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LETTER TO THE EDITOR

Superior Mesenteric Artery Syndrome in a Patient with Autism Spectrum Disorder: Case Report and Review of the Literature

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Dear Editor,

Superior mesenteric artery (SMA) syndrome is a gastrovascular disorder resulting from compression of the distal duodenum by the abdominal aorta and the overlying SMA. Its incidence is 0.013–0.3 %. There is a female predominance with a typical age of presentation between 10 and 30 years of age; however, onset in infancy has been reported (Mandarry et al. 2010). Symptoms may be acute or chronic and typically include epigastric pain, eructation, voluminous vomiting, and early satiety. SMA syndrome typically results from rapid and significant weight loss due to anorexia nervosa, hypercatabolic states, major surgery, malignancy, or malabsorption. It is associated with considerable morbidity and mortality.

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The SMA usually forms an angle of 45–60° with the abdominal aorta. The third portion of the duodenum crosses caudal to the origin of the SMA, coursing between it and the aorta. Fat and lymphatic tissues surround the SMA, protecting it from compression and maintaining the normal aortomesenteric angle and distance. With significant weight loss, these support tissues diminish, resulting in external duodenal compression. Predisposing conditions include short or abnormally inserted ligament of Treitz, low congenital origin of the SMA, neurological impairment, increased spinal lordosis, rapid linear growth, immobilization in a body cast, and peritoneal adhesions (Mandarry et al. 2010). Body mass index (BMI) below the fifth percentile is the best predictor of SMA syndrome.

On physical exam, patients may have a distended and tender epigastrium, with normal to hyperactive bowel sounds. Symptoms may improve in the left lateral decubitus or knee-to-chest position, or with pressure below the umbilicus in a superior-posterior direction, and worsen when supine (Marecek et al. 2010). Unrecognized or inadequately treated cases may progress to severe malnutrition, dehydration, electrolyte abnormalities, intestinal perforation, and death (Biank and Werlin 2006; Mandarry et al. 2010).

Diagnostic evaluation begins with an abdominal radiograph, which may show gastroduodenal dilatation, air-fluid levels, and occasionally, an abrupt vertical cutoff of air in the third portion of the duodenum. Upper gastrointestinal series may show delayed gastric emptying and distal duodenal obstruction. Computed tomography with contrast, magnetic resonance angiography, and endoscopic ultrasound can demonstrate vascular compression of the duodenum (Mandarry et al. 2010).

Treatment begins with conservative measures including nasogastric decompression, correction of electrolyte

imbalance, and nutritional optimization. The latter involves tube feeding or hyperalimentation, with gradual reintroduction of small, frequent oral feedings. Restoration of weight is the ultimate goal. When conservative management fails and the predominant symptom is vomiting, surgery is recommended.

Methods

Our patient is an 11-year-old former 36-week preterm male who has pervasive developmental disorder, not otherwise specified (PDD-NOS), mixed receptive/expressive language disorder, epilepsy, attention-deficit/hyperactivity disorder (ADHD) symptoms, and chronic constipation. His full-scale intelligence quotient (IQ) improved from 71 at age 6 years to 99 at present. During that same interval, his communication skills remained significantly impaired with standard scores of Receptive Language 58 and Language Expression 63. His responses to verbal prompts are slow and inconsistent. Over the previous year, his family relocated and his father deployed overseas for the military. There were no recent changes to his medications: divalproex sodium, imipramine, olanzapine, and amphetamine/dextroamphetamine. During the previous 4 weeks, he became more socially withdrawn and physically aggressive. These symptoms rapidly escalated, necessitating inpatient psychiatric care for major depressive disorder, single episode, with catatonic features and refusal to eat. After 2 weeks, his neurovegetative symptoms improved with minor adjustment to his psychotropic medications. However, his appetite remained poor and he vomited once. He transferred to our pediatric ward for medical evaluation. Further questioning revealed a 2-month history of episodic dizziness accompanied by nonspecific headaches that improved with acetaminophen. He denied associated night wakings, auras, nausea, photophobia, or phonophobia. He complained of easy fatigability and occasional leg pain. He denied fever, chills, or abdominal pain.

Results

Physical examination revealed a comfortable, cachectic male with no focal findings. Review of systems revealed a 2.73-kg weight loss over the preceding 12 months, with an increase in height of 3.05 cm. His body mass index (BMI) decreased from 14.8 kg/m^2 (11th percentile) to 12.8 kg/m^2 (<1st percentile). Laboratory studies included normal complete blood count, prealbumin, and C-reactive protein. Phosphate and alkaline phosphatase were slightly low, but renal function was otherwise unremarkable. We initially suspected a behavioral feeding disorder and attributed his weight loss to inadequate caloric intake. Thus, we planned for intensive feeding therapy and recommended placement of a gastrostomy tube to maximize his caloric intake. In preparation for the latter, we ordered an upper gastrointestinal series, which showed gastric distension with a normal contour and normal gastric emptying. The bulb, descending, and proximal aspect of the third portion of the duodenum had moderate distension. There was narrowing in the mid-aspect of the third portion of the duodenum, where it crossed between the abdominal aorta and SMA. The aorta-SMA angle and the aorta-to-SMA distance were abnormally decreased. We diagnosed SMA syndrome and placed a percutaneous endoscopic gastrostomy tube. Once he tolerated tube feedings we discharged him after 3 days on the pediatric ward. Over the subsequent weeks, his nutritional status improved, his physical aggression resolved, and he became more interactive than in the previous 2 years. At age 13 years, his BMI is 16.4 kg/m^2 (11th percentile). He tolerates more foods orally, but still requires supplemental tube feedings.

Discussion

Our patient presented atypically with 2 years of anorexia and minimal weight loss, complicated by autism spectrum disorder (ASD), ADHD symptoms, and inpatient psychiatric hospitalization for major depressive disorder, single episode, with catatonic features. Although SMA syndrome often presents acutely with abdominal pain, vomiting, and abdominal tenderness, some patients present with chronic vague abdominal symptoms, early satiety, anorexia, or waxing and waning episodes of abdominal pain with vomiting (Biank and Werlin 2006; Mandarry et al. 2010). In Biank et al.'s series, patients who had cerebral palsy or traumatic brain injury lost only 2 kg, whereas neurotypical patients lost 7.4 kg (Biank and Werlin 2006). SMA syndrome requires a high degree of clinical suspicion supported by radiographic evidence of obstruction. Due to its non-specific symptoms, clinicians often investigate other causes, leading to delay in diagnosis (Mandarry et al. 2010).

Autism spectrum disorder is a complex neurodevelopmental disorder featuring repetitive behaviors and deficits in social interaction and communication. As many as 80 % of children who have ASD reportedly have sensory processing problems, including altered pain sensorium (Bromley et al. 2004). These are not included in the diagnostic criteria for autistic disorder in the *Diagnostic and Statistical Manual of Mental Disorders, 4th Edition, Text Revision* (*DSM-IV-TR*; American Psychiatric Association 2000). However, some autism rating scales include altered pain sensorium, and *DSM-5* proposes to include

"apparent indifference to pain" in its behavioral criteria for ASD (www.dsm5.org). Descriptions in the literature include increased pain sensitivity, not feeling pain as intensely as others do, and having an indifference to pain. However, these rely on anecdotal reports, clinical impressions, personal recall, subjective assessment, and questionable measures of pain. A small, controlled study comparing pain sensation in children who have ASD to matched neurotypicals found that despite no difference in objective (i.e., facial) expression of pain, parents whose children had ASD underestimated their child's pain (Nader et al. 2004). A second study by these authors found that non-parental observers correctly identified pain based on observation of facial expression in children who have ASD (Messmer et al. 2008). Because both studies examined pain intensity based on an acute pain reaction to a specific medical procedure (venipuncture), findings may not generalize to observations of everyday or chronic pain in children who have ASD. Regardless, caregivers may discount or deny signals of distress in children who cannot clearly express their pain, especially if they believe that the child does not feel pain. In their 2009 study comparing 73 children who had ASD to 115 neurotypical children, Tordjman and colleagues found no difference in behavioral pain reactivity in response to venipuncture (Tordjman et al. 2009). In fact, the change in heart rate and the plasma beta-endorphin levels were higher in the ASD group. They proposed that the apparent decreased pain sensitivity attributed to individuals who have ASD derive not from analgesia but a different mode of pain expression, related to deficits in communication and cognition.

Beliefs about pain insensitivity in children who have ASD put them at risk for substandard pain management. In their 2010 review of the literature, Dubois and colleagues concluded that more knowledge about pain in persons who have ASD should enable the development of specific assessment tools and improved pain management in daily care (Dubois et al. 2010). Because we considered our patient's symptoms behavioral or psychological in nature, we initially attributed them to a behavioral feeding disorder and ASD. His recent inpatient psychiatric hospitalization for major depressive disorder, single episode, with catatonic features reinforced that assumption. His poor language skills limited his ability to communicate effectively, altering our interpretation of his symptoms.

Marecek et al. previously reported SMA syndrome in a patient who had ASD; however, their patient had recurrent bouts of SMA syndrome that resolved with correction of his complex spinal deformity (Marecek et al. 2010). To our knowledge, ours is the first report of SMA in a patient with ASD and no contributing medical problems.

Our findings emphasize the importance of considering discomfort, pain, and illness in patients who have developmental disabilities presenting with maladaptive behaviors, especially if they differ from the patient's usual pattern. In particular, providers should consider the possibility of SMA syndrome in patients experiencing unexplained weight loss, early satiety, and food refusal, even in the absence of other symptoms suggesting duodenal obstruction. Earlier recognition may allow for conservative management with improved outcome.

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