Annals of Hepatology 29 (2024) 101500

Contents lists available at ScienceDirect

Annals of Hepatology

journal homepage: www.elsevier.es/annalsofhepatology

Opinions

Liver rupture in patients with amyloidosis: Clinical features and treatment

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ARTICLE INFO

Article History:
Received 31 January 2024
Accepted 19 February 2024
Available online 4 April 2024

Hepatic amyloidosis is a rare disease caused by the extracellular deposition of various amyloid fibrils, such as immunoglobulin light chain (AL), serum amyloid A (AA), apolipoprotein A1 (ApoA1), and transthyretin (TTR) type of fibrils, in the liver tissue, which disrupts tissue architecture and leads to organ dysfunction [1,2]. In patients with hepatic amyloidosis, amyloid is deposited within the space of Disse along the hepatic sinusoids, within the walls of hepatic blood vessels, or in the walls of the extrahepatic and intrahepatic large bile duct [2,3]. The most common clinical manifestations of hepatic amyloidosis were fatigue, weakness, weight loss, and hepatomegaly, and the median survival of these patients was 8.5 months [1]. Although rare, hepatic amyloidosis can present with potentially fatal complications, such as liver failure and hepatic rupture [4,5].

Hepatic rupture in amyloidosis patients can be spontaneous or induced by trauma or liver biopsy [6–8]. The clinical manifestations were abdominal pain, abdominal distension, fatigue, emesis and weight loss [6,9,10]. Hepatic rupture in amyloidosis was displayed as perihepatic hematoma, parenchymal bleeding and abdominal effusion by CT or ultrasonic examination [6,11]. The pathological types of liver biopsy were AL amyloidosis, lysozyme amyloidosis, AA amyloidosis and TTR amyloidosis [12–15]. Lysozyme amyloidosis is an exceedingly rare hereditary amyloidosis [16], but this type of amyloidosis is prone to spontaneous hepatic rupture [5,14,17]. Several mechanisms may have been incriminated in the spontaneous rupture of the liver with amyloidosis, such as hepatic parenchyma infiltration with amyloid, vascular fragility from amyloid involvement, and defects in blood coagulation [5,14].

Given the rarity of hepatic rupture with amyloidosis, there is no current standardized treatment. The clinical treatment of hepatic amyloidosis with rupture can be classified into four aspects. Firstly, supportive care, such as blood transfusions, is mandatory for patients with serious hemorrhage. However, some patients did not respond to transfusion and needed further treatment to control the bleeding [6,7]. Conservative therapy without hemostatic therapy is not likely to be effective in hepatic rupture patients [18]. Secondly, controlling liver bleeding is a critical part of treating hepatic rupture with amyloidosis. Currently, angiographic embolization and surgery remain the significant treatments of choice for the control of liver bleeding. Amyloidosis patients with hepatic rupture successfully achieve hemostasis by using hepatic artery embolization [6,11,12], suggesting angiographic embolization is feasible and effective for the treatment of hepatic rupture with amyloidosis patients. Except for angiographic embolization treatment, surgery treatment, including abdominal exploration for hemostasis, liver lobectomy and hepatic artery ligation, was often used to control the hemorrhage [7,12]. However, patients fail to hemostasis through surgery treatment, which may be attributed to liver fragility and difficulty in hemostasis [6,9,11,14,15].

Moreover, percutaneous microwave ablation was also used to achieve liver hemostasis when angiographic embolization or surgery was difficult [8]. Thirdly, etiological treatment, which aimed to reduce the supply of amyloid fibrils, resulted in improved mean survival [19,20]. Targeting the primary disease may improve the amyloidosis of patients whose hepatic amyloidosis is attributable to an underlying condition. For example, in patients with a monoclonal plasma cell proliferative disorder, treatment of their monoclonal plasma cell proliferative disorder with chemotherapeutics and autologous stem cell transplant resulted in a hematological and organ response [7]. Lastly, liver transplantation is suitable for liver rupture in amyloidosis, which is massive parenchymal disruption and surgically untreatable or complication of liver failure. Combination of supportive measures, surgical treatment, angiographic embolization, liver transplantation with etiological treatment was used to decreasing the mortality rate in hepatic rupture with amyloidosis. However, most of the patients had serious bleeding with difficult to control hemorrhage.

Hepatic rupture in the setting of amyloidosis is a rare and life-threatening disorder, which is often a spontaneous rupture of the liver without abdominal trauma. In many cases, patients present with abdominal pain and abdominal distension. CT and ultrasonic findings helped detect hepatic rupture. Although a standardized treatment has not been established, supportive therapy, angiographic embolization and surgery, etiological treatment, and liver transplantation contribute to the survival of patients. However, most patients had serious bleeding and were particularly susceptible to poor outcomes.

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https://doi.org/10.1016/j.aohep.2024.101500

Declaration of competing interest

None.

Funding

Wei Wang is supported by grants from the Natural Science Foundation Project of Chongqing (cstc2020jcyj-msxmX0145).

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