CASE REPORT

Use of the AngioJet percutaneous thrombectomy system for the treatment of acute Budd-Chiari syndrome

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SUMMARY

A 31-year-old woman presented to our emergency department with an acute liver injury secondary to acute Budd-Chiari (BC) syndrome from hepatic vein thrombosis. After a thorough discussion of the risks involved, we proceeded to treatment with a novel approach, performing a mechanical hepatic vein thrombectomy with the AngioJet percutaneous thrombectomy system. Restoration of hepatic vein flow was confirmed with on-table Doppler ultrasound. There were no complications following the procedure. The patient was initiated on anticoagulation, and showed progressive clinical and laboratory improvement. She was discharged home on day 20 with normal liver function and biochemistry. This is the first reported case of successful mechanical thrombectomy in acute BC syndrome without the addition of angioplasty or chemical thrombolysis.

BACKGROUND

Budd-Chiari (BC) syndrome is a rare cause of hepatic injury, because of occlusion of the hepatic veins or inferior vena cava (IVC), leading to hepatic congestion. It is characterised by pain, nausea, anorexia, jaundice and deranged liver biochemistry. In some cases, the condition can lead to fulminant liver failure and requirement for transplantation. Anticoagulation is an effective treatment for this disorder; yet is associated with an increased risk of portal hypertensive haemorrhage, particularly in patients with severe hepatic synthetic dysfunction; the risk quantified is between 17 and 34% of anticoagulated patients in various retrospective analyses.^{1–3}

Insertion of a transjugular intrahepatic portosystemic shunt (TIPS) has also shown benefit in acute BC syndrome, yet risks severe complications including hepatic encephalopathy, pulmonary oedema, intrahepatic or intraperitoneal haemorrhage, TIPS thrombosis and IVC injury.^{2–6}

We describe a case of acute BC syndrome because of hepatic vein thrombosis from a systemic thrombophilia, which was successfully treated with a novel approach using radiologically guided percutaneous mechanical thrombectomy.

CASE PRESENTATION

A 31-year-old Caucasian female with quiescent Crohn's disease, diagnosed 6 months prior to presentation, was admitted to hospital with a 1 month history of intractable headache and neck stiffness,

culminating in a generalised tonic-clonic seizure. Her only medication was prednisolone 20 mg daily. She had no personal or family history of thromboembolic disease, and denied using any hormonal contraception.

On examination, the patient was alert and orientated with no discernible postictal phase or encephalopathy. Peripheral nerve examination identified significant left upper limb weakness with hyperreflexia. Sensation to pinprick and vibration was intact in all limbs, and cranial nerve examination was normal. Abdominal examination revealed significant distension with shifting dullness. There was no jaundice or asterixis. There were no peripheral stigmata of chronic liver disease.

INVESTIGATIONS

Laboratory investigation demonstrated markedly deranged liver function tests, with an alanine transaminase (ALT) of 752 IU/l, aspartate transaminase (AST) of 439 IU/l, alkaline phosphatase (AP) of 114 IU/l, γ -glutamyl transpeptidase (GGT) of 66 IU/l and total bilirubin of 29 μ mol/l. The prothrombin time ratio (international normalised ratio, INR) was recorded at 2.1. An urgent CT image of the brain showed extensive superior sagittal sinus and bilateral cortical vein thrombosis, with haemorrhagic venous infarcts in the right frontal and left parietal lobes. A triphasic abdominal CT scan demonstrated concurrent hepatic vein and right posterior segment portal vein thrombosis with moderate ascites. There was no evidence of malignancy, nor extrinsic hepatic or IVC compression or stenosis. Hepatic parenchymal enhancement during the portal venous phase was consistent with acute BC syndrome. There was no other evidence of haemorrhage despite the elevated INR.

TREATMENT

The patient was started on a heparin infusion with a target APTT of 50–60 s, measured every 4 h. Concerns nevertheless remained regarding the safety of anticoagulation in an already coagulopathic patient with deteriorating liver function and possible portal hypertension.

Consultation with the hospital's radiology department yielded a proposition of catheter-mediated extraction of the hepatic vein thrombi, in an attempt to ameliorate the patient's acute liver injury and more reliably anticoagulate the patient. The procedure was undertaken with the patient's consent after a thorough discussion of the risks

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Figure 1 Middle hepatic vein with large central thrombus before percutaneous thrombectomy.

involved, and an understanding that we would proceed to TIPS insertion if her liver function continued to deteriorate.

A 10-French catheter was inserted via the transjugular route into the patient's hepatic veins under fluoroscopic guidance (figure 1). The AngioJet percutaneous thrombectomy system (MEDRAD Inc, Pennsylvania, USA), currently licensed for use in peripheral arterial, deep venous and native coronary/bypass graft thrombectomy, was inserted into the hepatic veins using the Expedior catheter (Possis Medical Inc, Minnesota, USA) (4–12 mm vessel size). The device was used to successfully fragment and aspirate thrombi residing within the right and middle hepatic veins. Fluoroscopic evidence of restored flow was achieved for all treated vessels (figure 2). Patency was confirmed with on-table Doppler ultrasound postprocedure.

OUTCOME AND FOLLOW-UP

There was a steady improvement in the patient's liver biochemistry and markers of synthetic function (see table 1). A therapeutic abdominal paracentesis was performed, and spironolactone 100 mg daily and frusemide 40 mg daily were initiated. Ascitic fluid biochemistry was consistent with portal

Figure 2 Middle hepatic vein after percutaneous thrombectomy.

Table 1 Progression of liver biochemistry and international normalised ratio following treatment

	ALT	AST	INR	Bili
Day 1	752	439	2.1	29
Day 2	685	288	1.9	29
Day 7	409	137	1.7	17
Day 11	184	65	1.6	16
Day 21	65	30	2.3*	12
Day 50	34	24	3.3*	12

*Values measured after initiation of warfarin.

ALT, alanine transaminase (IU/ml); AST, aspartate transaminase (IU/ml); Bili, total bilirubin ($\mu\text{mol/l}$); INR, international normalised ratio.

hypertensive transudate based on a serum-ascites albumin gradient >11 g/l. The patient did not reaccumulate ascites, and her diuretics were progressively weaned until cessation 14 days later. There was no hepatic encephalopathy during her admission. The patient's left upper limb weakness progressively improved and no further seizures occurred.

The patient was discharged home on day 20 of her admission. She was reviewed again as an outpatient 16 weeks later, with complete normalisation of liver biochemistry, and no clinical evidence of encephalopathy or ascites. A follow-up ultrasound scan of the liver showed normal flow within all branches of the hepatic and portal veins. No aetiology of the patient's thrombophilia was identified, despite testing for pregnancy, protein C/S deficiency, antithrombin III deficiency, Factor V Leiden, antiphospholipid antibody syndrome, prothrombin gene mutation, hyperhomocysteinaemia, paroxysmal nocturnal haemoglobinuria and JAK2 V617F gene mutation. The only risk factor found for the extensive thrombosis and thrombophilia was Crohn's disease. She remains on lifelong warfarin (target INR 2.0–3.0) given the significant sequelae of her thrombosis.

DISCUSSION

This is the first report of the use of the AngioJet percutaneous thrombectomy system for acute BC syndrome without the addition of angioplasty or chemical thrombolysis. Thrombectomy has been used successfully in only a small number of reported BC cases to date.

One case of acute BC because of IVC stenosis and thrombosis was successfully treated using the Arrow percutaneous thrombectomy system (Arrow International, Pennsylvania, USA), followed by immediate angioplasty to the IVC.⁷ Recurrence of thrombotic IVC occlusion 9 months later led to a repeat percutaneous thrombectomy using the same device, followed by stenting of the IVC stenosis. The patient remained well on follow-up 12 months later.

Three additional cases of acute BC, each owing to paroxysmal nocturnal haemoglobinuria (PNH), were treated with a combination of catheter-mediated thrombolysis and thrombectomy.⁸ Thrombectomy in each case was performed using the AngioJet system, and in two cases was immediately followed by the Helix Clot Buster (Microvena, Minnesota, USA) system. Urokinase was administered directly into the hepatic veins following clot removal. Each case experienced progressive improvement in liver biochemistry. Two patients remained well at 3 months; however, one patient had passed away from unrelated comorbid illness.

Current American Association for the Study of Liver Diseases guidelines recommend the use of a tiered approach to treatment,

involving anticoagulation as first-line therapy, with angioplasty to stenotic vessels should they be present.⁹ If deterioration continues, then consideration of TIPS is recommended. Liver transplantation may ultimately be necessary in patients that fail to improve. The potential risks and limitations of these treatments were highlighted in this case. Although no randomised controlled trials have been performed on anticoagulation in acute BC, significant rates of anticoagulation-related haemorrhage have been reported in retrospective studies.^{1–3} There are also documented cases of severe complications from TIPS in this setting, most pertinently hepatic encephalopathy, bleeding and vascular injury. Several procedure-related deaths have also been reported.^{2–4} Hepatic vein clearance with percutaneous thrombectomy may offer another potential therapy alongside these other well-established treatments.

An unfortunate limitation of this case is that we cannot comment upon the relative contributions of anticoagulation and hepatic vein thrombectomy towards this patient's recovery. Further studies will be needed to determine if this treatment holds value in routine implementation alongside anticoagulation in the treatment of acute BC syndrome.

Learning points

- ▶ Acute Budd-Chiari (BC) syndrome has the potential to cause severe liver injury/failure.
- ▶ Anticoagulation is the recommended first-line treatment for acute BC syndrome.
- ▶ Consideration of transjugular intrahepatic porto-systemic shunt (TIPS) is recommended in patients who continue to deteriorate despite anticoagulation.
- ▶ Mechanical thrombectomy is a novel approach to the treatment of acute BC syndrome that may warrant consideration in the deteriorating or high-risk patient, where TIPS may be contraindicated or considered overly aggressive.

Contributors AD completed the initial draft. AN and RD contributed to revisions of the manuscript. All authors contributed to the management of the patient described in this case report.

Competing interests None.

Patient consent Obtained.

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