case report presents a 13-year-old female with Marfans syndrome with cardiovascular manifestations including aortic root dilation and ocular manifestations-chorioretinal atrophy. The patient was admitted to emergency with acute onset of colicky pain in epigastric region and peri umbilical region, abdominal distension, non-bilious and non-bloody vomiting for last 2 days. Patient had normal bowel movements 24 hours prior to admission. Patient experienced similar episodes in the past. On per abdomen examination, patient had mild distension and no signs of peritonitis on palpation and bowel sounds were exaggerated. She was hemodynamically stable on admission. Her complete blood profile, liver function tests (LFTs), renal function tests (RFTs) were normal except raised serum alkaline phosphatase. X-Ray FPA showed multiple air fluid levels. Computed tomography (CT) scan suggested inversion of superior mesenteric artery and superior mesenteric vein relationship with SMA on right and SMV on left side. This is the reliable finding for diagnosis of malrotation.5 There is clockwise rotation of SMV around SMA axis giving a classical "whirlpool sign" appearance (Figure 1). Stomach and proximal duodenum are over distended with twisting of the distal duodenum and dudeno-jejunal junction along with SMV. Hence the diagnosis of acute midgut volvulus was made.

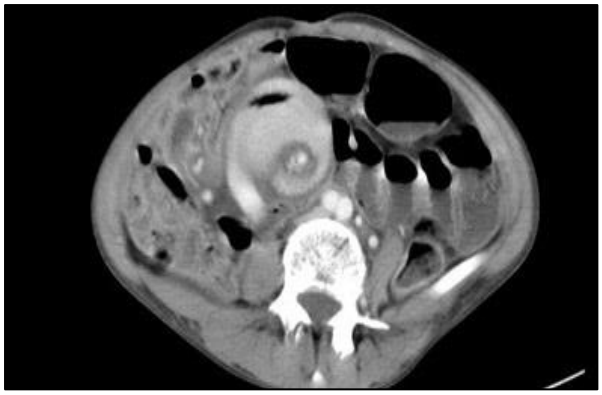

Figure 1: Clockwise rotation of SMV around SMA axis giving a classical "whirlpool sign" appearance.

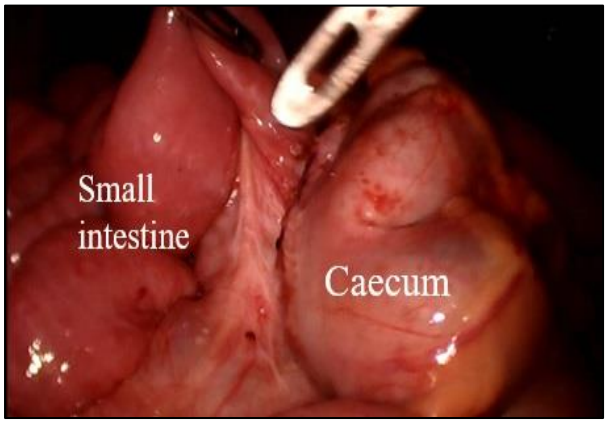

Figure 2: Small intestine was mobilized to right while large intestine to left.

Diagnostic laparoscopy was done 36 hours after admission (5 ports were created) which showed twisted small intestine loops with clockwise twisting of midgut with ladd's band (caecum to ascending colon). There were no signs of ischemia.3 Then Laparoscopic Ladd's procedure was carried out; counter clockwise detorsion of the midgut volvulus done, division of Ladd's bands overlying the duodenum with widening of the narrowed root of the small bowel mesentery by mobilizing the duodenum and division of the adhesions around the SMA to prevent further volvulus with appendicectomy. Small intestine was mobilized to right while large intestine to left (Figure 2). Other abdominal viscera were found to be normal. The procedure was two hours long. Postoperatively patient's recovery was uneventful. She was discharged on postop day five. This is the first reported case of marfans syndrome having midgut volvulus due to malrotation treated by laparoscopic approach.

DISCUSSION

Marfans syndrome is an inherited connective tissue disorder with mutation in Fibrillin gene (FBN1) on chromosome 15. Fibrillin is a glycoprotein which have structure similarity with TGF-β binding proteins that sequester TGF-β in the extracellular matrix. As a result, some the manifestations of MFS have been shown to arise from alterations in binding sites that modulate TGF-β bioavailability during development of skeleton and other tissues.1 Fibrillin is responsible for production of elastic microfibrils which are in turn manage availability of TGF-β (manages production and apoptosis of cells). Mutation is Fibrillin gene results in impairment of connective tissues throughout the body leading to disorder of various systems of body.[6]

In our patient, there was involvement of cardiovascular system (aortic root dilatation), skeletal system and ocular system with gastrointestinal manifestations. Review of the literature showed only a few reported cases of small bowel diverticulosis, particularly of the jejunal area and some cases of sigmoid volvulus and one documented case of primary midgut volvulus in 80-year old female in conjunction with a diagnosis of Marfans syndrome.[7,8] Incidence of malrotation is estimated to be 1 in 6000 live births. 64-80% of malrotation cases present in the first month of life and 90% within the first year. Adult presentation is very rare accounting for only 0.2-0.5% of cases, of which only 15% present with midgut volvulus.[14] There are no reported cases of midgut volvulus due to malrotation in a patient with Marfans syndrome. We used laparoscopic approach for correction which showed favourable results as there were no complications, early initiation of oral intake, early return of peristalsis and early discharge. Literature (case series and case reports) suggested the same safety and excellent outcome of this procedure while others disagree and consider same as standard open technique (as there is large operating time and laparotomy remains favoured for patients presenting with volvulus).[10-12] Midgut volvulus due to malrotation is common in paediatric population but it is rarely found in conjunction with Marfans syndrome.

CONCLUSION

Marfans syndrome is a multisystem inherited connective tissue disorder affecting mainly three systems cardiovascular, skeletal and ocular but uncommon with gastrointestinal manifestations. To our knowledge this is the first case of midgut volvulus due to intestinal malrotation associated with Marfan syndrome treated laparoscopically. Acute midgut volvulus is an emergency, urgent exploratory laparoscopy/laparotomy with resection of gangrenous bowel should be done. In doubtful viability bowel preservation with relook laparotomy is very wise decision. Chronic recurrent midgut volvulus requires high index of suspicion, CECT abdomen is investigation of choice, symptomatic cases should be managed with laparoscopic Ladd's Procedure. Laparoscopic surgery is better than open technique as there were no postoperative complications and early return to normal lifestyle.9,10,13 This case report will facilitate further studies in gastrointestinal manifestations in Marfan's syndrome and its management laparoscopically.

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