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Original article

Superior mesenteric artery syndrome in a child with acute gastric dilatation with refractory Guillain–Barré syndrome

Ehsan Ghaedi [a, b], Hamidreza Kianifar [c], Behnoush Imani [d], Bahareh Imani [e, *]

[a] Department of Cellular and Molecular Nutrition, School of Nutritional Sciences and Dietetics, Tehran University of Medical Sciences, Tehran, Iran
[b] Students' Scientific Research Center (SSRC), Tehran University of Medical Sciences (TUMS), Tehran, Iran
[c] Department of Pediatric Gastroenterology, Mashhad University of Medical Sciences, Mashhad, Iran
[d] Department of Radiology, Shahid Beheshti University of Medical Sciences, Tehran, Iran
[e] Department of Pediatrics, Mashhad University of Medical Sciences, Mashhad, Iran

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SUMMARY

Superior Mesenteric Artery Syndrome (SMAS) resulting from proximal partial small bowel obstruction is one of possible causes of acute gastric dilatation (AGD). A child with refractory demyelinating Guillain–Barré syndrome which received 5 times IVIG and consequently 5 times plasmapheresis intubated until 59th day of admission. Because of complicated treatment and cardiopulmonary resuscitation (CPR) nutrition was completely neglected which lead to severe SMAS because of weight loss. Gradual advancements of continuous enteral and parenteral nutrition improved patient's symptoms significantly. Hypophosphatemia complicated the weaning from ventilator which after nutrition therapy resolved and patient extubated. Present case is the first report of pediatric demyelinating GBS suspected to SMAS.

1. Introduction

Acute gastric dilatation (AGD) is a rare medical condition in which the stomach becomes gradually hypotonic and overstretched [1]. Superior mesenteric artery syndrome (SMAS) also known as Willkie's syndrome is a rare gastro-vascular disorder resulting from proximal partial small bowel obstruction can cause AGD. Treatment is initially conservative therapy, which includes nutrition and electrolyte therapy and specific position [2]. Guillain–Barré syndrome (GBS) is the most frequent cause of acute flaccid paralysis in children [3].

Here we present a patient who admitted for demyelinating GBS whose hospital course was complicated due to poor nutritional management. His condition managed with nutritional intervention.

2. Case presentation

A nine years old boy (131 cm, 30 kg, and BMI was 18.3 kg/m^2) with profound weakness in lower and upper extremities presented to the emergency department. Severe progressive pain in extremities commences and escalates gradually. The patient had a history of upper respiratory infection for 6 days before admission. The patient was never hospitalized before this time and there was no history of gastrointestinal symptoms and surgery.

Upon arrival to the hospital, the patient's vital signs were stable, with a 36 °C temperature, blood pressure of 100/70 mm of Hg, and a pulse rate of 88 beats per minute and respiratory rate 22 and O_2 Saturation 99%. Laboratory tests were just abnormal for Hemoglobin and mild Iron deficiency anemia. Patient appeared well-nourished. He was awake and alert but unable to obey commands due to sever muscle weakness. Electromyography (EMG)/Nerve Conduction Study (NCV) was performed for patient and compatible with polyradiculoneuropathy. Neurological findings revealed bilateral flaccid paralysis and hyperreflexia affecting all extremities. Muscle force of upper extremities was 3/5 and also Deep Tendon Reflexes (DTR) decreased and lower extremities were 0/5 without DTR. Cranial neurological examination and Gag reflex was also normal and there were no signs of ataxia or nystagmus. He had decreased light touch, temperature, and pinprick sensation bilaterally from his feet to his thighs, and in his hands ascending to his shoulders the patient developed by hypo nasal speech and poor gag reflex in 2nd day of admission. Demyelinating GBS was suspected. The patient's treatment with IVIG (Intravascular Immunoglobulin)

* Corresponding author. Imam Reza Hospital - Department of Pediatrics; Mashhad University of Medical Sciences, Mashhad, Iran.
E-mail address: imanibh@mums.ac.ir (B. Imani).

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(1gr/kg/day*2) was started. Also the patient developed by respiratory insufficiency and cardio respiratory arrest leading intubation in 2nd day of admission. Successful cardiopulmonary resuscitation (CPR) was done at the same time.

The patient was refractory (with 5 times IVIG and methylprednisolone). In 14th day of admission patient received plasmapheresis (five PE were performed). A slow improvement of respiratory function and peripheral force were observed after the first PE. This resulted improvement in his proximal upper and lower extremity motor strength. Then, left upper extremity sensory and motor nerve conduction studies (NCS) were performed because of persistent generalized areflexia. Mechanical ventilation was weaned on the 59th day and the same time he started to move his shoulder.

In hospital course the patient developed recurrent feeding intolerance and bilious vomiting, sever nosocomial sepsis (Sepsis ESR = 60, WBC = 38400, NEUT = 84%, LYM = 2%) repeated convulsion, autonomic involvement, failure to weaning from mechanical ventilation, weight loss (10 kg) during hospitalization, nutritional disturbance (hypomagnesaemia and hypophosphatemia).

During intubation in critical care unit, severe abdominal distention and bilious vomiting along with upper GI bleeding on 53rd day of admission occurred. Enormous secretion from Naso-Gastric Tube and biliary secretion was reported. Abdominal X-ray image showed Gastric Outlet Obstruction (GOO) and AGD. Based on the patient's clinical course, we suspected a diagnosis of SMAS due to sever weight loss and bilious vomiting, but because of clinically ill and intubated patient, we were not able for doing abdominal CT scan or upper GI series. Thus a nasogastric tube was placed, draining approximately 200 mL of stomach contents. The surgeons and gastroenterologists were consulted and conservative management for SMAS started via enteral and parenteral nutrition (Fig. 1).

2.1. Medical nutrition therapy

During hospitalization nutritional status was not checked and patient did not receive enteral nutrition (EN). It must be mentioned that EN indicated many times but due to gastrointestinal bleeding and food intolerance EN discontinued. Furthermore, parenteral nutrition neglected until we commence PPN at 53rd day of admission. So in fact patient were *nil per os* (NPO) for 53 days. Based on following formula resting energy expenditure (REE) calculated. The final goal estimated approximately 90 kcal/kg/day but at first started with 30 kcal/kg/day and gradually increased. And also 2–2.5 g/kg protein were added (Table 1).

Gradual advancements of continuous enteral feeding by infusion pump were introduced to prevent refeeding syndrome. Other nutritional therapy includes treatment of hypomagnesaemia and hypophosphatemia and PN. Hypophosphatemia complicated the weaning from ventilator which after nutrition therapy resolved and patient extubated. Thereafter, the patient's symptoms significantly improved and after 20 days' gastric dilatation relieved and pt gain 80 g' weight. Patient extubated on day 59 (Table 2).

2.2. Post discharge follow up

At discharge to rehabilitation after 78 days of hospitalization, the patient's force had returned to slightly normal in the arms and power (4/5) in the distal legs and he was able to walk with assistance. Prior to discharge, the patient was broadly counseled on diet modification and oral supplementation with pediatric sip feed (NutriniDrink). He was eventually discharged with the ability to tolerate oral feeding and weighed 25 kg (BMI: 14.793 kg/cm2). The patient gained approximately 3 kg over a period of 3 months follow up, his symptoms resolved and was able to maintain his weight. The ethical committee of MUMS approved the study protocol and patient provided written informed consent.

3. Discussion

The critically ill patients are in the risk of negative energy balance (because of hypermetabolic state and increment in the nutritional requirements). This condition leads to catabolism of lean body mass and worsening of the clinical outcome and recovery. There are many barriers to adequate nutritional supply in the Pediatric Intensive Care Unit (PICU) such as gastrointestinal disorders, pause for procedures and mechanical ventilation. Moreover, individual assessment of a patient's nutritional status is not part of

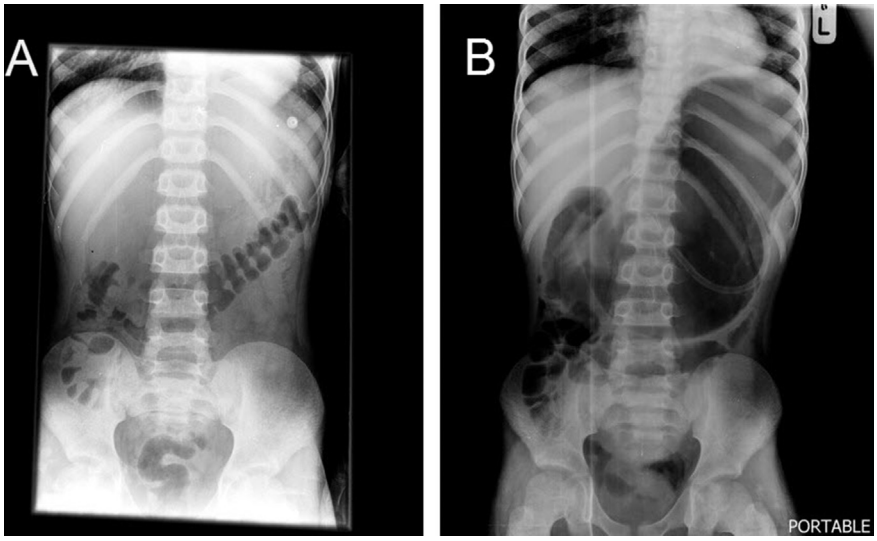

Fig. 1. A: Transverse and descending colon are seen with almost normal diameter while other portions of abdomen appear gasless (fluid filled small bowels?). B: In serial abdominal X rays performed in supine position, progressive gastric dilation is noted. Evidence of dilation of first and 2nd part of duodenum and proximal jejunum is also seen. The pelvic cavity is not gasless. Soft tissue and bones are unremarkable.

Table 1
Nutrition intervention after superior mesenteric artery syndrome diagnosis.

Nutrition Management	EN	PN
Intervention	Was Started at 20 cc every 2 h and increased gradually to about 250 cc every 3 h, Continuous feeding via infusion pump was used if there was feeding intolerance. Formula: (MCT Oil-calorie dense formula (Nutrinidrink powder and solution) Glutamine: 0.5 mg/kg/day	**Carbohydrate:** Was Started at 5 g/kg/day and increased to 20 g/kg/day. **Amino acid** 10%: Was Started at 0.5 g/kg/day and increased to 1.5 g/kg/day. **Fat (SOMFLipid, Fresenius Kabi):** Was Started at 0.5 g/kg/day and increased to 2 g/kg/day. **Selenium:** 20 μg/kg/day for first day and then followed by 10 μg/kg/day daily for 10 days. **Zinc:** 8 mg/day IV Trace element and Multi Vitamins (Soluvit, Fresenius Kabi)

Table 2
Nutrition Assessment at admission, before nutrition therapy and after nutrition therapy.

Nutrition assessment	Before	During Intubation in ICU	At discharge after Initiation of Nutrition therapy
Weight kg	31	21	25
Height cm	130	130	130
BMI kg/cm2	18.3	12.42	14.793
Triglyceride (Mg/dl)	74	122	82
Glucose (Mg/dl)	95	110	103
Magnesium (Mg/dl)	1.2	2.3	2.5
Phosphorus (Mg/dl)	2.2	3.8	4.5
Vit D ng/ml	29	35	35
Creatinine	0.3	0.6	0.5
Liver enzyme IU/dl	NL	NL	NL
Stool	NL	NL	NL
UA/UC	NL	NL	NL

the usual procedures upon admission to PICU and also most of them have not been validated for critically ill children [4]. Currently anthropometry is the best tool in PICU for nutrition assessment [5].

The etiology of SMA syndrome in this patient is most probable associated to the acute and dramatic nature of his weight loss. Others reported same conditions in ICU setting which lead to SMAS because of postoperative ICU stay [6] and cachexia [7,8].

Nutrition risk factors in GBS patient including poor intake due to bulbar palsy, poor gag reflex and poor oral intake, poor digestion and absorption and hypermetabolic state. It has been recommended that GBS patients receive minimally 1.35 to 1.58 X the calculated REE (Resting Energy Expenditure) superimposed on a metabolic state plus an additional 30%–50% for weight stabilization [9]. Therefore, it is probable if our nutrition team involved earlier, length of ICU stays and duration of ventilator-dependant state reduced. Interestingly nutrition assessment was not done at all. Staffs without weighing patient in treatment course, (before entrance of nutrition experts), mentioned just one weight which record at admission, repeatedly. More important serum albumin will be an unreliable gauge of nutritional status because patient undergoes PE with albumin replacement.

AGD is faced most often as a postoperative complication such as ileus and plenty of other disorders. Although its incidence is quite rare, it can have overwhelming results [10]. Acute angulations of the SMA caused by paucity of mesenteric and retroperitoneal fat pads leads to compression of the third part of the duodenum between the overlaying SMA and underlying abdominal aorta because of rapid weight loss (see Fig. 2). SMAS commonly presents chronic in nature [11] due to severe weight loss, such as eating disorders, cardiac cachexia, bariatric surgery and prolonged bed rest have been reported. This is the first

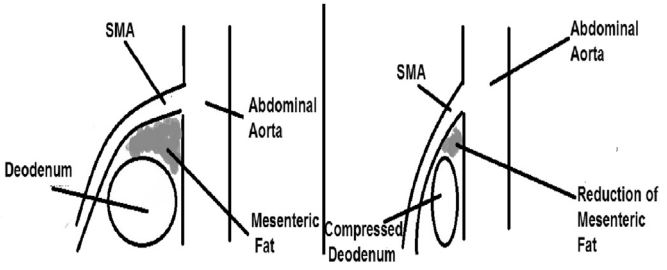

Fig. 2. Wilkie's syndrome. Schematic image of aortomesenteric angle reduction by attenuating the fat pad between vascular structures.

reports of SMAS in patient receiving treatment for GBS after ICU stay. Conservative treatment is the first step in managing these patients to reconstitute the fat pad that normally surrounds the mesenteric bundle [2]. Here we showed that the patient responds to nutrition therapy and symptoms relieve after weight gain.

4. Conclusion

Our search of the English medical literature revealed no reports of SMAS in patients receiving treatment for GBS. In addition, SMAS is found more commonly in females than males and previous reports show that SMAS commonly presents chronic in nature [11]. Therefore, this is very rare condition and to the best of our knowledge the first case of SMAS due to poor nutritional management in PICU during GBS management. This case highlights the

importance of adequate nutrition during critical care admission; including nutritional screening and intervention by a multidisciplinary team that includes dietitian.

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Conflicts of interest

None.

Ethical considerations

Ethical issues (including plagiarism, misconduct, data fabrication, falsification, double publication or submission, redundancy) have been completely observed by the authors.

Author contribution

E.G collected the data and write the primary manuscript, B.I report the graphs and add comments, H.K. edited manuscript and B.I. approve the manuscript with last edition.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.clnesp.2019.05.007.

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