Recurrent Portal Vein Thrombosis after Liver Transplantation for Budd-Chiari Syndrome

Miriam Cortes Cerisuelo MD PhD
Consultant Liver Transplant Surgeon
Institute of Liver Studies
King’s College Hospital
London
Case report

-29 years-old male
-Ulcerative colitis (poor response to steroids) in 2006
-Budd-Chiari syndrome (ascites) in 2007
Abdominal CT:

- Heterogeneous big liver
- Partial occlusion of the hepatic veins
Full prothrombotic screening:

- Negative for JAK-2 mutation
- Bone marrow normal
- Antiphospholipid negative, Protein S normal.
- Protein C deficiency

Myelodysplastic syndrome

Anticoagulated with Warfarin
Cerebral venous thrombosis in 2008 and PE

Cerebral CT venogram:

Venous thrombosis in the sagittal sinus
- Initial improvement after TIPS in 2009
- Initial improvement after TIPS in 2009

- Progression of the symptoms:
  Ascites, abdominal pain, diarrhea, lethargy

- Worsening liver function:
  Bb 300 µmol/L
  AST 1835 IU/L
  Albumin 25-35 g/L
Abdominal CT: ascites, heterogeneous liver, hypertrophy of the caudate lobe, thrombus in the TIPS and retro hepatic cava
- First liver transplant on March 2009
DBD whole graft 67 years-old
Venous-venous bypass
Caval replacement
Duct to duct
- Not fully compliant with Warfarin or Clexane
- Recurrent Budd-Chiari syndrome in 2013
- Worsening ascites, renal and synthetic dysfunction.

Warfarin/Clexane switched to Rivaroxaban
- Second LT on September 2014

DBD 54 years-old

Whole graft

Piggy-back 3 veins

Duct to duct
Hepatic vein thrombosis

*Courtesy of Dr Alberto Quaglia, Consultant Histopathologist. Institute of Liver Studies*
One month later, presented with tonic-clonic seizures.
Head CT

Intracranial fungal abscess

Voriconazol 6 months
-After stopping the antifungal, the dose of Tacrolimus was not increased resulting in severe ductopenic rejection

-Bilirubin 426 umol/L and no response to medical treatment
-Third re-transplantation 4 months later: January 2015
DBD whole graft 68 years-old
Venous-venous bypass
Caval replacement
Jump graft to restore portal flow from SMV to donor PV
Duct to duct
Skin closure only
-Third re-transplantation 4 months later: January 2015

Jump graft between the SMV and donor portal vein with iliac vein from a deceased donor.
Fourth abdominal surgery:

Biliary reconstruction with a hepatico-jejunostomy for anastomotic stricture for failed endoscopic treatment on October 2015
+ full muscle closure
One year later presented with abdominal pain, renal dysfunction and ascites.

- CT showed narrowing of the venous jump graft
- PTLD? Lymph node compressing the jump graft
- Dilatation declined by radiologist “High risk”
Few months later re-admitted in hospital with:

- Gastro-intestinal bleeding
- Persistent abdominal pain, distension
- Blood transfusion requirements.
Abdominal CT

Complete thrombosis of the previous jump graft
Fifth laparotomy for meso-Rex shunt on November 2016

Findings:
Multiple varices and moderate portal hypertension
1.6L ascites

Complex surgery:
7 L blood loss
Pericardium opened during the surgery
Fifth laparotomy for meso-Rex shunt on November 2016
Fifth laparotomy for meso-Rex shunt on November 2016
Fifth laparotomy for meso-Rex shunt on November 2016

Mean velocity: 26.8 cm/s
Fifth laparotomy for meso-Rex shunt on November 2016

Pulp pressure
Before: 40 mmHg
After: 23 mmHg

Pressure in the SMV
Before: 23 mmHg
After: 5 mmHg
Complications:

- re-laparotomy for bleeding 2 days later.
- Pericardiocentesis by cardiologist 25/11/2016, 900mls aspirated
- Chyle leak
- CMV viremia
- Pulmonary emboli
Fibrous septum

Hepatic plate disarray

Courtesy of Dr Alberto Quaglia, Consultant Histopathologist, Institute of Liver Studies
Discharged home 6 weeks later but...
-After 8 months, admitted in his local hospital with:
  abdominal pain and distension

-Abdominal CT

Thrombosis of the new jump graft
- Normal LFT:

**Bilirubin**: 3 µmol/L

**ALP**: 197 IU/L

**AST**: 27 IU/L

**GGT**: 87 IU/L
Abdominal CT:

Thrombosis of the new jump graft

Several non-occlusive thrombi

within the portal vein branches
What to do next?
What to do next?

- Wait and see...
- Refashioning of the Rex shunt + tying all possible spontaneous shunts?
- Liver re-transplantation + tying all possible shunts (left gastric...)
- Combined liver and small bowel transplant+/- colon
  +/− colectomy
What to do next?

Why is he still procoagulant?
Why is he still procoagulant despite Liver transplantation?

-Protein C polymorphism in the donor liver: development of antibodies by the recipient

-Is Rivaroxaban enough?

-Thrombotic tendency related to UC +/- episodes of dehydration?

-Donor acquired pro-thrombotic status
Thrombotic tendency in UC

Table 1: Acquired risks factors for thrombosis in IBD

1. Fluid depletion
2. Surgery
3. Central venous catheters
4. Immobilization
5. Steroid therapy
6. Oral contractive/hormone replacement therapy
7. Vitamine defiency
8. Hyperomocystenemia
9. Cigarette smoking

Table 2: Abnormalities in coagulation, anticoagulation and fibrinolytic system in IBD

<table>
<thead>
<tr>
<th>Coagulation factors</th>
<th>Fibrinolytic factors</th>
<th>Plasma coagulation inhibitors</th>
</tr>
</thead>
<tbody>
<tr>
<td>↑ Fibrinogen</td>
<td>↓ tPA</td>
<td>↓ AT III</td>
</tr>
<tr>
<td>↑ Prothrombin</td>
<td>↑ PAI-1</td>
<td>↓ TFPI</td>
</tr>
<tr>
<td>↑ Factors: Va, Vlla, Vllla, Xa,Xla, XIIa</td>
<td>↑ TAFI</td>
<td>Conflicting data about PS and PC</td>
</tr>
<tr>
<td>↑ Prothrombin factors 1+2</td>
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<tr>
<td>↑ Thrombin-antithrombin III complex (TAT)</td>
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<td></td>
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<tr>
<td>↑ Fibrinopeptide A and B</td>
<td></td>
<td></td>
</tr>
<tr>
<td>↑ Microparticles</td>
<td></td>
<td></td>
</tr>
<tr>
<td>↓ Factor XIII</td>
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</tbody>
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*Gianotta et al. Thrombosis Journal 2015*
Thank you