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Recurrent Variceal Hemorrhage in Non-Cirrhotic Portal Hypertension with Thymoma-Related Myasthenia Gravis : A Case Report

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ABSTRACT

Background: Extrahepatic portal vein obstruction (EHPVO) in adults is uncommon and usually is accompanied by portal hypertension but preserved liver function. Its association with thymoma-associated myasthenia gravis (MG) presents with distinct therapeutic challenges.

Case Presentation: We present a 33-year-old man with EHPVO and high-risk esophageal varices that needed to be treated with repeated endoscopic variceal ligation (EVL). He also had invasive thymoma complicated by MG with high titers of acetylcholine receptor antibodies. The management was done via a multidisciplinary team including hepatology, neurology, and surgery. Beta-blockers and plasmapheresis stabilized the patient before thymectomy.

Conclusion: The simultaneous management of risk of variceal bleeding and MG-associated thymoma underscores the importance of individualized, multidisciplinary care in multisystem disorders.

Keywords: *Extrahepatic Portal Vein Obstruction, Esophageal Varices, Myasthenia Gravis, Thymoma, Plasmapheresis, Portal Hypertension, Case Report*

1. INTRODUCTION

Extrahepatic portal vein obstruction (EHPVO) is a rare vascular condition with extrahepatic portal vein obstruction, causing portal hypertension, splenomegaly, and formation of varices without cirrhosis [6]. Treatment is aimed at the prevention of gastrointestinal bleeding and maintenance of hepatic function.

Myasthenia gravis (MG) is an autoimmune chronic neuromuscular disease found frequently in association with thymoma, for which thymectomy is curative [7]. The combination of MG and EHPVO in one patient complicates perioperative management, particularly because it poses a risk for bleeding complications and hemodynamic instability [8].

2. CASE PRESENTATION

A 33-year-old man came with a history of pain in the abdomen and was diagnosed incidentally with EHPVO in the year 2022.

Imaging showed splenomegaly and severe portal-systemic collaterals. Upper GI endoscopy showed grade II–III esophageal varices.

The patient received repeated sessions of EVL for 9 months because of recurrence of high-risk varices with signs of red color [9].

Table 1. Summary of Hepatic and Neuromuscular Findings

Parameters	Findings
Liver Function (Bilirubin)	1.12 mg/dL
INR	1.1
Albumin	4.4 g/dL
Liver Stiffness (Elastography)	4.2 kPa (no fibrosis)
Imaging	Cavernomatous transformation of portal vein
Endoscopy	Grade II – III varices (multiple EVLs)
Neurological symptoms	Slurred speech, fatigability
AChR Antibody	12.45 nmol/L (Elevated)
Thoracic CT	Invasive anterior mediastinal thymoma

Neurological assessment established Myasthenia gravis (MG) with high levels of acetylcholine receptor antibodies. Thoracic imaging revealed a large anterior mediastinal mass, which is consistent with invasive thymoma [10].

Since the patient had high titers of AChR antibodies and bulbar signs, the patient was scheduled for five courses of plasmapheresis spread over 10 days before proceeding to thymectomy. Portal hypertension was started on carvedilol, although the titration was withheld because of hypotension. Surgical clearance was postponed until follow-up endoscopy revealed low-risk varices [11].

3. DISCUSSION

EHPVO raises special management problems that differ from cirrhotic portal hypertension. While liver function is intact, patients have a high risk of variceal bleeding through portosystemic collaterals [12]. Endoscopic variceal ligation is still the first-line treatment, with the addition of beta-blockers to decrease portal pressure [13]. Yet, recurrence of varices is a problem, particularly in non-cirrhotic patients [14].

Myasthenia gravis (MG) with thymoma usually necessitates thymectomy. Immunomodulation prior to surgery, e.g., plasmapheresis, is necessary in patients with bulbar weakness or elevated antibody titers to reduce perioperative risk [2,15].

Table 2. Multidisciplinary Management Plan and Rationale

Discipline	Intervention	Rationale
Hepatology	EVL, Carvedilol	Prevent recurrent variceal bleeding
Neurology	Plasmapheresis (5 sessions)	Lower AChR antibodies; Reduce

		perioperative complications
Surgery	Thymectomy (post stabilization)	Definitive treatment for thymoma-associated MG
Anaesthesia	Perioperative MG precautions	Minimize respiratory / hemodynamic complications

This case demonstrates how multisystem diseases may pose conflicting management priorities: bleeding hazards from EHPVO vs. MG's immunological instability. Coordination between specialties was essential in order to tackle these challenges safely [4,5].

4. LIMITATIONS

This case is also limited by absence of data on long-term outcomes following thymectomy. Histological diagnosis of thymoma type and genetic or immunohistochemical studies were not done.

5. CONCLUSION

This case emphasizes the need for multidisciplinary management of patients with rare, co-morbid conditions. Successful coordination of hepatology and neurology management plans permitted safe planning of thymectomy in a patient with EHPVO and MG. More work is necessary to define best practices in such complex clinical situations.

Conflicts of Interest

None declared.

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Patient Consent

Informed consent was received from the guardians of the patient for publication. Identifiers have been removed.

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