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Colonic Volvulus in Patient with Marfan Syndrome: Rare Gastrointestinal Presentation

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The Marfan syndrome (MFS) is a pleiotropic, autosomal dominant disorder of connective tissue with highly variable clinical manifestations [1]. It primarily involves the skeletal, cardiovascular, and ocular systems; however, gastrointestinal presentation (GP) are rare [2]. We present an unusual case of colonic volvulus in a patient with Marfan syndrome and with abnormal anatomy of various segments, malrotations and colonic volvulus. A 62-year-old male presented to the emergency department of our medical center with a four-day history of abdominal pain. He has a history of coronary artery bypass graft, MFS, and pre-existing gastrointestinal disturbances, in fact about a month ago in another center following a period of intestinal obstruction from colonic volvulus he underwent a colonoscopy that reduced the volvulus. Physical examination revealed hypoactive bowel sounds and diffuse abdominal tenderness with rebound with no abdominal rigidity. Plain abdominal radiograph demonstrated a greatly dilated sigmoid colon that almost filled the entire abdomen. A computed tomography (CT) scan of the abdomen revealed a distended colon without haustral markings (Figure 1). He underwent left hemicolectomy with colo-colic anastomosis (Figure 2). Intraoperatively we found a transverse colon and a dilated left solon with a maximum diameter of 15 cm, abnormal interlocking between told and gerota, floppy tissues, with rotation of the large intestine in two points (Figure 3). The postoperative course was uneventful and patient was discharged 6 days after surgery. GP is not commonly found in patients with Marfan syndrome compared to higher occurrences of cardiovascular, ocular and soft tissue pathology. GP are infrequent and there are only a few cases reported of differing diagnoses, most of the cases presenting with abnormal anatomy of various segments of the gastrointestinal system such as diverticulosis, hernias, atresias, malrotations and sigmoid volvulus [3].



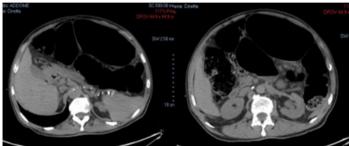


Figure 1: Sagittal and coronal images showing colonic distention, especially of the transverse and descending colon

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Figure 2: Surgical piece with loss of the typical austrature of the colon



Figure 3: Detail intra-operative with rotation

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