Images in Pediatric and Congenital Heart Disease

Duodenal Lymphangiectasia in a Patient Suffering from Protein-Losing Enteropathy after Undergoing Fontan Operation

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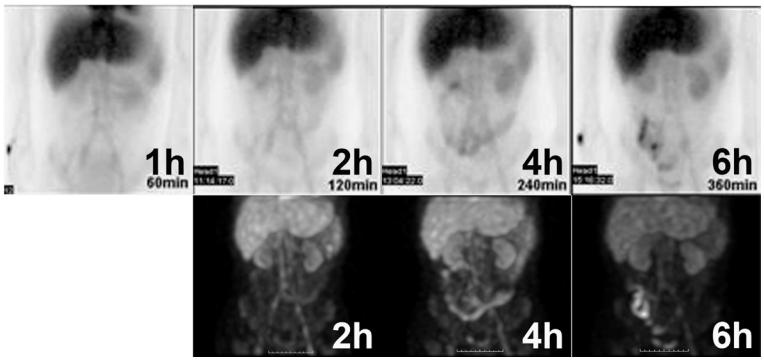

Fig. 1 Albumin scintigraphy
Albumin scintigraphy revealed albumin loss from the duodenum and jejunum even in early phase.

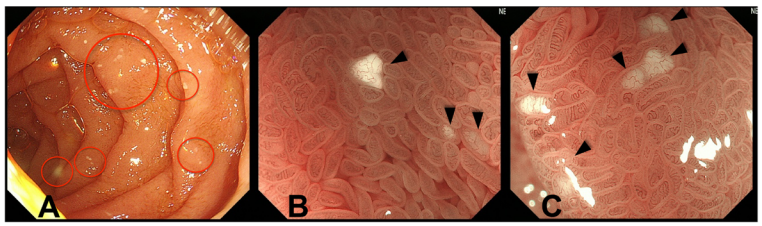

Fig. 2 Endoscopic images of the duodenum
(A) Macro-image of the intestinal mucosa. Intestinal mucosa and duodenal villi were edematous. White spots on the surface were lymphedema (inside the circles). (B) and (C) Magnified images of duodenal villi. White spots on the villi (arrowheads) indicate enlarged lymphatic vessels.

The Fontan operation is a radical surgical procedure that is used to treat congenital heart disease patients with a single functional cardiac ventricle. Recently, patients who undergo the Fontan operation have a good prognosis. However, several severe complications have been reported during long-term follow-up of

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these patients, particularly those that have undergone atriopulmonary connection (APC). Among these, protein-losing enteropathy (PLE) is one of the complications which is characterized by abnormal loss of protein from the intestinal tract and results in hypoalbuminemia, hypogammaglobulinemia, tetany, edema, ascites, pleural effusion, and diarrhea.[1] Although abnormal lymphatic circulation and focal inflammation due to high venous pressure is thought to be one of the causes of PLE as well as intestinal edema, its pathophysiology is still unclear.[2] Using endoscopy, we observed lymphangiectasia and lymphatic congestion in duodenum of a patient who was suffering from PLE after undergoing the APC Fontan operation.

A 34-year-old female complained of diarrhea possibly due to PLE, as her mean right atrial pressure was 19 mmHg. She was born with tricuspid atresia and pulmonary atresia, and underwent the APC Fontan operation when she was 9 years old. She had suffered from severe hypo-albuminemia since she was 20 years old, and had a serum albumin level of approximately 1.2–1.4 mg/dL. Albumin scintigraphy revealed intestinal albumin loss including descending portion of duodenum (Fig. 1). Gastroduodenal endoscopy was performed to elucidate the causes of the albumin loss, during which edematous intestinal mucosa and duodenal villi were observed. Many villi were white, which was presumably related to enlargement of the lymphatic vessels (Fig. 2 A–C).

Although the causes of PLE remain still uncertain, these findings indicate that one cause of PLE is increased lymphatic pressure due to Fontan circulation. This is the first clear evidence image indicating that lymphatic congestion after Fontan operation can cause PLE. To improve PLE, total cavopulmonary connection conversion procedure is planning to reduce high venous and lymphatic vessels pressure in the near future.

Conflict of Interest

None.

References

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