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Superior Mesenteric Artery Syndrome in a Patient with Duchenne Muscular Dystrophy: A Case Report

Yu Hui Won[1,2*], Da-Sol Kim[1,2], Gi-Wook Kim[1,2], Sung-Hee Park[1,2], Myoung-Hwan Ko[1,2], Jeong-Hwan Seo[1,2]

[1]Department of Physical Medicine & Rehabilitation, Jeonbuk National University Medical School, Jeonju, Korea
[2]Research Institute of Clinical Medicine, Biomedical Research Institute of Jeonbuk National University Hospital
Yu Hui Won; wonyh@jbnu.ac.kr

Introduction

Superior Mesenteric Artery (SMA) Syndrome is a rare condition in which the third duodenum is compressed between the superior mesenteric artery and the aorta, with rapid weight loss being a major trigger. Patients with Duchenne Muscular Dystrophy (DMD) are at increased risk due to muscle atrophy and malnutrition, especially after spinal deformity correction surgery, which can alter body shape and lead to weight loss. Here, we report a case of recurrent SMA Syndrome in a DMD patient and its surgical management.

Case presentation

A boy born in 2004 was diagnosed with DMD at age three through genetic testing. He began using non-invasive ventilation (NIV) in 2019 (age 15) and underwent scoliosis correction surgery in December 2021 (age 17). Soon after, he developed persistent vomiting and was diagnosed with SMA Syndrome in January 2022. Due to severe respiratory impairment (vital capacity 0.26 L, 7%), surgery was considered high-risk and was deferred in favor of conservative management with nutritional support.

Because the patient was using a nasal mask for NIV, a standard L-tube could not be used. Instead, a thin 12 French enteral tube was inserted into the jejunum through nose by an interventional radiologist, and taping was applied to minimize air leakage at the nasal mask. His weight increased from 31.35 kg to 34.1 kg, but oral intake remained insufficient, so he was discharged with a naso-jejunal tube, which was removed four months later after clinical improvement.

Three months later (July 2022), he was hospitalized for recurrent SMA Syndrome with severe vomiting and weight loss (27.7 kg). A naso-jejunal tube was reinserted, and enteral feeding, parenteral nutrition, and oral feeding were maintained at home. After weight stabilization, the naso-jejunal tube and PICC were removed seven months later (Feb 2023).

However, a third relapse occurred in April 2023, and conservative treatment was no longer effective. A laparoscopic gastrojejunostomy was performed under general anesthesia. Oral feeding began the next day, and tube feeding was gradually tapered as symptoms improved. His weight, which had dropped to 29 kg, increased to 32.6 kg postoperatively, and he maintained a stable weight (~31 kg) without further recurrence.

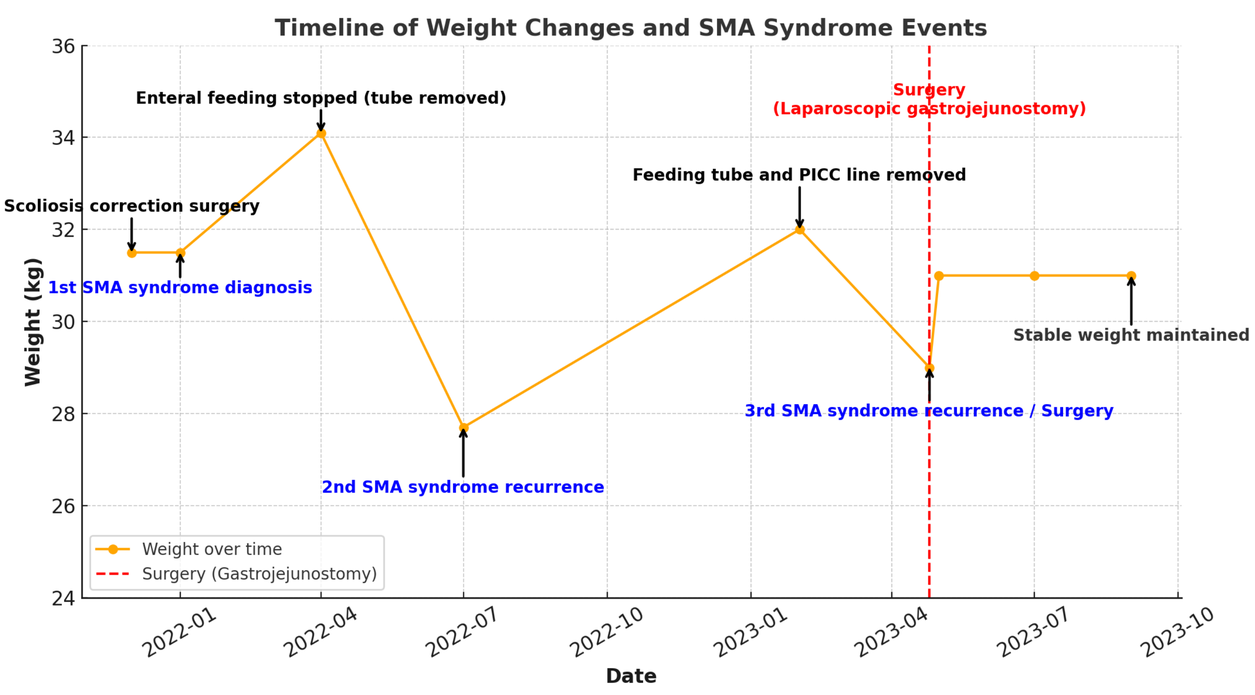

Figure 1. Timeline of weight changes and major clinical events in a Duchenne Muscular Dystrophy (DMD) patient with recurrent Superior Mesenteric Artery (SMA) syndrome
The first and second episodes of SMA syndrome were treated conservatively using nasojejunal tube placement. Following the third recurrence, the patient underwent laparoscopic gastrojejunostomy, which successfully prevented further recurrence, and he could maintain his weight afterwards.

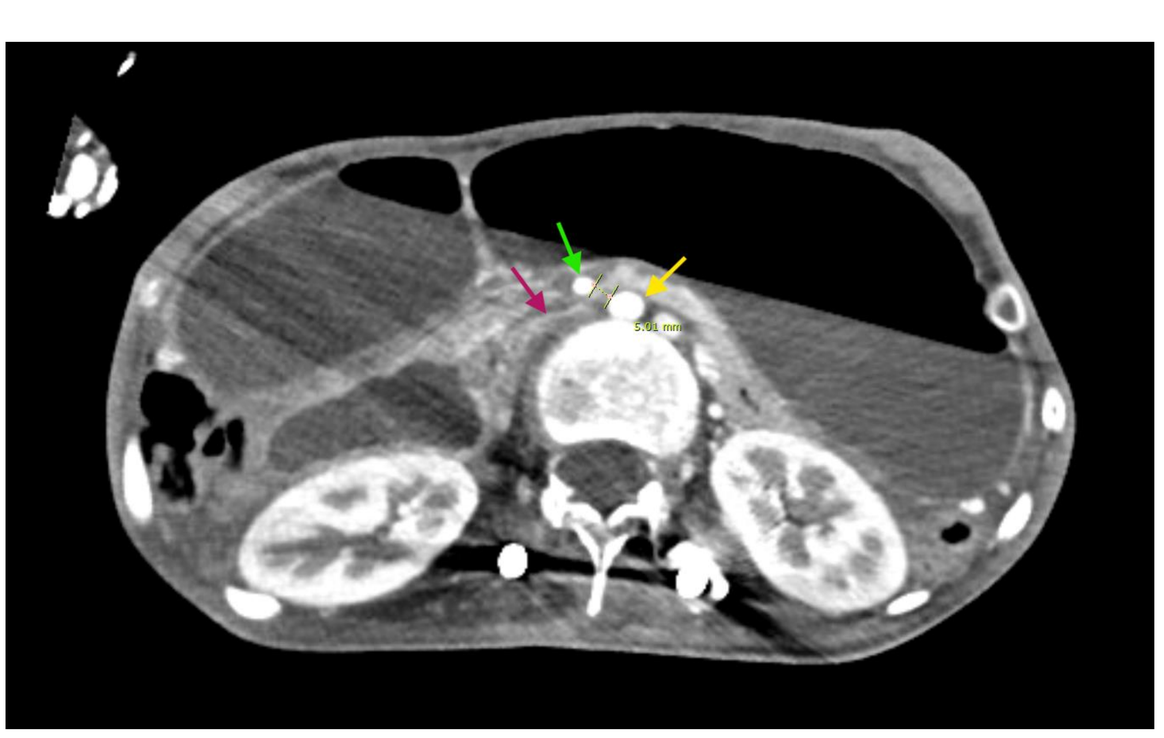

Figure 2. Axial contrast-enhanced abdominal CT scan demonstrating anatomical features of Superior Mesenteric Artery (SMA) syndrome.
The aorta (yellow arrow) and superior mesenteric artery (SMA) (green arrow) form a narrowed aortomesenteric distance, measuring 5.01 mm in this patient.
The third portion of the duodenum (pink arrow) is compressed between these two vessels, consistent with findings of SMA syndrome

Conclusion

In DMD patients, SMA Syndrome can initially be managed conservatively, but surgical treatment can be actively considered if relapses occur. Surgical correction is often avoided in patients with respiratory failure and severe paralysis. However, as in this case, surgery may be necessary when oral and enteral feeding fail to ensure weight gain or when nasal tube feeding is difficult due to NIV use.