CLINICAL CASE

Unusual neonatal case of superior mesenteric artery syndrome with Meckel's diverticulum and literature review

Caso inusual neonatal de síndrome de la arteria mesentérica superior con divertículo de Meckel y revisión de la literatura

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Abstract

Superior mesenteric artery syndrome (SMAS) is a rare cause of duodenal obstruction which is characterized by compression of the duodenum due to narrowing of the space between the superior mesenteric artery and aorta. Incomplete duodenal obstruction due to SMAS in neonates is rarely reported in the literature. In this case, it is a full-term 2-day-old male with the complaint of recurrent vomiting starting soon after birth. The patient was diagnosed with SMAS and duodenoduodenostomy was performed. Accompanying Meckel's diverticulum was excised.

Keywords: *Duodenal obstruction. Newborn. Superior mesenteric artery syndrome. Vomiting.*

Resumen

El síndrome de la arteria mesentérica superior (SMAS) es una causa rara de obstrucción duodenal que se caracteriza por la compresión del duodeno debido al estrechamiento del espacio entre la arteria mesentérica superior y la aorta. La obstrucción duodenal incompleta por SMAS en recién nacidos rara vez se informa en la literatura. En este caso se trata de un varón de 2 días nacido a término que presenta vómitos recurrentes desde poco después del nacimiento. El paciente fue diagnosticado de SMAS y se le realizó duodenoduodenostomía. Se extirpó el divertículo de Meckel que lo acompañaba.

Palabras clave: *Obstrucción duodenal. Recién nacido. Síndrome de la arteria mesentérica superior. Vómitos.*

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Introduction

Superior mesenteric artery syndrome (SMAS), also known as aortomesenteric duodenal compression, cast syndrome, chronic duodenal ileus, or Wilkie syndrome, was first described in 1861 based on an autopsy case by Von Rokitansky[1]. David Wilkie published the largest series of 75 patients in 1927, made a detailed clinical and pathophysiological description, and recommended treatment approaches; hence, it is also called Wilkie Syndrome[2]. Cast syndrome was used by Dorph in 1950[3]. Finally, it was named as SMAS by Kaiser et al. in 1960[4].

SMAS is one of the rare causes of upper gastrointestinal tract obstruction. It is characterized by the narrowing of the space between the superior mesenteric artery (SMA) and the abdominal aorta causing compression of the third segment of the duodenum. Although the exact prevalence of the disease is unknown, its incidence is estimated to be between 0.1% and 0.3%. The SMAS occurs in adolescents and mostly young adults between the ages of 10 and 39 years, but can ultimately occur at any age. It is observed 3:2 more frequently in females than in males. An ethnic predisposition has not been described, but familial cases have been reported[5,6]. Patients usually present after orthopedic surgical procedures or acute weight loss due to hyperthyroidism, anorexia nervosa, or gastroenteritis. Symptoms typically consist of chronic intermittent abdominal pain, vomiting (sometimes bilious), nausea, early satiety, and anorexia[7]. In neonates and infants, SMAS is extremely rare and presents as a rare cause of feeding intolerance or incomplete duodenal obstruction. The diagnosis of SMAS is usually confirmed by upper gastrointestinal radiography, but can also be diagnosed by computed tomography or diagnostic laparotomy/laparoscopy.

Ethical approval

Ethical consent was obtained from the patient's family for the publication of the medical case and accompanying images. The ethical consent form was archived by the first author of this article.

Case presentation

A 3065-g male baby was born at 38 + 3 gestational weeks from a healthy 31-year-old mother with gravida 2 and 1 abortion. He was admitted to our hospital on the 2nd postnatal day due to recurrent bilious vomiting soon after delivery. There were no abnormalities on his prenatal follow-up. On physical examination, the abdomen was mildly distended, there was no tenderness, and no mass was noted. Bowel sounds were normoactive. Other systemic examinations were normal. The patient was followed up with an orogastric tube and had 13 cc/h of bilious fluid. All laboratory tests were normal. Antibiotic treatment was started empirically in terms of neonatal sepsis. The abdominal ultrasound was normal. The patient's complaints persisted and therefore an upper gastrointestinal series with contrast was performed. A stenosis in the third part of the duodenum and marked dilatations in the second and first parts of the duodenum were observed (Fig. 1). The late babygram revealed passage of the contrast material to the colon (Fig. 2).

Diagnostic laparotomy was performed since the patient did not improve clinically. The patient was examined for duodenal stricture, and external compression, especially the duodenal web and annular pancreas. It was found that the duodenum was trapped under SMA and this segment was stenotic (Fig. 3). In addition, Meckel's diverticulum was detected incidentally and excised (Fig. 4). The ligament of Treitz was examined and the position was found normal. After meticulous dissection of the duodenum, the stenotic segment was excised and duodenoduodenostomy was performed. A penrose drain was left in place.

Following the operation, the patient was admitted to the neonatal intensive care unit, and total parenteral nutrition and antibiotherapy were redefined. Defecation occurred on the post-operative 2nd day. Minimal enteral feeding was initiated on the 5th post-operative day orally. The patient gained weight and was discharged on the post-operative 14th day.

Discussion

Intestinal obstructions either partial or complete occur approximately one in 1500 live births. Among the rarest cases, SMAS is characterized by the obstruction of the duodenum beneath the SMA causing gastroduodenal dilatation[8]. Predisposing factors leading to a reduction in the angle between the SMA and the aorta resulting in SMAS include diseases associated with significant weight loss with loss of peritoneal and mesenteric adipose tissue responsible for the cushion effect, acute or prolonged trauma, spinal disease or deformity, peritoneal adhesions due to inflammation or thickening of the mesenteric root[7].

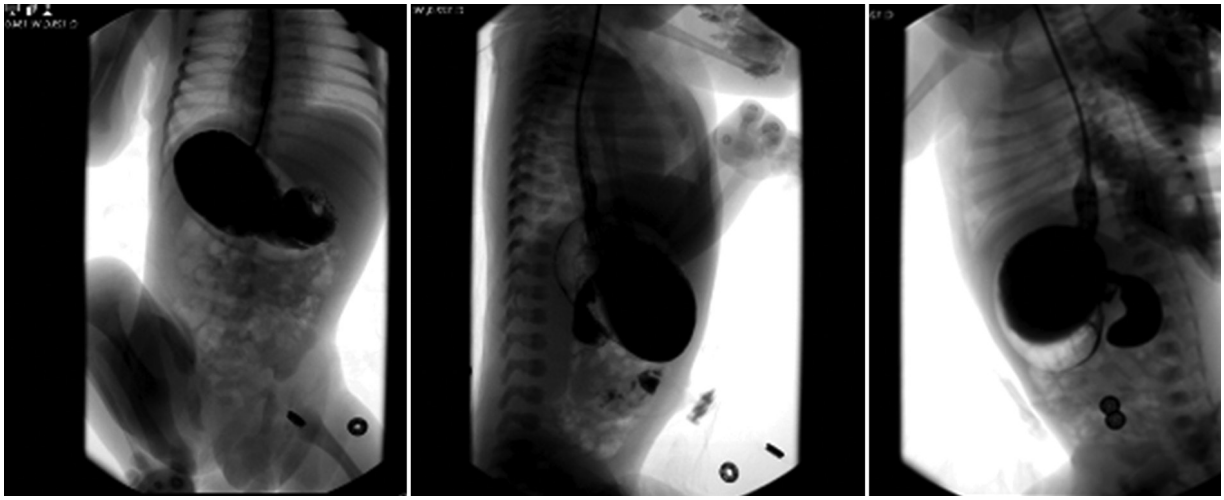

Figure 1. *Upper gastrointestinal system series, obstruction in the 3rd portion and dilatation in the 1st and 2nd portions of the duodenum.*

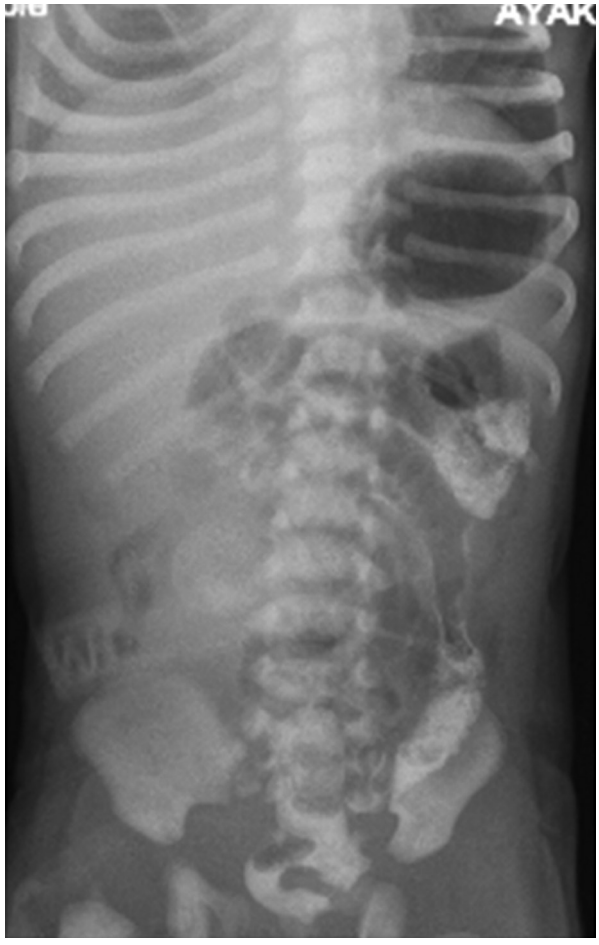

Figure 2. *The babygram performed 6 hours after the administration of contrast agent. Passage of the contrast material beyond the 3rd portion of the duodenum.*

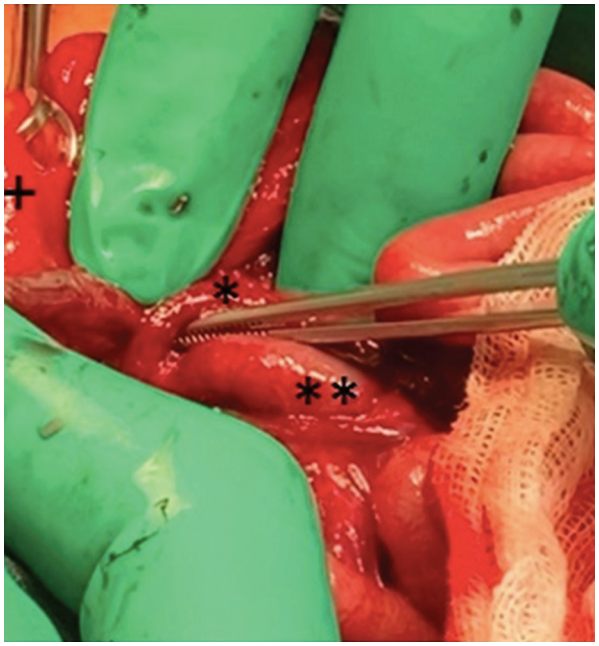

Figure 3. *During the operation, it was observed that the superior mesenteric artery passed over the third portion of the duodenum and caused duodenal obstruction (+: proksimal part of duodenum, *: superior mesenteric artery, **: third portion of the duodenum).*

There are only two familial case series in the literature. The first case included a mother and her daughter; the second included a father and his four daughters[9].

SMAS should be distinguished from the SMA-like syndrome, in which the pressure exerted on the duodenum from SMA is secondary to the duodenal dilatation. The variant with normal aortomesenteric angle and reduced aortomesenteric distance may be associated with diminished mesenteric venous drainage[10].

Table 1. The reported data of superior mesenteric artery syndrome in neonates

No	Author/year	Age at diagnosis	Sex	Symptom	Surgery
1	Caspi et al./2003	0 day (prenatal)	Female	Polyhydramnios is, bilious vomiting	Divided Treitz, stricturoplasty for duodenal stenosis
2	Sözübir et al./2006	1 day	Female	Bilious vomiting	Duodenojejunostomy
3	Mosalli et al./2011	7 days	Male	Abdominal distension, feeding intolerance, deterioration after diarrhea	Divided Treitz
4	Our case	2 days	Male	Feeding intolerance, bilious vomiting	Duodenoduodenostomy

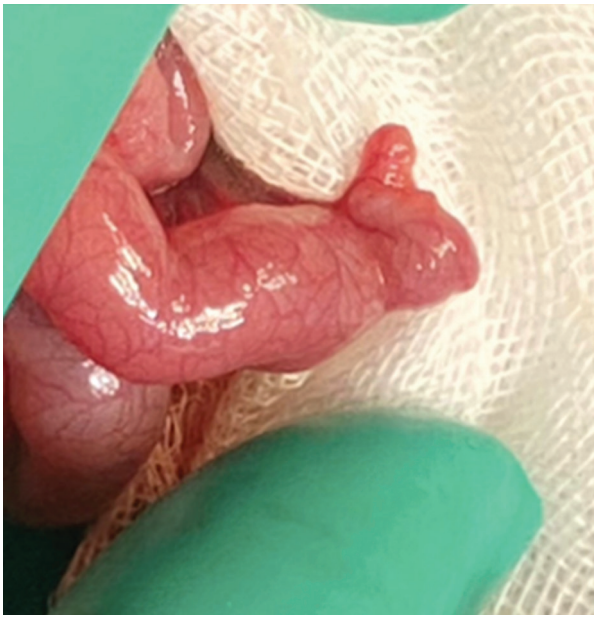

Figure 4. *Meckel's Diverticulum.*

SMAS has been described mainly in adults and rarely in children, but neonatal SMAS is extremely rare. Few cases in infancy and only four neonatal cases including this case have been documented in the literature (Table 1). All these neonatal cases were presented with bilious vomiting and incomplete bowel obstruction soon after birth[7,11,12]. The third case is a newborn SMAS which was provoked after an attack of diarrhea[12]. Clinical signs include gastric enlargement, nausea, and vomiting (may contain bile), aggravated by feeding. The main symptoms in the newborn are vomiting and poor weight gain.

SMAS can be difficult to diagnose and is usually diagnosed by exclusion or laparotomy. Radiological features suggestive of SMAS are enlargements of the proximal part of the duodenum and stomach on plain abdominal radiography. On upper gastrointestinal contrast series, obstruction in the third part of the duodenum, significant delay of gastroduodenal passage by 4-6 h, retrograde movement of contrast agent may be observed. Postural change, left lateral, or prone positioning may resolve the obstruction. In cases with complete obstruction, polyhydramnios, and double bubble signs like duodenal atresia can be detected on prenatal ultrasonography[9].

Contrast-enhanced abdominal computed tomography determines the anatomic location of the obstruction site and the angle formed by the SMA and the aorta. Magnetic resonance angiography is also useful for measuring the aortomesenteric angle. Fiberoptic endoscopy is effective in distinguishing intraluminal causes of occlusion if it can be passed through the obstruction[7].

There are two main goals of surgical treatment; either bypassing the obstruction site or duodenal release from the compression. It is known that persistent pain and blind loop syndrome may be observed in duodenojejunal bypasses while iatrogenic malrotation and entrapment may complicate the lysis of the ligament of Treitz[13]. Unfortunately, considerably high failure rates of these procedures provoked us to think of a way to preserve the duodenal function and anatomy while preventing blind loop and re-entrapment. In this case, we preferred to perform end-to-end duodeno-duodenal anastomosis to prevent any long-term complications. The meticulous freeing of the duodenum and the anastomosis went well and both the duodenum and SMA appear fine after 1 year of post-operative evaluation. The patient is thriving.

Conclusion

Although extremely rare in neonates, SMAS should be considered in cases presenting with obstructive

upper gastrointestinal symptoms. Delayed diagnosis will result in prolonged hospital stay and delay in feeding. While duodenoduodenostomy appears as a promising technique, more data is needed further.

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Conflicts of interest

The authors have no conflicts of interest to declare.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The first author (not corresponding author) of article is in possession of this document.

Use of artificial intelligence for generating text. The authors declare that they have not used any type of generative artificial intelligence for the writing of this manuscript, nor for the creation of images, graphics, tables, or their corresponding captions.

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