CASE REPORT

Spontaneous intracerebral haemorrhage: a rare complication of aortic aneurysm endoleak

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SUMMARY

We present a rare case of intracerebral haemorrhage secondary to consumptive coagulopathy in relation to ongoing endoleak after thoracic endovascular aneurysm repair (TEVAR). A 68-year-old man underwent elective TEVAR for an 18 cm diameter Crawford type II thoracoabdominal aneurysm. He was subsequently shown to have a type 1b endoleak and a short episode of disseminated intravascular coagulation (DIC) perioperatively. Two months after the procedure, he experienced a consumptive coagulopathy leading to intracerebral haemorrhage and ultimately his death. Endoleak-related DIC is an underappreciated phenomenon within the medical literature. Currently, management is reliant on general DIC principles and anecdotal experiences of others within the case report literature.

BACKGROUND

Endoleaks are a known complication of endovascular repair of aortic aneurysm. It occurs when the aneurysmal sac continues to receive systemic blood flow despite stent graft placement. An infrequent complication of ongoing endoleak is the excessive consumption of platelets and clotting factors within the sac, sometimes resulting in clinically significant coagulopathies.^{1–3} In the current article, we describe the case of a 68-year-old man who suffered a fatal intracerebral haemorrhage as a catastrophic example of an underappreciated phenomenon; endoleak-related consumptive coagulopathy. We then survey current approaches within the literature to the management of this entity and identify the need for further research to recognise high-risk endoleak populations for haemostatic screening.

CASE PRESENTATION

A 68-year-old man with a known Crawford type II thoracoabdominal aneurysm underwent an elective fenestrated endovascular aneurysm repair (EVAR) with T-branch procedure in 2016. His medical background included hypertension, hyperlipidaemia, atrial fibrillation and a previous history of smoking. Full blood count, liver function and coagulation profile were normal apart from a slightly raised activated partial thromboplastin time (APTT) (39s). The procedure was abandoned after stenting of the thoracic aneurysm had been completed due to difficulty cannulating the visceral branches of the abdominal aorta and the excessive procedure length. Digital subtraction angiography confirmed

stent graft placement from just proximal to the left subclavian to the upper abdominal aorta. Images showed evidence of a type I endoleak at the lower pole of the stent. Subsequent CT aortogram showed contrast leaking into an 18 cm diameter aneurysm at least two-thirds of which represented thrombus of various age (see [figures 1 and 2](#)). He was to be followed clinically as an outpatient with a definitive procedure to be scheduled for approximately 3 months later. The patient was transferred to a regional hospital for ongoing recovery and discharge planning on continued dual antiplatelet therapy. Nine days later, he experienced word-finding difficulty. CT head showed he had had a small left-sided intracerebral haemorrhage. Antiplatelet therapy was ceased and he underwent stroke rehabilitation prior to discharge home a short time later with minimal residual symptoms.

Two months following the EVAR procedure, the patient presented to the emergency department with a sudden-onset headache associated with nausea and vomiting. On examination, he was disoriented, had a Glasgow Coma Scale (GCS) score of 14/15 (E4, V4, M6) without localising neurological deficits and a blood pressure of 260/160 mm Hg.

Urgent CT head scan showed a new left-sided occipital intracerebral haemorrhage and small bilateral subdural haematoma. Blood and coagulation abnormalities included thrombocytopenia and prolonged coagulation profile (see [table 1](#)). Blood film was examined showing occasional red cell fragments, decreased haptoglobin, increased bilirubin (and raised lactate dehydrogenase (LDH)) providing evidence of a haemolytic process.

Tranexamic acid (1g) was commenced along with 2 units fresh frozen plasma (FFP), 1 unit (1 adult dose, $>2.4 \times 10^{11}$) platelets and 1 unit of cryoprecipitate (~1g/unit). During the transfer to a tertiary centre, his condition deteriorated with his GCS score rapidly falling to 9/15 (E2, V3, M4) at which stage he was intubated. Repeat CT head showed an enlarging left-sided frontoparietal subdural haematoma (18 mm thick with 13 mm midline shift). The left occipital lobe haemorrhage had enlarged with extensive intraventricular blood, uncal herniation and complete effacement of the basal cisterns (see [figures 3 and 4](#)). Despite administration of a further 3 units of platelets, 4 units of cryoprecipitate, 4 units of FFP and 1 unit of red blood cells, repeat laboratory studies indicated further deterioration in platelets and the coagulation profile (see [table 1](#)).

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Figure 1 Stented thoracoabdominal aortic aneurysm (coronal CT) with evidence of endoleak inferiorly.

OUTCOME AND FOLLOW-UP

Given the radiological findings and refractory coagulopathy, surgery was not appropriate and the decision was made to withdraw treatment. He passed away 36 hours after presentation.

DISCUSSION

The current case illustrates how endoleak may lead to a consumptive coagulopathy causing bleeding abnormalities and, for the first time to our knowledge, fatal intracerebral haemorrhage. Our literature search showed nine previously reported cases of endoleak-related disseminated intravascular coagulation (DIC), all patients were men, had an average age of 69 years and a general medical background including some combination of the typical risk factors for aortic aneurysm (eg, hypertension, dyslipidaemia, current/ex smoker) as well as five patients having preoperative thrombocytopenia and two with liver cirrhosis.¹⁻⁷

Endoleak is diagnosed radiologically as blood flow outside the bounds of the endograft. It is a common complication of endovascular repair of aortic aneurysms, identified in up to 30% of thoracic EVARs.⁸ There are four types of endoleak each indicating the point

at which systemic blood is in communication with the aneurysmal sac.⁹ Of the cases of endoleak-related DIC within the literature, four were type I, two type II and three type III.¹⁻⁷

Type I endoleak makes up 12% of all endoleaks and relate to the ongoing filling of the aneurysmal sac from either the proximal (1a) or distal (1b) stent attachment sites.¹⁰ Due to potential re-pressurisation of the sac, initial management will usually be repeat endovascular intervention to obtain complete closure.^{10 11} Type II endoleaks are the most common (76% of all endoleaks) and denote ongoing perfusion of the aneurysmal sac via retrograde flow from the inferior mesenteric artery, lumbar or intercostal branches.¹⁰ Type II endoleaks rarely cause rupture (approximately 1%) and as many as 60% will spontaneously close within 6 months.¹⁰ For this reason, type II endoleaks are managed conservatively in the first instance. Type III endoleak (3% of all endoleaks) is due to either a junctional leak between endograft components or a hole within the endograft fabric itself.¹⁰ Increasingly rare, type III endoleak significantly increases the risk of aneurysmal rupture and are repaired with deployment of additional stent graft components.¹⁰ Type IV endoleak (3% of all endoleaks) refers to excessive porosity of the

Figure 2 Stented thoracoabdominal aortic aneurysm (axial CT) with evidence of endoleak.

Table 1 Clotting indices before and after blood product replacement

Blood parameter	Platelets	INR	PT	APTT	Fibrinogen
Baseline	152	1.2	–	39	3.3
Presentation	90	2.4	25	35	<0.5
Repeat	50	2.9	30	43	<0.5
Reference range	150–400	0.8–1.2	9–13	22–32	1.8–4.0

APTT, activated partial thromboplastin time. INR, international normalised ratio. PT, prothrombin time.

endograft wall material allowing blood migration across the endograft wall.¹⁰ This type of endoleak is self-limiting in the current generation of endografts, generally resolving within 24 hours.¹⁰

DIC is an acquired syndrome characterised by hyperactivation of the blood coagulation system, resulting in the consumption of platelets, coagulation factors and an increased risk of bleeding.¹² This is reflected by laboratory findings of increased INR and APTT as well as reduced fibrinogen.¹² The International Society on Thrombosis and Haemostasis (ISTH) working group has produced a now widely adopted DIC scoring classification based on these laboratory results.¹³ The ISTH score stratifies patients either into non-overt (low risk) and overt (high risk) DIC groups. Coagulopathy has been reported to occur at a subclinical level in up to 26% of all post-EVAR patients during the perioperative period.¹⁴ A further 2%–4% will show biochemical evidence of overt DIC placing them at significantly increased bleeding risk.^{2,14} Risk factors for overt DIC in the acute phase included duration of the operation, renal failure, age and endoleak.¹⁴ Stimulation of procoagulant activity is thought to occur via endothelial damage by radiological instrumentation and contrast medium, as well as by disturbance of pre-existing intramural thrombus and the direct effect of the stent graft material itself.^{1–3} Long-term, successful endovascular repair leads to a reduction in the prothrombotic state reflected in a shift towards normalisation of platelets and the coagulation profile.¹⁵ In the current case, the patient had normal preoperative platelets and coagulation studies. After EVAR, laboratory findings were consistent with overt DIC, which was treated at the time and, as seen in previous studies, resolved a short time later.¹⁵ However, almost 2 months post-EVAR, overt DIC was again evident. Apart from

endoleak, there was no other identified source to drive the coagulopathy observed (eg, infection, pharmacological causes, liver and nutritional disease or very recent blood transfusion). When endoleak occurs, it is suggested that ongoing perfusion of the aneurysmal sac exposes blood to denuded endothelium and tissue factor.^{1,2} Turbulent blood flow in the sac causes red cell haemolysis and release of procoagulant entities, such as phospholipids, resulting in persistent thrombotic activity.^{1,2} Local consumption of clotting factors causes a systemic deficiency of coagulation precursors, which if severe may lead to significant or even fatal bleeding disorders.⁶

In general, treatment for DIC includes both definitive intervention to address the drivers of coagulopathy, as well as supportive measures aimed at reversing the specific abnormalities within the coagulation system.^{12,13} The acute approach to correction of the haemostatic abnormalities is dependent on the particular type of coagulopathy. Treatment may include replacement of platelets, FFP and cryoprecipitate.^{12,13} Antifibrinolytic agents such as tranexamic acid may also be used where DIC is a predominantly a fibrinolytic bleeding type.¹² For predominantly thrombotic DIC (organ failure type), low molecular weight heparin may be used to terminate ongoing coagulation, thereby preventing further thrombosis, consumption of platelets and coagulation factors.^{12,13} Other authors have shown that coagulopathy can be corrected by closure of the endoleak via stent graft extension, aortic banding and even antifibrinolytic administration.^{1–3}

In the absence of high-quality evidence to guide specific management of endoleak-related consumptive coagulopathy, general treatment principles are to be derived from recognised DIC guidelines and the anecdotal experiences of other endoleak cases within the literature. A study relating post-EVAR endoleak to readily available haemostatic laboratory tests (such as those included in the ISTH score) against the incidence of DIC and clinically adverse outcomes would provide better insight to the risk factors for overt DIC in endoleak. This information could help to identify low-risk and high-risk populations for DIC and guide future screening efforts.

In conclusion, persistent endoleak can lead to overt DIC and catastrophic outcomes as demonstrated here. Currently, there are no widely accepted guidelines to direct DIC screening specific to the endoleak population. In practice, if overt DIC is identified, then coagulopathy should be immediately addressed while endoleak closure is expedited.

Figure 3 Repeat axial CT head scan (axial—level of midbrain).

Figure 4 Repeat CT head scan (axial—level of septum pellucidum).

Learning points

- ▶ Endovascular repair is associated with an acute non-overt disseminated intravascular coagulation (DIC) in 26% of patients and overt DIC in 2%–4%.
- ▶ DIC can develop chronically in relation to ongoing endoleak (particularly with large aneurysms).
- ▶ DIC may be diagnosed and risk stratified by application of the ISTH score.
- ▶ DIC management addresses the underlying cause driving coagulopathy as well as acute replacement of platelets, fibrinogen and other clotting factors where bleeding is present or a major risk.

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