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# Myasthenia Gravis and Abdominal Aortic Aneurysm: A Rare Combination

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## Abstract

Abdominal aortic aneurysm in a patient with myasthenia gravis (MG) is extremely rare. We present a 64-year-old male with MG and an asymptomatic abdominal aortic aneurysm treated endovascularly. After extubation, he suffered a cardiac arrest due to an acute myocardial infarction. Cardiopulmonary resuscitation and a primary coronary angioplasty led to a satisfactory outcome. Special care is needed due to higher rates of postoperative complications in these patients.

**Keywords**

- myasthenia gravis
- abdominal aortic aneurysm
- endovascular abdominal aneurysm repair
- acetylcholine receptor

## Introduction

Endovascular abdominal aneurysm repair (EVAR) is widely adopted due to the associated lower morbidity and mortality. Nevertheless, specific comorbid conditions like myasthenia gravis (MG) may lead to increased complication and mortality rates. We report a 64-year-old male with MG and an abdominal aortic aneurysm (AAA), a scarce combination in the literature. There is limited evidence regarding the outcome of MG patients after vascular procedures. We report our postoperative complications and discuss the relevant literature.

## Case Presentation

A 64-year-old male was admitted for elective repair of an asymptomatic infrarenal AAA. He was a current smoker, and he suffered from osteoporosis and acetylcholine receptor (AChR) antibody-positive MG diagnosed 14 years ago. He underwent a thymectomy 13 years ago. At present,

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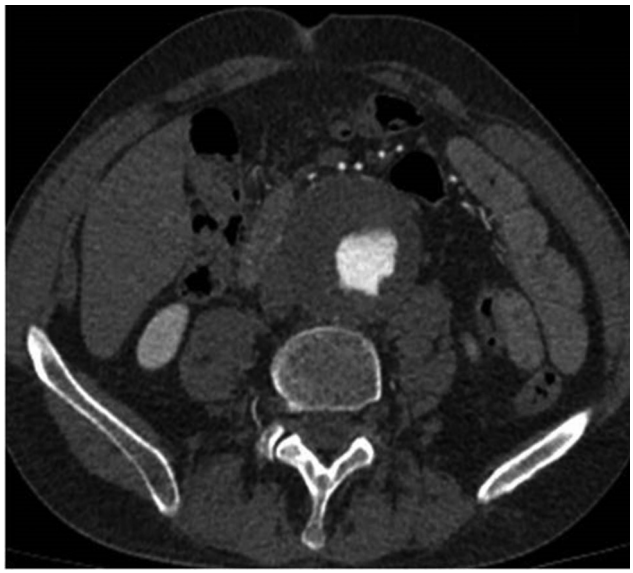

**Fig. 1** Axial view of the preoperative computed tomography angiography depicting the abdominal aortic aneurysm.

myasthenia remained asymptomatic under pharmacological treatment which included pyridostigmine bromide 60 qd and prednisolone 5 bid.

Preoperative computed tomographic angiography (CTA) showed an infrarenal AAA measuring 5.7 cm in diameter (►**Fig. 1**). Preoperative echocardiographic stress test was normal with an ejection fraction of 60%. Pedal pulses were palpable and neurological status was normal.

Anesthesia was induced with bolus propofol and remifentanil, without muscle relaxants using a laryngeal mask (LMA). General anesthesia was maintained by total intravenous anesthesia (TIVA) with target-controlled infusion of propofol and remifentanil. The patient underwent EVAR with the placement of an ALTO Abdominal Stent Graft System (Endologix Inc., Irvine, CA) by the aid of a portable C-arm, through common femoral arteries cutdown (►**Figs. 2** and **3**). He received 4,000 IU unfractionated heparin, intravenously. The duration of the operation was 2.5 hours. Kerma-area product and fluoroscopy time were 2.36 mGy·m$^2$ and 26.39 minutes, respectively, with the use of 170 mL contrast I.V. Perioperatively, steroids were given to protect from the surgical stress.

Postoperatively, TIVA was discontinued, and after a few minutes, the patient was fully awake (bispectral index = 95), breathing calmly on his own (tidal volume > 250 mL). He was hemodynamically stable, and LMA was removed. He was transferred to the postanesthesia care unit, where 5 minutes later he suddenly became unresponsive, hemodynamically unstable, and finally, pulseless. Advanced cardiac life support was immediately initiated, and the patient was intubated. Return of spontaneous circulation occurred after 6 minutes and three cycles of cardiopulmonary-resuscitation. ECG revealed ST-segment elevation in anterolateral leads, indicative of an anterior acute myocardial infraction (AMI). Transthoracic echo showed adequate ejection fraction (55%). He was hemodynamically stable with low doses of noradrenaline. Blood gases showed mild metabolic acidosis, which was reversed. He was sedated with low doses of

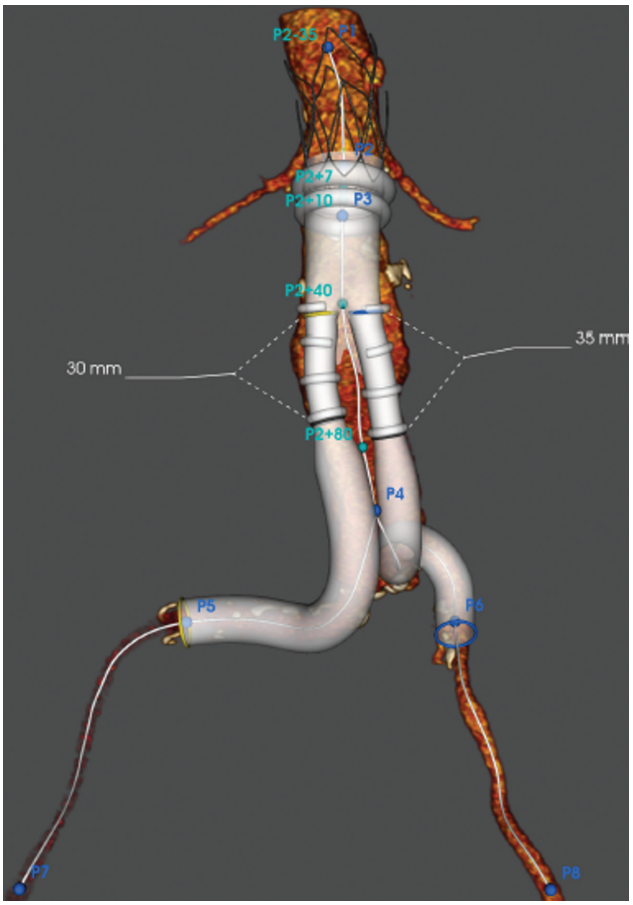


**Fig. 2** Preoperative graft sizing.

propofol and midazolam, and he was transferred to the Radiology Department for urgent imaging.

Brain CT and carotid, thoracic and abdominal CTA (►**Fig. 4**) were normal, except for a bilateral chronic cervicocranial arterial dissection (CCAD).

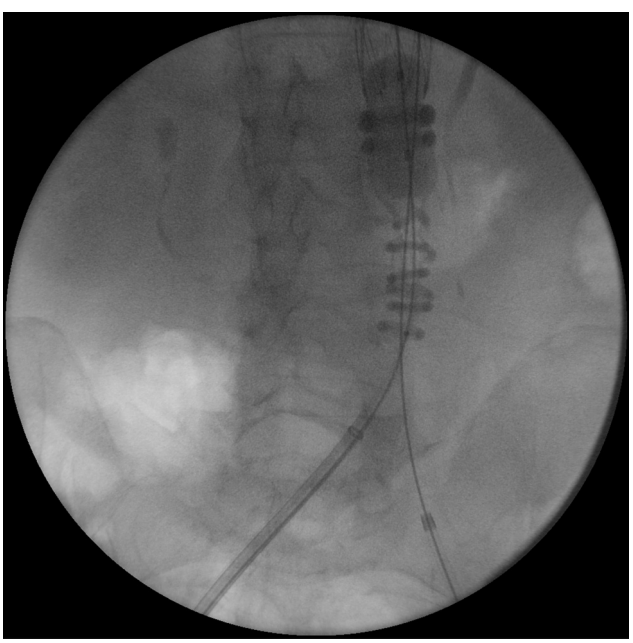

**Fig. 3** Intraoperative main trunk ballooning.

 

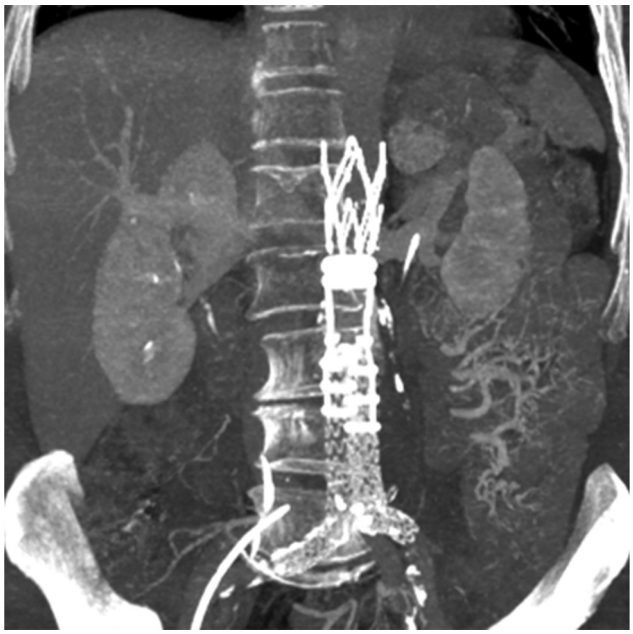

**Fig. 4** Postoperative computed tomography angiography depicting the graft placement.

Consequently, he was transferred to the catheterization laboratory where a coronary angiogram revealed a 90% stenosis of the left anterior descending artery with fresh thrombus. We assumed that an atheromatous plaque causing moderate stenosis that was not identified in the preoperative stress test had ruptured postoperatively and that accumulated thrombus worsened the lumen stenosis to a critical level. A drug-eluting stent (Resolute Integrity DES, $3 \times 18$mm, Medronic, Minneapolis, MN) was placed with satisfactory result (►**Video 1** and ►**Fig. 5**). Brain-CT was repeated after 24 hours and showed no signs of cerebral ischemia or edema. After 48 hours the patient was fully alert, neurologically normal, and was extubated successfully. Cardiac enzymes were found to be elevated (HS Troponin I: 5393.40 pg/mL). He was discharged on the 12th postoperative day on dual antiplatelet regimen (acetylsalicylic acid qd and ticagrelor 90 mg bid).

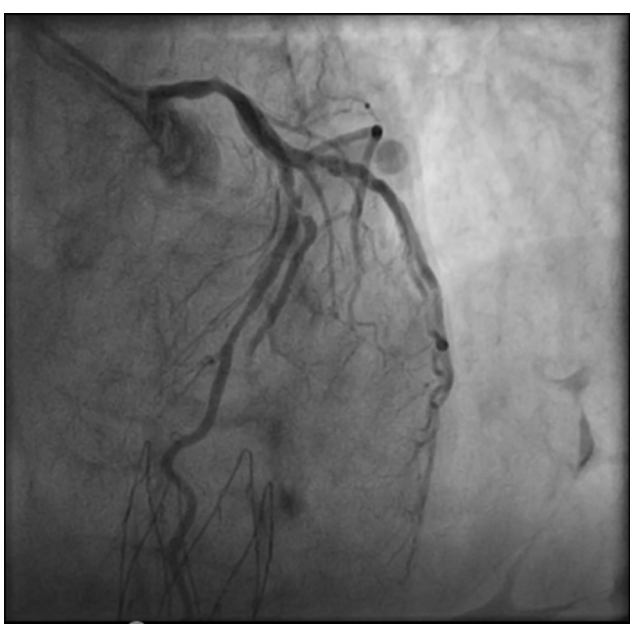

**Fig. 5** Coronary angiogram.

**Video 1**

Coronary angioplasty. Online content including video sequences viewable at: https://www.thieme-connect.com/products/ejournals/html/10.1055/a-2051-7678.

## Discussion

MG is an autoimmune disorder that targets the neuromuscular junction, characterized by fluctuating fatigue and weakness, initially affecting the extraocular muscles but with frequent generalization encompassing bulbar and respiratory musculature.[1] AChR antibodies (Abs), which were present in our patient, are highly specific in confirming the diagnosis.[2]

Although cardiovascular events due to MG or its treatment are rare, it is reported that in 47% of patients Abs against striatal myocardial antigens are present. These Abs act outside the neuromuscular junction, inciting cardiac muscle inflammation.[2,3] This makes the heart a potential second autoimmune target, especially in the presence of a thymoma.[4]

Cardiac manifestations in MG include arrhythmias, giant cell myocarditis, heart failure, and cardiac arrest.[2,4] Stress cardiomyopathy or Takotsubo (broken heart) syndrome in MG may be exacerbated by physical or emotional stress.[4] Moreover, MG treatment with anticholinesterase inhibitors (AChEi) may also trigger acute cardiac events. This is due to a greater supply of acetylcholine (Ach) at the synaptic cleft.[3] Excessive Ach has arrhythmogenic effects or may cause coronary vasoconstriction when endothelial damage occurs. Only four such cases of AChEi-mediated coronary vasospasm have been reported so far.[3] Of course, cardiac symptoms may be unrelated to MG, as in our case, where an AMI occurred due to atherosclerotic coronary obstruction.

Generally, AMI occurs in 1 to 12% of MG patients.[3] Furthermore, these patients face a higher risk of complications, including AMI after major surgical procedures. The general risk was 1.6 to 15% higher after hip and knee arthroplasty, while AMI had an odds ratio: 7.4 during hospitalization in a recent report.[1] Surprisingly, the widespread use of preoperative stress testing in many centers did not reduce perioperative major adverse cardiovascular events.[5]

MG and AAA coexistence is extremely rare. In the English literature, only one case report has been published so far, presenting a ruptured inflammatory infrarenal AAA treated by open repair. The patient eventually died after a 3-month hospitalization.[6] Another case report describes a thoracoabdominal aneurysm after chronic dissection treated successfully by open repair.[7] Spontaneous CCAD occurs more

frequently in autoimmune diseases, possibly caused by an immune-related local inflammation.[8]

Postoperative AMI is reported to have an incidence of 0.8 to 1% after EVAR,[5] with a 5.9% mortality rate, while cardiac arrest presents in 0.35% after EVAR, leading to death in 33%.[5] Although AMI after major surgery in MG patients was not increased in one report by Chang et al[9], there is a paucity of data regarding AMI after vascular procedures in MG patients.

EVAR can be performed under general, epidural, spinal, or local anesthesia. In one report by Bakker et al [10] cardiac events were observed in 6.4% of patients receiving general anesthesia versus 0.8% in patients receiving local or regional anesthesia. General anesthesia increases the systemic inflammatory response induced by surgery.

It is interesting that the low-profile ALTO graft inserted in small iliac artery diameters of only 5 mm showed excellent behavior during our cardiopulmonary resuscitation and profound hypotension. No limb or graft thrombosis occurred, despite the fact that the patient was not on regular antiplatelets.

In conclusion, we suggest that surgeons inform the patients of the higher surgical risk in MG, control meticulously the MG preoperatively, avoid muscle relaxants and other medications that are contraindicated in MG, and give special care during the postoperative period.

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Conflict of Interest
The authors declare no conflict of interest related to this article

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