Interactive CardioVascular and Thoracic Surgery 18 (2014) 230–233
doi:10.1093/icvts/ivt481 Advance Access publication 7 November 2013

Haemolytic anaemia resulting from the surgical repair of acute type A aortic dissection

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Received 26 June 2013; received in revised form 11 September 2013; accepted 17 September 2013

Abstract

OBJECTIVES: Haemolytic anaemia after acute aortic dissection surgery is extremely rare. We report 4 cases of haemolytic anaemia with different aetiologies.

METHODS: Four patients underwent emergency operation for acute type A aortic dissection and subsequently developed haemolytic anaemia.

RESULTS: Case 1: a 41-year old man underwent hemiarch replacement. We performed total arch replacement 3 years postoperatively, which revealed that haemolytic anaemia was induced by proximal anastomotic stenosis caused by inverted internal felt strip. Case 2: a 28-year old man diagnosed with Marfan syndrome underwent total arch replacement. Five months postoperatively, we noted severe stenosis at the previous distal anastomotic site, which caused the haemolytic anaemia, and performed descending thoracic aortic replacement for a residual dissecting aneurysm. Case 3: a 49-year old man underwent hemiarch replacement. Three years postoperatively, we performed total arch replacement for a residual dissecting aortic arch aneurysm and repaired a kinked graft responsible for haemolytic anaemia. Case 4: a 42-year old man underwent total arch replacement. Eighteen months later, we performed descending thoracic aortic replacement. We repaired a portion of the ascending aorta as haemolityc anaemia was induced by kinking of a total arch replacement redundant graft.

CONCLUSIONS: All the haemolityc anaemia patients were successfully released after surgical reintervention.

Keywords: Haemolytic anaemia • Acute aortic dissection • Reoperation • Inverted internal felt strip • Kinked graft

INTRODUCTION

Operative survival for acute aortic dissection has recently improved because of remarkable advancements in surgical techniques and perioperative management.

Although, most intraoperative concern is directed towards bleeding from the anastomotic site, anastomotic site-related intraoperative complications persist. Various techniques for the reinforcement of anastomoses, such as using two layers of Teflon felt, gelatin–resorcinol–formalin (GRF) glue and the elephant trunk technique are available. Haemolysis associated with prosthetic grafts or felt strips is an extremely rare complication. Here, we report 4 cases of haemolytic anaemia resulting from the surgical repair of acute type A aortic dissections.

CASE REPORTS

Case 1

A 41-year old man underwent emergency ascending aortic replacement for acute type A aortic dissection at another institution 4 years previously, where the proximal and distal anastomotic sites were reinforced using internal and external felt strips and GRF glue. His postoperative course was uneventful. Three years postoperatively, he presented with haemolytic anaemia and systolic murmur. His haemoglobin (Hb), lactate dehydrogenase (LDH) and haptoglobin levels were 9.8 g/dl, 953 IU/l and 5 mg/dl, respectively, and the reticulocyte percentage was 45%. Haemolytic anaemia was suspected. Transthoracic echocardiography failed to demonstrate abnormal acceleration flow at the proximal and distal anastomotic sites. Computed tomography (CT) revealed no signs of graft stenosis at the proximal anastomotic site or an anatomical kinked graft (Fig. 1).

Although definitive diagnosis could not be obtained, we strongly suspected haemolytic anaemia, so we decided to reoperate. We found severe stenotic aortic lumen at the proximal anastomotic site caused by an inverted internal felt strip. We removed the felt strip and performed proximal reanastomosis and total arch replacement for the residual arch aneurysm. His postoperative course was uneventful and he was discharged on postoperative day 20.

Case 2

A 28-year old man, diagnosed with Marfan syndrome underwent emergency total arch replacement and the elephant trunk

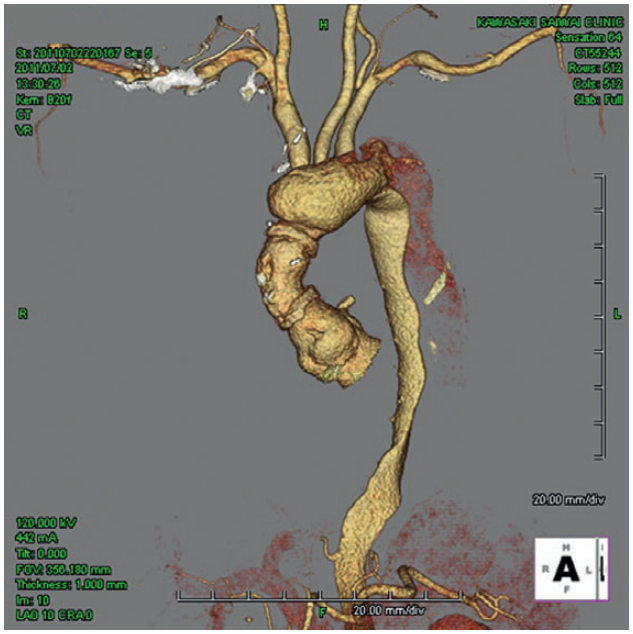

Figure 1: Preoperative CT revealed no abnormality at the anastomotic site and no kinked graft. Thus, the CT scan did not reveal the cause of haemolytic anaemia.

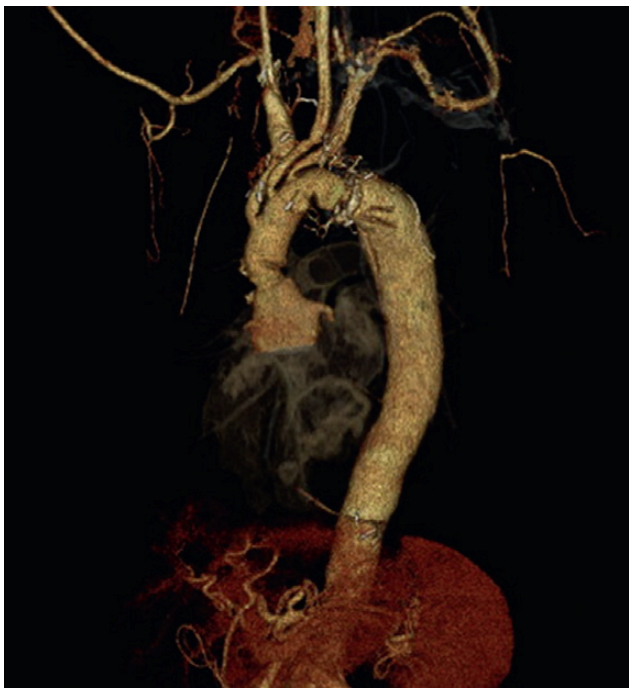

Figure 2: Preoperative CT revealed severe stenosis at the distal anastomotic site and a residual dissecting thoracic descending aortic aneurysm.

technique for acute type A aortic dissection at our institution 5 months previously. Postoperative laboratory data revealed his Hb and LDH levels to be 7.2 g/day and 1598 IU/l, respectively. Although he was discharged on postoperative day 50, he was re-admitted for examination and blood transfusion because of severe worsening anaemia and an LDH level spike 60 days later. Enhanced CT revealed severe stenosis at the distal anastomotic site and mild stenosis at the replaced aortic arch (Fig. 2). Ankle brachial pressure index tests indicated severe lower extremity pressure decrease. A 70-mmHg pressure gradient was noted between the upper and lower extremities by catheterization. We diagnosed haemolytic anaemia caused by distal anastomotic stenosis. During reoperation, we approached the distal aortic arch and descending thoracic aorta using left thoracotomy. We found severe stenosis at the distal anastomotic site and a severely compressed elephant trunk graft. We removed the distal anastomosis and performed descending thoracic aortic replacement. His postoperative course was uneventful. He was discharged on postoperative day 21.

Case 3

A 49-year old man underwent emergency ascending aortic replacement for acute type A aortic dissection at another institution 3 years previously. He was referred to our institution for treatment of residual aortic arch to thoracoabdominal aortic aneurysm.

Preoperative laboratory data revealed his Hb, LDH and haptoglobin levels to be 8.3 g/dl, 892 IU/l and 2 mg/dl, respectively, and a reticulocyte percentage of 37%.

Enhanced CT revealed residual aortic arch and thoracoabdominal aortic enlargement and severe kinking at the graft-to-graft anastomotic site (Fig. 3). During reoperation, we found severe stenosis at the anastomotic site, released the graft kinking and performed total arch replacement and elephant trunk insertion.

Although the patient developed transient neurological dysfunction postoperatively, he had a full recovery. He was discharged on postoperative day 40.

Case 4

A 42-year old man underwent emergency total aortic arch replacement for acute type A aortic dissection at another institution 18 months previously. He was referred to our institution for

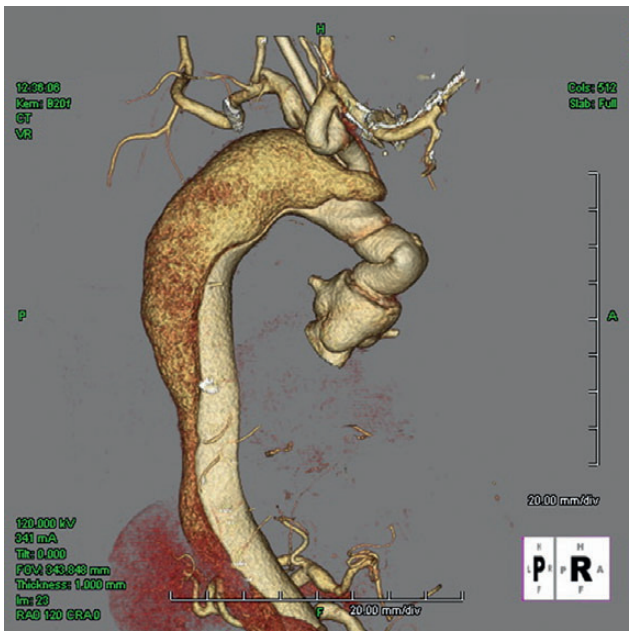

Figure 3: Preoperative CT revealed a severe kinked previous graft and residual dissecting aortic arch and thoracoabdominal aortic aneurysm.

NEGATIVE RESULTS

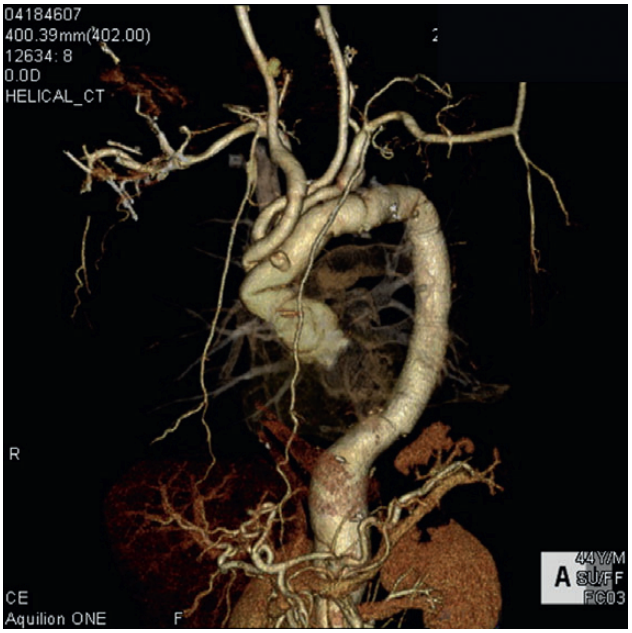

Figure 4: Preoperative CT revealed severe kinked previous graft and replaced descending thoracic aorta by prosthetic graft.

treatment of impending ruptured descending thoracic aorta. The preoperative laboratory data revealed the following: Hb 9.3 g/dl, LDH 299 IU/dl, reticulocyte percentage 29.2% and haptoglobin 158 mg/dl. Enhanced CT revealed a 69-mm dilated descending thoracic aorta and a severely kinked previous graft at the ascending portion. We performed descending thoracic aortic replacement via left thoracotomy at the first reoperation because of impending rupture (Fig. 4).

During the second operation, we found a severely kinked graft caused by inappropriate length tightly wrapped by the residual aortic wall. Ascending aortic replacement released the stenosed portion. His postoperative course was uneventful and he was discharged on postoperative day 20.

DISCUSSION

Although paravalvular leakage after mitral valve surgery is a well-known cause of postoperative haemolytic anaemia, complication with this condition after acute type A aortic dissection surgery is quite rare. A few such cases of haemolytic anaemia have been previously reparted [1–7].

The main causes of haemolysis are inverted internal felt strips [1–5], severely kinked grafts and extravascular factors resulting from graft compression by haematoma or proximal anastomotic pseudoaneurysm [6, 7]. The four causes of haemolytic anaemia in our cases can be divided into two types (Table 1).

One type is anastomotic site stenosis. In particulary, inverted internal felt strips cause severe anastomotic stenosis and accelerated turbulent flow. Teflon felt strips are widely used to reinforce anastomosis of the dissected aortic wall and to prevent residual dissection and bleeding. However, some reports describe related complication. The possibility of infection, distal embolization and haemolytic anaemia should be considered. To prevent these complications, the strip should be narrow and stitches should be placed in the more proximal portion of the felt strip so the proximal part will not turn upwards. In Case 4, we encountered anastomotic stenosis caused by a small native aorta, relatively large elephant trunk graft and inappropriate anastomotic handling.

Table 1: A summary of operative procedures and cause of haemolytic anaemia in 4 patients

Case	First procedure	Cause of haemolysis	Preoperative Hb (g/dl)	Second procedure
1	HAR	inverted internal felt	9.8	TAR
2	TAR + elephant trunk	Distal anastomotic stenosis	7.2	DSR
3	HAR	Graft kinking	8.3	TAR
4	TAR	Graft kinking	9.3	ASR

HAR: hemiarch replacement; TAR: total arch replacement; DSR: descending thoracic aortic replacement; ASR: ascending aortic replacement.

The other haemolytic anaemia type is graft kinking. We experienced two different patterns of kinking. Inappropriate graft length causes graft bending and graft-to-graft anastomotic sites are easily bent because of different flow directions. Graft length should be carefully decided to avoid graft kinking.

The important clues for diagnosis of this rare complication are systolic ejection murmur, laboratory findings that are suggestive of red cell fragmentation syndrome, including progressive anaemia, elevated LDH level, elevated reticulocyte percentage and decreased haptoglobin level and three-dimensional CT findings.

Peripheral blood smears are also useful for differential diagnosis of haemolytic anaemia [6]. In our cases, we could not detect accelerated flow by transthoracic echocardiography, although other reports suggest that echocardiography is more useful than CT. For more precise definitive diagnosis, transoesophageal echocardiography and magnetic resonance imaging should be performed.

In conclusion, haemolytic anaemia caused by an inverted internal Teflon felt or stenosed graft is an extremely rare complication. To aid in early detection of this serious complication, haemolytic anaemia should be considered if progressive anaemia, remarkable LDH level and severely kinked graft on CT scans are detected.

Because the onset period of haemolytic anaemia varies from a few months to a few years, a long and careful follow-up period is needed.

Conflict of interest: none declared.

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