Haemolytic anaemia due to stenosed double-reinforced grafts after surgical repaired aortic dissection

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Abstract

Haemolytic anaemia due to a stenosed graft is a rare complication after surgery for aortic dissection. We present the case of a patient with haemolytic anaemia and heart failure, who had undergone emergent ascending aorta replacement for type A acute aortic dissection 5 years earlier. Chest computed tomography revealed severe graft stenosis of the proximal anastomosis and transthoracic echocardiography showed severe aortic regurgitation. Surgical treatment was necessary because of heart failure and myocardial ischaemia due to haemolytic anaemia and aortic regurgitation. During the operation, we found an inner graft surrounded by an outer graft and a dilated lumen between the double-reinforced grafts compressing the inner graft. We successfully reconstructed the aortic root with a total arch replacement. To the best of our knowledge, there are no cases in which haemolytic anaemia and AR developed in a patient with acute aortic dissection surgically treated by such a mechanism.

Keywords: Haemolytic anaemia • Surgical repaired aortic dissection and aortic regurgitation

INTRODUCTION

Type A acute aortic dissection (AAD) is the most lethal disease of the aorta, with high morbidity and mortality. Usually, the fragility of the aortic wall makes surgical treatment difficult. Thus, reinforcement of the dissected aortic wall at the anastomosis site is key to prevent bleeding. We report a case of successful treatment of haemolytic anaemia and aortic regurgitation (AR) with stenosed double-reinforced grafts.

CASE REPORT

A 63-year old woman had undergone emergent ascending aorta replacement because of type A AAD 5 years earlier at another hospital, with difficulty in controlling bleeding during the operation. The patient's postoperative course was uneventful, and she returned for a follow-up visit. Although she was totally asymptomatic, laboratory data indicated haemolytic anaemia with a serum haemoglobin (Hb) level of 7 g/dl for the past 3 months. Furthermore, she had chest pain on exertion and general fatigue with macroscopic haematuria for a week, and was referred to the haematology specialists in our hospital. The Hb and serum lactate dehydrogenase (LDH) concentrations were 5.3 g/dl and 3974 IU/l, respectively, and macroscopic haematuria was seen. Peripheral blood smear revealed numerous schistocytes and the Coombs' test was negative. These findings suggested a mechanical rather than immunological destruction of red blood cells. Furthermore, she complained of chest pain at rest with ST-segment depression on electrocardiogram following a worsening of the anaemia. Transthoracic echocardiography (TEE) showed no asynergy, but severe AR without dilation of the aortic root. Chest computed tomography (CT) revealed a severe graft stenosis of the proximal anastomosis (Fig. 1). Although we suspected a fragmentation of red blood cells by a turbulent flow at the proximal anastomosis, we could not detect an accelerated flow by TTE or by transoesophageal echocardiography. Chest pain and ST segment depression on electrocardiogram resolved by red blood cell transfusion. However, heart failure caused by AR was exacerbated the next day. Thus, we decided to do a reoperation to repair the proximal anastomosis and to explore the stenosed lesion causing haemolysis and AR. We found that there were double-reinforced grafts; the inner graft was surrounded by an outer graft (Fig. 2A), and the dilated lumen between the inner graft and outer graft was compressing the inner graft. This double-reinforced graft was resected, the aortic root was reconstructed (Fig. 2B), and a total arch replacement was performed using a 26-mm InterGard (Intervascular S.A., France). She did not need an aortic valve repair because the improvement of AR was confirmed by TEE during the operation. Postoperatively, macroscopic haematuria and haemolytic anaemia resolved completely. At discharge, the LDH level had decreased to 213 IU/l, and she returned for a follow-up visit.

DISCUSSION

Haemolytic anaemia is a rare complication for patients who undergo surgical treatment for aortic disease. In this case, two
important factors, haemolytic anaemia and AR, led to the reoperation.

Firstly, in relation to haemolytic anaemia after an operation for AAD, perivalvular leakage after mitral valve replacement is a well-known cause of haemolytic anaemia. Stenosed graft or kinked prosthetic graft is also a rare cause of haemolytic anaemia after aortic surgery. Graft constriction due to the dilation of the aorta proximal to the graft was pointed out as a cause of haemolytic anaemia by Izumi et al. [1]. Vilacosta et al. [2] reported a patient with external graft compression and supravalvular aortic stenosis induced by a pseudoaneurysm following dehiscence of the suture line. However, in the most recent reports, haemolytic anaemia was due to stenosis at the site of anastomosis by inverted felt strips, which was used to reinforce the aortic stump [3, 4]. Teflon felt strips have been used to reinforce the anastomosis of the dissecting aortic wall and to avoid residual dissection and serious bleeding, but it might be a cause of supravalvular aortic stenosis at the proximal anastomosis site. Therefore, there have been several recommendations for reinforcement of the anastomosis. In the present case, the inner graft was surrounded by an outer graft in order to control bleeding during the previous operation. However, the sutures of the proximal anastomosis had recently failed, resulting in dehiscence of the double-reinforced grafts. Blood flow to the false lumen, which was constructed between the inner graft and outer graft, caused the false lumen to dilate, and the dilated lumen compressed the inner graft; this increased the shear stress on the red blood cells, causing haemolysis. Although we could not detect accelerated flow induced by supravalvular aortic stenosis by TTE or transoesophageal echocardiography, we speculate that the compressed inner graft was the cause of supravalvular aortic stenosis and haemolytic anaemia.

In this case, heart failure and myocardial ischaemia caused by severe AR led us to perform a reoperation. Although AR is a rare complication occurring in the chronic phase of surgically treated type A AAD, it is serious because it often requires reoperation. The aortic valve had not been replaced at the prior operation for AAD, or recurrent dissection at the original site or at a second site, which is suspected to be a cause of progressive AR in the chronic phase. In this patient, we suspected that the cause of AR was not redissection, but supravalvular aortic stenosis. The reason was that incomplete coaptation of the aortic leaflets, or aortic leaflet prolapse by invasion of the dissection flap into the aortic leaflet, was not observed in preoperative transoesophageal echocardiography. The mechanism responsible for the development of AR in the present case appears to be elevated aortic pressure proximal to the stenosed graft.

As suggested by our case report, haemolytic anaemia in the chronic phase of a surgically repaired AAD is a rare but lethal complication requiring a reoperation with a varied mechanism. It combined not only supravalvular aortic stenosis but also AR. Thus, a simple and sure method to reinforce the dissected wall is necessary despite its fragility.

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