Veno-occlusion disease after liver transplantation: Case Report

Jae Ryong Shim, Je-ho Ryu, Kwang-ho Yang, Byung-hyun Choi, Tae-beom Lee
Department of General Surgery, Pusan National University Yangsan Hospital, Korea, Republic of

Introduction

- Venous occlusive disease (VOD) as known as sinusoidal obstruction syndrome (SOS) is extremely rare disease that can occur within 30 days after hematopoietic stem cell transplantation (HSCT).

- VOD is relatively well-known in other countries where bone marrow transplantation has become a universal treatment for hematologic malignancy, but VOD after liver transplantation (LT) is more extremely rare. VOD can be caused by liver disease before HSCT, radiation-induced liver disease, massive pretreatment before LT, usage of azathioprine.

- Some researchers revealed that VOD after LT can be diagnosed by hepatomegaly, right upper quadrant pain, jaundice, and ascites. Several studies revealed that heparin, tissue plasminogen activator (t-PA), ursodeoxycholic acid, prostaglandin E1, pentoxifylline is not helpful for prophylaxis of VOD. And recently, some studies showed that defibrotide can be used as treatment of VOD after LT.

- This case is about the patient who was diagnosed as VOD after LT, and the patient was treated with defibrotide.

Case Presentation

- Forty years old female patient was diagnosed as primary biliary cirrhosis (PBC). PBC was managed with several medications including azathioprine. Liver transplantation was considered as her condition was getting worse. And she was taking azathioprine just before operation.

- Her sister donated her liver and living donor liver transplantation was performed.

- Clinical acute rejection was considered as liver enzyme was elevated after operation. Steroid pulse therapy was applied and rejection was considered as treated.

- Transjugular liver biopsy was performed due to recurrent RUQ pain, jaundice and elevated liver enzyme. The patient was transferred to ICU after biopsy for management of fluid retention.

- Liver biopsy was confirmed as veno-occlusion disease (VOD). And conservative management was applied such as low-molecular weighted heparin, PEG E1, diuretics. And percutaneous drainage was applied for pleural effusion and ascites control.

- But jaundice and elevated liver enzyme were not satisfactorily improved, and a condition of the patient was getting worse.

- And next, we searched published studies for treatment of VOD after transplantation. Several studies showed that defibrotide was considered as effective treatment for VOD. We administered defibrotide to the patient with reference to the studies, and the patient was slowly recovered from jaundice and elevated liver enzyme was also slowly normalized.

Figure 1. After LDLT, acute rejection was suspicious due to elevated AST, ALT and TB.

Figure 2. After administration of defibrotide, total bilirubin was recovered slowly.

Discussion

- In the postliver-transplant setting, VOD is considered to be a rare complication. There have been a few publications describing this condition in detail or its prevalence after liver transplantation. In a previous study on liver transplant recipients, VOD occurred in approximately 2% of a large study population.

- Pathologic findings of liver VOD (or SOS) are subintimal edema and hemorrhage of the terminal hepatic venule, centrilobular congestion, hepatocyte degeneration in an early stage. Late lesions include complete or partial fibrous obstruction of the distal hepatic vein and sinusoid fibrosis of the central lobule.

- This disease was initially called VOD because of the occlusive fibrosis of the small hepatic vein. However, recently, the sinusoidal lesion was found to be the main lesion and is called SOS.

- The frequency of SOS varies from report to report but varies from 0 to 60% and the average incidence is 14%. More than 90% of severe VOD cases are known to die from multi-organ failure (MOF).

<table>
<thead>
<tr>
<th>Criteria</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Seattle</td>
<td>At least two of the following within 30 days after transplantation - Hepatomegaly and right upper quadrant pain - Jaundice - Unexplained weight gain or ascites</td>
</tr>
<tr>
<td>Modified</td>
<td>At least two of the following within 20 days after transplantation - Hepatomegaly and right upper quadrant pain - Unexplained weight gain of more than 2% or ascites</td>
</tr>
<tr>
<td>Baltimore</td>
<td>Bilirubin greater than or equal to 2 within 21 days after transplant and at least two of the following - Painful hepatomegaly - Ascites - Weight gain of more than 5% of baseline</td>
</tr>
</tbody>
</table>

Table 1. Defining criteria for sinusoidal obstruction syndrome