J Metab Bariatr Surg. 2025 Apr;14(1):81-84
https://doi.org/10.17476/jmbs.2025.14.1.81
pISSN 2287-2930·eISSN 2508-5956

Journal of Metabolic and Bariatric Surgery JMBS

Case Report

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Bariatric Surgery for a Patient With Myosin Heavy Chain 9-Related Disorders (MYH9RD): A Case Report

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OPEN ACCESS

Received: Dec 16, 2024
Revised: Apr 11, 2025
Accepted: Apr 12, 2025
Published online: Apr 23, 2025

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ABSTRACT

Myosin heavy chain 9-related disorders (MYH9RD) are rare autosomal dominant genetic conditions caused by *MYH9* mutations, leading to macrothrombocytopenia, renal complications such as focal segmental glomerulosclerosis (FSGS), and other systemic manifestations. We report a case of 28-year-old male with MYH9RD and body mass index exceeding 47 kg/m^2, who successfully underwent laparoscopic sleeve gastrectomy. Despite challenges from bleeding tendency caused by macrothrombocytopenia and renal impairment caused by FSGS, thorough preoperative evaluation and management, including platelet transfusion, enabled surgery to proceed without complications. The patient achieved significant weight loss, from 147.6 kg preoperatively to 90.15 kg at 1 year postoperatively, with improvements in hypertension and metabolic parameters, including aspartate aminotransferase/alanine aminotransferase, hemoglobin A1c, triglycerides, and low-density lipoprotein levels. While MYH9RD is not directly associated with morbid obesity, this case highlights that comprehensive preoperative evaluation and risk management can lead to successful outcomes in bariatric surgery for MYH9RD patients.

Keywords: MYH9-related disorders; Morbid obesity; Bariatric surgery

INTRODUCTION

Myosin heavy chain 9-related disorders (MYH9RD) are a type of autosomal dominant genetic disorder caused by mutations in the *MYH9* gene on chromosome 22q12-13. This gene encodes the non-muscle myosin heavy chain class IIA (NMMHC-IIA) protein, which has 3 subtypes: A, B, and C. NMMHC-IIA plays a crucial role in maintaining the structure and function of cells, particularly in renal podocytes, platelets, and leukocytes [1-3]. MYH9 mutations lead to various clinical manifestations, including end-stage renal disease (ESRD), macrothrombocytopenia with leukocyte inclusion bodies, lens detachment, and sensorineural hearing loss [4]. Renal involvement often manifests as focal segmental glomerulosclerosis (FSGS), leading to proteinuria and progressive renal failure, potentially culminating in ESRD. Since platelets possess only the A subtype of the NMMHC-IIA protein, *MYH9* mutations result in giant, fragile platelets, causing macrothrombocytopenia [2].

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Funding
No funding was obtained for this study.

Conflict of Interest
None of the authors have any conflict of interest.

Author Contributions
Conceptualization: Park DJ; Data curation: Kim SH; Formal analysis: Kim SH; Supervision: Park DJ; Writing - original draft: Kim SH, Park DJ; Writing - review & editing: Park K, Kim C, Kim J, Kong SH, Lee HJ, Yang HK, Park DJ.

While several genes have been linked to increased body mass index (BMI) and fat distribution [5,6], no interactions between *MYH9* mutations and morbid obesity have been reported. In this case report, we present a patient with an *MYH9* gene mutation, complicated by FSGS-related impaired renal function and a bleeding tendency due to macrothrombocytopenia, who successfully underwent bariatric surgery and achieved significant weight loss.

CASE REPORT

A 28-year-old obese male (height: 177 cm, weight: 130 kg) presented to the nephrology outpatient clinic in October 2017 with proteinuria and hypertension (145/98 mmHg). His weight had significantly increased from 85 kg in his early 20s. He had a smoking history and was on antihypertensive medication. Laboratory tests revealed thrombocytopenia (platelet count: 79,000/μL) and impaired renal function (creatinine: 1.54 mg/dL, glomerular filtration rate [GFR]: 53.3 mL/min). The patient was advised to lose weight, quit smoking, and was prescribed dual renin-angiotensin system blockers for hypertension management. However, adherence to lifestyle changes and medication was poor, leading to further weight gain (145.7 kg by January 2020). Concurrently, renal function deteriorated (creatinine: 1.97 mg/dL, GFR: 44 mL/min), and a kidney biopsy performed in February 2020 confirmed FSGS, with 76.2% of glomeruli globally sclerosed (16 out of 21) and 9.5% segmentally sclerosed (2 out of 21). Gene sequencing revealed heterozygous mutation in the *MYH9* gene where a cytosine is replaced by thymine at position 3493, resulting in a change from arginine to cysteine at position 1165 in the protein sequence (MYH9, c.3493C>T, p.Arg1165Cys, heterozygote).

By September 2020, the patient was referred to the obesity clinic of familial medicine department, presenting with a weight of 147.6 kg (BMI: 47.1 kg/m^2). Despite the initiation of Saxenda (liraglutide) and lifestyle modification, compliance remained suboptimal. Consequently, he was referred to gastrointestinal surgery department for surgical treatment for morbid obesity. Preoperative blood test after admission (February 2021) showed thrombocytopenia (platelet count: 60,000/μL) with giant platelets in peripheral blood smear (hematoxylin and eosin stain, ×100; **Fig. 1**). After receiving platelet transfusion (platelet count: 74,000/μL), he underwent laparoscopic sleeve gastrectomy on February 15, 2021 (**Fig. 2**).

The patient adhered to a structured dietary plan postoperatively, starting from water intake on the second day to a semi-fluid diet by the fifth day. He was discharged on the postoperative fifth day without complications. Subsequent follow-ups revealed significant weight loss: 138 kg at 10 days, 124 kg at 2 months, 112.1 kg at 4 months, 101.7 kg at 7 months, 96.25 kg at 10 months, and 90.15 kg at 1 year postoperatively (**Fig. 3**). Correspondingly, blood pressure improved, recorded with 122/78 mmHg at 1 year postoperatively. Laboratory tests at 1 year

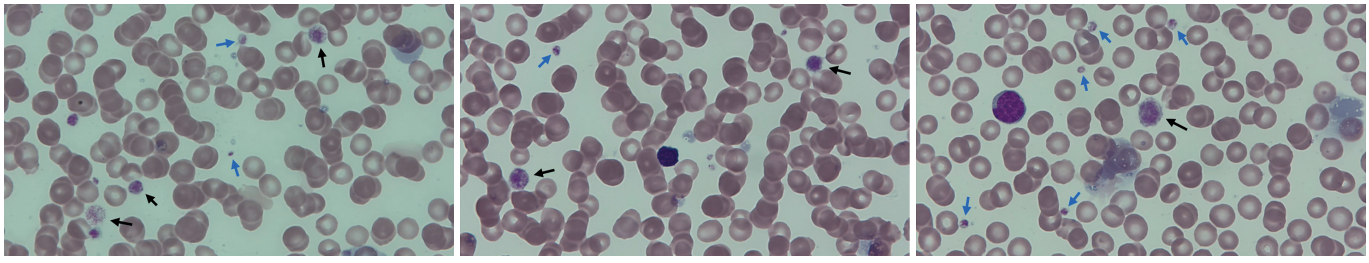

Fig. 1. Peripheral blood smear demonstrated physiological and pathological platelet morphology (black arrow: giant platelets, blue arrow: normal platelets; hematoxylin and eosin stain, ×100).

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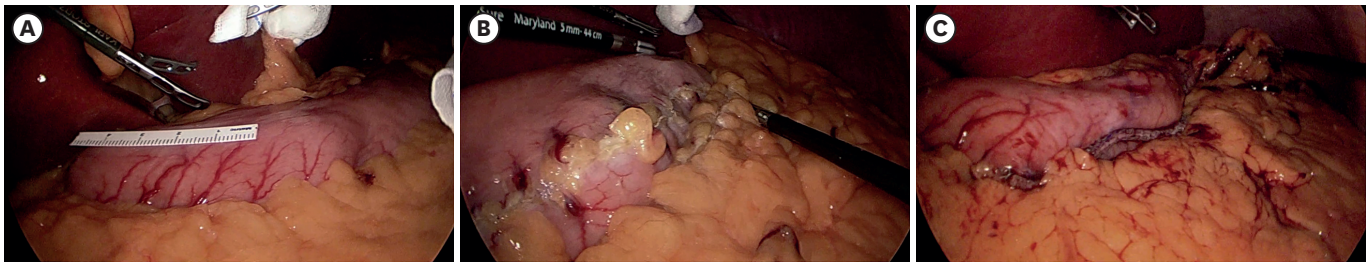
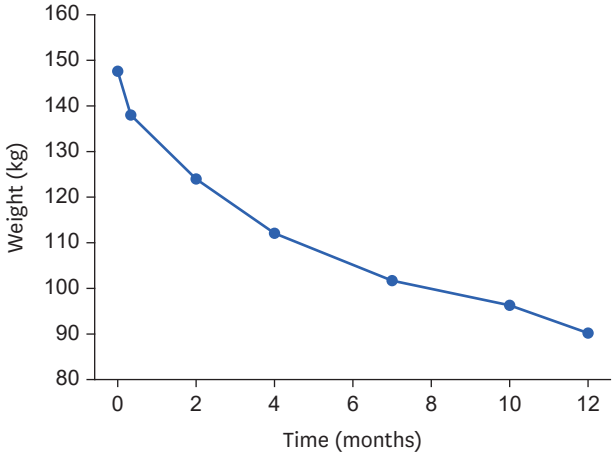


Fig. 2. Laparoscopic sleeve gastrectomy; description of each step.
(A) A 5 cm-distance from the pylorus was measured, which is the start point of greater omentum clearing. (B) Greater omentum was cleared from gastric greater curvature with energy-based device (LigaSure™; Covidien). (C) From the 5 cm-measured point, sleeve gastrectomy was conducted with laparoscopic linear stapler (ECHELON FLEX™; Ethicon); 38-Fr calibration bougie was inserted along the gastric lesser curvature before beginning gastric resection.



Time (months)	Body weight (kg)
Preoperative	147.6
Postoperative	
10 days	138.0
2 months	124.0
4 months	112.1
7 months	101.7
10 months	96.3
12 months	90.2

Fig. 3. Body weight loss during one-year period after bariatric surgery.
The patient achieved significant weight loss, from 147.6 kg preoperatively to 90.15 kg at one year postoperatively.

showed normalized aspartate aminotransferase/alanine aminotransferase, hemoglobin A1c, triglycerides, and low-density lipoprotein levels. However, renal function showed only minimal change (creatinine: 1.95 mg/dL, GFR: 44 mL/min). The patient developed postoperative gastroesophageal reflux symptoms, managed with proton pump inhibitors (PPI) for 1 year. At the 1-year follow-up, gastroscopy confirmed Los Angeles classification B reflux esophagitis, necessitating continuous PPI therapy.

1. Ethical approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the Institutional Review Board (IRB) of the Seoul National University Hospital (IRB No.: 2408-146-1565) and with the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards. Informed consent was obtained from the patient in this case report.

DISCUSSION

Patients with MYH9RD can encounter significant perioperative challenges due to bleeding tendency associated with macrothrombocytopenia (giant, fragile, and dysfunctional platelets) and impaired renal function. In surgical patients, preoperative assessment of bleeding risk is essential. In this case, we consulted the hematology department regarding

https://doi.org/10.17476/jmbs.2025.14.1.81

perioperative risk in the context of macrothrombocytopenia. It was recommended by the hematology department to maintain a platelet count of 70,000–80,000/μL throughout the perioperative period, with platelet transfusion as needed. In the event of postoperative bleeding, tranexamic acid, additional platelet transfusion, and desmopressin were suggested as part of the hemostatic strategy. Given the presence of FSGS, renal considerations were also critical. Careful fluid management was undertaken to avoid both volume overload and depletion. Nephrotoxic agents were avoided, and potassium-free intravenous fluids were administered. Cardiovascular risk assessment was also performed due to the patient's history of hypertension and macrothrombocytopenia. Following a comprehensive evaluation, the cardiology department considered the patient to have an acceptable cardiac risk for surgery.

In terms of surgical technique, cautious tissue handling and the use of hemo-clips for vessel ligation were implemented. A bipolar electrothermal vessel sealing device (LigaSure™; Covidien, Mansfield, MA, USA) was used to enhance hemostasis and facilitate tissue dissection. During gastric resection, laparoscopic surgical staplers (ECHELON FLEX™; Ethicon, Somerville, NJ, USA) were employed, and a hemostatic agent was applied along the staple line to minimize the risk of postoperative bleeding.

The patient developed gastroesophageal reflux disease postoperatively. However, in Korea, where the incidence of gastric cancer is relatively high, sleeve gastrectomy is generally preferred over Roux-en-Y gastric bypass, as it allows easier long-term endoscopic surveillance.

In this case report, the patient with MYH9RD, complicated by macrothrombocytopenia, FSGS, and morbid obesity, successfully underwent sleeve gastrectomy, resulting in significant weight loss and improved hypertension control over a year with thorough preoperative evaluation and risk management.

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