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CONGENITAL PERITONEAL BAND CAUSING TRANSVERSE COLONIC OBSTRUCTION, MASQUERADING AS SIGMOID VOLVULUS AT INITIAL PRESENTATION IN A PEDIATRIC PATIENT WITH DIGEORGE SYNDROME: A CASE REPORT.

Radiology

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ABSTRACT

We present a case of a 4-year-old child with DiGeorge syndrome who presented to the emergency department with abdominal distention and recurrent vomiting. Initial radiographic findings raised suspicion of sigmoid colon volvulus due to a coffee bean sign. However, further evaluation through a CT scan raised a possibility of congenital band causing intestinal obstruction at the level of the transverse colon. Surgical intervention confirmed the congenital peritoneal band and successfully released the obstruction, highlighting the significance of multidisciplinary collaboration for prompt diagnosis and management.

KEYWORDS

DiGeorge syndrome, 22q11.2 deletion syndrome, congenital band, intestinal obstruction, radiological investigations, multidisciplinary collaboration.

INTRODUCTION

DiGeorge syndrome, also known as 22q11.2 deletion syndrome, is a congenital disorder resulting from a microdeletion on chromosome 22(1). It is associated with various anomalies such as cardiac defects, immune system dysfunction, vertebral anomalies and craniofacial abnormalities(2). Gastrointestinal manifestations are less commonly reported. Congenital bands, fibrous connections that cause adhesions and obstructive complications, are a relatively rare but important cause of intestinal obstruction. We present a case where a colonic obstruction caused by a congenital band was diagnosed through sequential radiological imaging and managed surgically.

Case Presentation

A 4-year-old child with DiGeorge syndrome presented with abdominal distention and recurrent vomiting for 12 hrs.There is no history of previous abdominal surgeries. Clinical examination revealed a distended abdomen with visible peristaltic waves. Initial plain radiograph demonstrated a grossly distended loop of large bowel, resembling a coffee bean sign indicative of sigmoid colon volvulus(3).However, the presence of haustrations raised suspicion about this initial diagnosis.

Radiological Investigations and Acumen

In this case, the initial radiographic findings were inconclusive, suspicious of sigmoid colon volvulus. However, the presence of haustrations, which is not seen in the sigmoid colon volvulus, prompted further investigation. Given the complex clinical scenario and the child's underlying condition, a CT scan was deemed appropriate. CT imaging allowed for a more detailed assessment of the abdominal structures, revealed a transition point with obstruction at the level of transverse colon (4). The possibility of a congenital band causing intestinal obstruction at this level was raised.

Surgical Intervention, Intraoperative Findings and Management

Prompt collaboration between radiologists, pediatricians, and surgeons played a pivotal role in determining the accurate diagnosis and guiding management. Based on the radiological findings, the patient was urgently taken to the operative theatre. Intraoperative exploration revealed the presence of a congenital band connecting the mesentery to the small bowel, specifically the mid part of the jejunum(5). This band was causing mechanical obstruction of the transverse colon, resulting in gross dilation of the ascending and proximal transverse colon (Figures 5 and 6). The congenital band was carefully dissected and released, relieving the obstruction.

DISCUSSION

DiGeorge syndrome presents with a myriad of clinical manifestations, and gastrointestinal abnormalities should be considered, albeit less common. Congenital bands, often overlooked, can lead to significant obstructive complications and require a high index of suspicion for timely diagnosis. Anomalous congenital band has no relationship with previous abdominal problems, such as laparotomies, inflammatory bowel disease, peritonitis or embryonic remnants, such as the vitelline vessels or omphalomesenteric duct. These bands seem to be congenital in origin(6,7) and cause obstruction when the intestine becomes trapped between the band and the mesentery, which can cause an internal hernia. The aetiology of anomalous congenital band is obscure and has no identifiable embryonic cause, since its location is not similar to that of remnants.

CONCLUSION

In pediatric patients with rare syndromes like DiGeorge syndrome, colonic complications can present unique diagnostic challenges. This case emphasizes the importance of a multidisciplinary approach in diagnosing and managing complex cases .Radiologists, alongside pediatricians and surgeons, should maintain a vigilant approach to guide appropriate investigations and interventions. This case report highlights the successful collaboration between radiology and surgery in diagnosing and managing a colonic obstruction caused by a congenital band in a patient with DiGeorge syndrome.

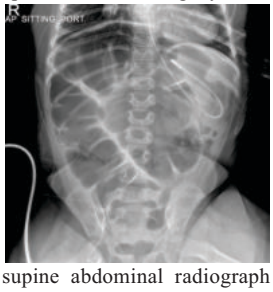
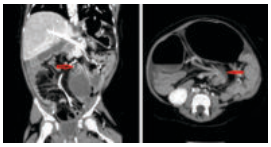


Figure 1: Plain supine abdominal radiograph showing grossly distended large bowel with preserved haustrations.



Figures 2 and 3: Coronal and axial CT scan image showing transition point (marked in red arrow).

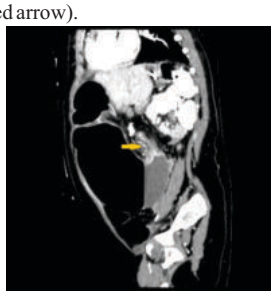
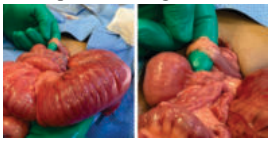


Figure 4: Sagittal CT scan images showing transition point(marked in yellow arrow) and dilated proximal large bowel.



Figures 5 and 6: Intraoperative images showing band causing obstruction and grossly dilated proximal colon.

Acknowledgments
We extend our gratitude to the medical and surgical teams involved in the diagnosis and management of this case.

Conflict of Interest
The authors declare no conflicts of interest related to this case report.

Informed Consent
Informed consent was obtained from the patient's legal guardian for the publication of this case report and accompanying images.

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