Aortic dissection four months after SARS-CoV-2 infection in a patient with Fabry disease whose targeted treatment was stopped 2 months earlier

Marcin Kamil Dobrowolski[1], Magdalena Marczak[2], Rafał Dąbrowski[2]

[1]Department of Cardiology, WKK Westküstenkliniken Heide und Brunsbüttel, Heide, Germany
[2]National Institute of Cardiology, Warszawa, Poland

Correspondence to:
Marcin Kamil Dobrowolski, MD, Department of Cardiology, WKK Westküstenkliniken Heide und Brunsbüttel, Esmarchstraße 50, 25746 Heide, Germany,
phone: +49 160 143 34 38,
e-mail: dobmarcin@gmail.com

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Fabry disease is a rare storage disease involving the absence or decreased activity of alpha-galactosidase A enzyme [1]. This leads to the accumulation of glycolipids (globotriaosylceramides [GB3]) with consistent cell damage and fibrosis. It results mostly in renal, cardiac, neurological, and other systems' complications [2].

A 43-year-old male patient with a history of Fabry disease, hypertension, dyslipidemia, and obesity was transported by the ambulance service in a critical condition with suspected acute coronary syndrome to the emergency room of one of German hospitals. The patient was not vaccinated against COVID-19 and suffered from COVID-19 four months earlier. For this reason, he was not hospitalized. The ambulance service was called by the patient's mother after she noticed her son's speech disorders lasting about 2 hours and progressive disturbances of consciousness. For over 4 days, the patient had complained of chest pains radiating to the back, arms, and abdomen, as well as nausea and vomiting. Due to the prevailing COVID-19 pandemic, the patient had not received intravenous therapy (agalsidase alfa) for 2 months.

In the emergency room, the patient was deeply unconscious (Glasgow Coma Scale [GCS], 6–7), with saturation dropping to 60% despite intensive oxygen therapy. Pulse was regular but imperceptible, heart tones muted, pressure on the right arm was 130/80 mm Hg, on the left — 165/80 mm Hg. Significant stasis in the jugular veins has been observed. The electrocardiogram record showed significant ST-segment elevation in the leads from the inferolateral and anterior walls (Figure 1A, B).

Echocardiography was performed urgently due to suspected aortic dissection. A large amount of fluid was observed in the pericardium (Figure 1C).

Contrast-enhanced computed tomography of the thoracic and abdominal aorta was performed (Figure 1D–E). The diagnosis of aortic dissection was confirmed. It was decided to intubate the patient to avoid aspiration and to immediately transport him by air to the university department for further surgical treatment. While waiting for transport, the patient experienced cardiac arrest four times in the asystole mechanism. Despite the continuation of the resuscitation procedure, in the presence of the air ambulance service and the lack of recovery of the electromechanical activity of the heart, a decision was made to discontinue medical activities.

Despite his relatively young age, the patient had several risk factors for cardiovascular diseases: hypertension, dyslipidemia, obesity, and confirmed and treated Fabry disease. Aortic dissection is not a typical symptom of Fabry disease. In this complex clinical situation, it is difficult to unequivocally state, but also to exclude what would have the direct impact on extensive aortic dissection [3].

Article information

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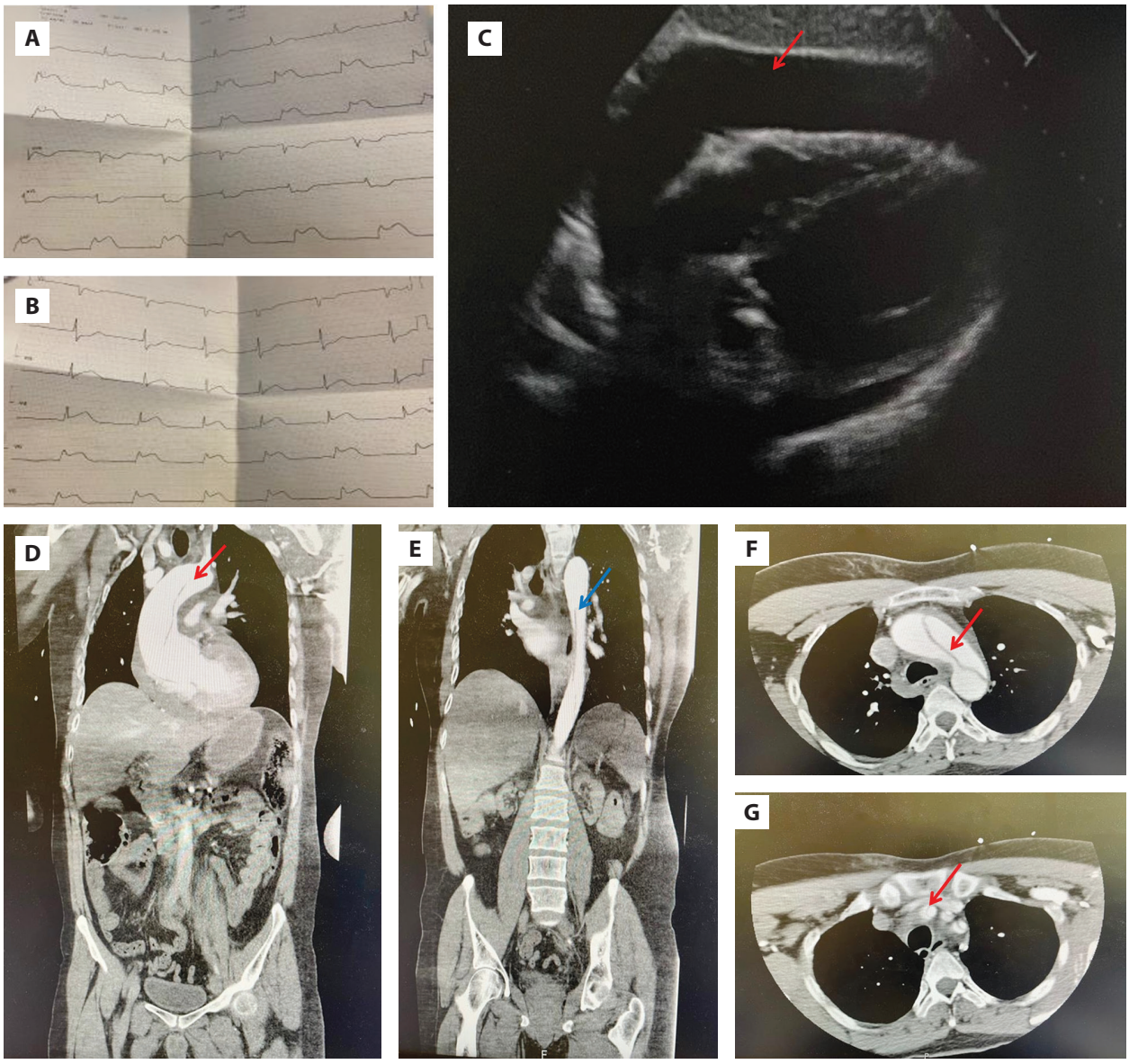

Figure 1. A, B. Electrocardiogram: significant ST-segment elevation in leads II, III, aVF, V3–6. **C.** Echocardiogram: a large amount of fluid in the pericardium (the arrow). **D, E.** Contrast-enhanced computed tomography with coronal plane showing dissection of the ascending aorta and the aortic arch (the red arrow) and the descending thoracic aorta (the blue arrow). **F, G.** Contrast-enhanced computed tomography with transverse plane showing dissection across the aortic arch and initial segment of the brachiocephalic trunk, the left common carotid, and the left subclavian artery (the red arrows)

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