Superior Mesenteric Artery Syndrome in Down Syndrome: A Case Report

Fuad Alshaibani

Corresponding author: Fuad Alshaibani, fuad.h.alshaibani@gmail.com

Abstract

Down syndrome is a common chromosomal abnormality, which is associated with a wide spectrum of multiorgan complications, including gastrointestinal disorders. Superior mesenteric artery syndrome is a rare type of small intestinal obstruction caused by compression of the duodenum between the superior mesenteric artery anteriorly and the abdominal aorta posteriorly [1]. It may present acutely with a clinical picture of proximal intestinal obstruction with severe nausea and bilious vomiting. Patients with Down syndrome are more prone to develop superior mesenteric artery syndrome [1]. In general, patients with Down syndrome have a relatively high prevalence of acute and chronic gastrointestinal disorders [2]. Here, I present the case of a young child with Down syndrome who had a seizure secondary to electrolyte disturbances related to recurrent vomiting due to superior mesenteric artery syndrome. Certain conditions have been associated with an increased risk of developing superior mesenteric artery syndrome which includes abdominal surgeries and eating disorders, which can cause significant weight loss that leads to a decrease in the amount of the mesenteric fat pad [3].

Introduction

Superior mesenteric artery syndrome is a rare type of small intestinal obstruction caused by compression of the duodenum between the superior mesenteric artery anteriorly and the abdominal aorta posteriorly [1]. It may present acutely with a clinical picture of proximal intestinal obstruction with severe nausea and bilious vomiting. Patients with Down syndrome were found to be more prone to develop superior mesenteric artery syndrome [1]. In general, patients with Down syndrome have a relatively high prevalence of acute and chronic gastrointestinal disorders [2]. Here, I present the case of a young child with Down syndrome who had a seizure secondary to electrolyte disturbances related to recurrent vomiting due to superior mesenteric artery syndrome. Certain conditions have been associated with an increased risk of developing superior mesenteric artery syndrome which includes abdominal surgeries and eating disorders, which can cause significant weight loss that leads to a decrease in the amount of the mesenteric fat pad [3].

Case Presentation

A 12-year-old boy with Down syndrome was brought to our emergency department with a complaint of jerky movements involving his upper and lower extremities associated with frothy secretions from the mouth and upward rolling of the eyes. This episode lasted for 2 minutes and had a spontaneous resolution. The child appeared tired after the episode and fell asleep. He had no previous similar episodes. The mother reported that the child had been having recurrent episodes of vomiting for the last five days. The vomiting occurred with every meal and was large in amount. She described the vomitus as coffee-ground in appearance. The mother reported that the vomitus did not have bowel movements for one week. The child was seen by a general practitioner one week ago and was diagnosed as having recurrent episodes of vomiting for the last five days. The vomiting occurs with every meal and was large in amount. She described the vomitus as coffee-ground in appearance. The mother reported that the child had been having recurrent episodes of vomiting for the last five days. The vomiting occurred with every meal and was large in amount. She described the vomitus as coffee-ground in appearance. The mother reported that the child had been having recurrent episodes of vomiting for the last five days. The vomiting occurred with every meal and was large in amount. She described the vomitus as coffee-ground in appearance. The mother reported that the child had been having recurrent episodes of vomiting for the last five days. The vomiting occurred with every meal and was large in amount. She described the vomitus as coffee-ground in appearance. The mother reported that the child had been having recurrent episodes of vomiting for the last five days. The vomiting occurred with every meal and was large in amount. She described the vomitus as coffee-ground in appearance. The mother reported that the child had been having recurrent episodes of vomiting for the last five days. The vomiting occurred with every meal and was large in amount. She described the vomitus as coffee-ground in appearance. The mother reported that the child had been having recurrent episodes of vomiting for the last five days. The vomiting occurred with every meal and was large in amount. She described the vomitus as coffee-ground in appearance. The mother reported that the child had been having recurrent episodes of vomiting for the last five days. The vomiting occurred with every meal and was large in amount. She described the vomitus as coffee-ground in appearance.
cells/μL, and a platelet count of 314,000/μL. Electrolyte levels were disturbed, including hyponatremia (127 mEq/L), potassium level of 3.3 mEq/L, low chloride level (70 mEq/L), and elevated bicarbonate level (43 mEq/L). The patient was admitted for further evaluation and management.

The plain abdominal radiograph showed a distended stomach with no abnormal small or large bowel dilatation (Figure 1). Computed tomography of the abdomen was performed to rule out intestinal obstruction. The scan demonstrated extreme dilation of the fluid-filled stomach and the proximal duodenum with a transition point in the aortomesenteric region. The stomach measures 20 cm x 14 x 18 cm on maximum dimensions (anteroposterior x mediolateral x craniocaudal). The distal part of the duodenum showed normal caliber. There was no pneumoperitoneum or ascites. The aortomesenteric angle measures 19 degrees (normal range: 28-65) and the aortomesenteric distance measured 8 mm (normal range: 10-34). A massively dilated stomach exerted a mass effect on adjacent structures and displaced the spleen and small intestinal loops inferiorly. The left renal vein was also compressed between the superior mesenteric artery and the aorta. The aforementioned findings were consistent with intestinal obstruction due to superior mesenteric artery syndrome together with nutcracker syndrome (Figures 2-5).

**FIGURE 1:** Plain abdominal radiograph shows markedly distended stomach with no small or large bowel dilatation.
FIGURE 2: Axial CT scan of the abdomen shows a markedly fluid-filled stomach (arrow) causing a significant mass effect on the left kidney and adjacent bowel loops.

CT: computed tomography
FIGURE 3: Sagittal CT scan of the abdominopelvic cavity shows a markedly distended stomach (asterisk) with a narrow aortomesenteric distance (arrow) consistent with superior mesenteric artery syndrome. CT: computed tomography

The patient had nasogastric tube insertion which drained 3.0 liters of bilious fluid. Aggressive intravenous fluid resuscitation was performed along with the correction of electrolyte disturbances. He received intravenous omeprazole 20 mg and ondansetron 8 mg as a symptomatic treatment for nausea and vomiting. A nasojejunal tube was inserted for enteral feeding. The patient showed a significant clinical improvement after five days, as he gained 0.8 kg. He was able to tolerate oral feeding and the nasojejunal tube was removed. The child protection team was involved in the patient’s management. The team concluded that the child received less care after the parents received genetic confirmation of the diagnosis of Down syndrome, leading to rapid weight loss and the development of superior mesenteric artery syndrome. The growth parameters record indicated that the patient lost 4 kg in two weeks since the diagnosis of Down syndrome.
syndrome was confirmed. The patient was discharged after extensive counseling with the parents together with a social worker. The child was seen in the clinic with regular visits and no active problems were observed.

Discussion

The present case describes a case of proximal intestinal obstruction secondary to superior mesenteric artery syndrome in a patient with Down syndrome. Superior mesenteric artery syndrome is a rare condition in children. Biank et al. [3] conducted a retrospective review of medical charts over 20 years and identified only 22 cases of superior mesenteric artery syndrome in children.

Typically, superior mesenteric artery syndrome develops after a history of acute weight loss before the diagnosis [3]. In the present case, it is assumed that the child was neglected after the parents were confronted with the diagnosis of Down syndrome, which has led to rapid weight loss leading to the development of superior mesenteric artery syndrome. However, in any case, the history of weight loss is not reported. For example, an interesting report by Okamoto et al. [4] describes a case of superior mesenteric artery syndrome in a healthy adolescent with insufficient weight gain relative to height growth. In the pediatric population, superior mesenteric artery syndrome is typically seen in patients after corrective spine surgery for scoliosis [5], as this procedure lengthens the spine cranially and decreases the angle of the aortomesenteric.

A high index of suspicion for superior mesenteric artery syndrome is required given the non-specific clinical presentation. Imaging criteria for superior mesenteric artery syndrome involve duodenal obstruction with an aortomesenteric angle of less than 25 degrees and a high fixation of the duodenum by the ligament of Trietz [6]. The management of superior mesenteric artery syndrome is usually conservative. The goal of treatment is to decompress the bowel and correct any fluid and electrolyte disturbances. Surgical treatment is attempted when conservative therapy fails to achieve any improvement [6].

Conclusions

Superior mesenteric artery syndrome is a rare cause of proximal intestinal obstruction in children. Weight loss is an important predisposing factor to this condition. Therefore, clinicians must maintain a high index of suspicion of superior mesenteric artery syndrome in any child who presents a clinical picture of intestinal obstruction after rapid weight loss for any reason. Conservative treatment with aggressive fluid resuscitation and correction of electrolyte disturbances is generally successful.

Additional Information

Disclosures

Human subjects: Consent was obtained or waived by all participants in this study. Governmental Hospital Ethics Committee issued approval N/A. Case reports are waived by the institutional review board. Informed consent was taken from the parents for the publication. Conflicts of interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following: Payment/services info: All authors have declared that no financial support was received from any organization for the submitted work. Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. Other relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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