




Case Report

Lymphedema in a patient with decompensated cirrhosis

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Abstract:

The lymphatic system is critical in body fluid homeostasis. Although lymphatic vascular expansion prevents the development of ascites and oedema in the early stages of liver cirrhosis, compensatory mechanisms cannot achieve this in the advanced stages due to lymphatic dysfunction. A 36-year-old male patient, who had been followed up for cryptogenic cirrhosis for 15 years, had been complaining of excessive swelling in his legs for the last 4 years. Excessive swelling in the legs was accompanied by skin rashes and ulcers, and the leg skin had an orange peel appearance characteristic of lymphedema. Bilateral lower extremity artery and venous system colour Doppler ultrasonography showed that the vascular structures of the right lower extremity arterial and venous system were open, and the flow rate, direction and spectrum were normal. The patient, who could not undergo liver transplantation due to organ limitations, died due to sepsis following lymphedema-induced wound infection. In conclusion, this case suggests that lymphedema should be considered in the presence of oedema in cases of decompensated cirrhosis, and the necessary conservative treatments should be applied.

Keywords: cirrhosis, lymphedema, lymphatic dysfunction, lymphatic system, oedema

Introduction

Ascites and hepatic oedema are major complications observed in patients with decompensated cirrhosis and are associated with increased morbidity (1) and poor quality of life (2). The lymphatic vascular system plays a critical role in ascite formation. The lymphatic vascular system removes interstitial fluid from tissues in the body and returns it to the bloodstream. Failure of normal lymphatic function results in a build-up of interstitial fluid and can lead to lymphedema and ascites (3).

The liver is the largest lymph-producing organ, generating 25% to 50% of the lymph flowing through the thoracic duct. (4) Recent studies have suggested that the progression of liver disease causes structural and functional changes in both hepatic and extrahepatic lymphatic vessels. This suggests a possible link between lymphatic vessel changes and disease progression (5-6).

In this case report, we aim to present the lymphedema we encountered in a patient with liver cirrhosis.

Case Report

A 36-year-old male patient, who had been followed up for cryptogenic cirrhosis for 15 years, had been complaining of excessive swelling in his legs for the last 4 years. Excessive swelling in the legs was accompanied by skin rash and ulcers (*Figure 1*). His sister underwent a living donor liver transplantation due to cryptogenic liver cirrhosis. The patient had been

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hospitalized twice before with the same complaint and had undergone debridement surgery at the plastic surgery department due to the wounds on his legs. In the last abdominal ultrasound examination, it was determined that the liver had a heterogeneous parenchymal echo, its contours were lobulated (chronic liver disease), the size of the spleen increased (16 cm), and there were massive ascites. Bilateral lower extremity artery and venous system colour Doppler ultrasonography showed that the vascular structures of the right lower extremity arterial and venous system were open, and the flow rate, direction and spectrum were normal. In the examination area, the subcutaneous tissue thickness is increased and edematous in the bilateral thigh and leg sections. *Figure 2* shows cirrhotic liver and massive ascites on dynamic liver tomography.

His laboratory values were creatinine 1.4 mg/dL, total bilirubin 11.95 mg/dL, direct bilirubin 7.19 mg/dL, prothrombin time INR 1.45, NA(Sodium):131 mmol/L, Albumine 1.4 G/DL, Cholesterol:52 mg/dL, Tg:96 mg/dL, VLDL 19.2. HBsAg and Anti-HCV were negative. MELD-Na and Child-Pugh scores 26 and 11, respectively.

Liver transplantation was planned due to decompensated cirrhosis. However, cadaveric or living donor liver transplantation could not be performed due to organ limitations. Cultures taken from the wounds on his legs grew *Klebsiella pneumoniae* and *Pseudomonas aeruginosa* bacteria. The patient did not respond to antibiotic treatments in the intensive care unit and died due to sepsis.



Figure 4 Excessive swelling in the legs was accompanied by skin rash and ulcers.



Figure 5 Dynamic Liver tomography shows cirrhotic liver and massive ascites.



Figure 6 Typical lymphedema appearance on the left leg.

Discussion

Ascites and oedema are common in patients with decompensated cirrhosis. Since lymphedema is rare, it is generally considered as classical oedema in this patient group at first and is not brought to mind. In addition, lymphedema is generally considered an untreatable and resistant chronic problem. All of these may lead to diagnostic delays. To diagnose lymphedema, attention should be paid to the patient's risk factors and specific findings on physical examination (7). Peripheral lymphedema manifests itself physically as an orange peel appearance on the skin, as seen in *Figure 3*.

The lymphatic system is critical in body fluid homeostasis, adaptive immunity, and transport of lipids. In patients with liver cirrhosis, capillary filtration increases markedly due to an increase in hydrostatic pressure, resulting in increased lymph production. In the early stages, expansion of the lymphatic vascular system attempts to prevent fluid accumulation by returning fluid to the systemic circulation. However, with the progression of cirrhosis, lymphatic functions become compromised and, as a result, the lymphatic compensatory mechanism becomes overloaded, contributing to the development and eventual worsening of ascites and oedema (8). Additionally, there are studies reporting that the progression of liver disease is accompanied by changes in hepatic and extrahepatic lymphatic vessels (5-6). Although there are very few publications on lymphedema in patients with cirrhosis, the frequency of peripheral lymphedema in patients with cirrhosis and refractory ascites was reported as 33.5% in one study (9). The paucity of literature on lymphedema in patients with cirrhosis is probably due to the fact that patients are considered to have classical oedema and are less noticed, as well as the difficulties in the diagnosis and treatment of lymphedema.

In the treatment of lymphedema, intensive bandaging and lymphatic massage are essential, as well as the use of compression clothing. Surgical interventions may be required in cases that do not respond to conservative treatments (7). Our case had to undergo surgery twice due to deep ulcers. Patients with lymphedema are more prone to cellulite due to their deep fissures, stagnant lymph and weak immunity (9). Our case with decompensated cirrhosis also died due to sepsis following lymphedema-induced wound infection.

In conclusion, this case suggests that lymphedema should be considered in the presence of oedema in cases of decompensated cirrhosis, and the necessary conservative treatments should be applied.

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