Case Report

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Splenic Duplication, a Rare Cause of Gastric Varices: A Case Report

비장 중복, 위 정맥류의 드문 원인: 증례 보고

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Splenic duplication, also known as polysplenia syndrome, is a condition occasionally observed in which the spleen is divided into segments of similar size. However, gastric fundic varices arising from a duplicated spleen are exceedingly rare, and this medical anomaly has been infrequently reported in the literature. This case report presents a 40-year-old male with a rare instance of gastric fundic varices secondary to splenic duplication. Comprehensive imaging studies, including endoscopy, CT, Doppler US, and radioisotope splenic scans, were performed to confirm the diagnosis. This case contributes to valuable information in the medical literature, shedding light on a seldom-discussed condition.

Index terms Splenic; Duplication; Varices; Sinistral Portal Hypertension

INTRODUCTION

Splenic duplication, also known as polysplenia syndrome, is a rare condition characterized by the division of splenic tissue into two or more segments of similar size (1, 2). While splenosis-causing gastric varices have been described, gastric varices arising from congenitally duplicated spleens are exceedingly rare, with few reported cases in the literature (2, 3).

This report presents a rare case of splenic duplication leading to a gastric varix that was incidentally discovered during a routine health screening. The diagnosis was confirmed by endoscopy, CT, Doppler US, and radioisotope splenic scans.

CASE REPORT

A 40-year-old asymptomatic male with no prior medical history, including viral hepatitis, was referred to our hospital after the incidental discovery of gastric vari-

ces during a routine health screening endoscopy. Physical examination and laboratory test results were unremarkable.

Upper gastrointestinal (GI) endoscopy revealed isolated gastric fundus varices (IGVs) without esophageal varices or active bleeding. The varices were classified as IGV1 (large) and RCS0 according to the Sarin classification (Fig. 1A).

Contrast-enhanced CT revealed a 10 cm wedge-shaped spleen in the left upper quadrant (LUQ) and a 7 cm round mass on the medial side, both of which demonstrated similar enhancement patterns. The mass exhibited a zebra-like enhancement pattern and was supplied by branches of the splenic artery, suggesting a duplicated spleen. An enlarged vein draining from the mass led to dilated short gastric and coronary veins, forming fundic gastric varices that extended to the posterior wall of the stomach (Fig. 1B).

A cinematic volume-rendering technology image was added to illustrate the anatomical and structural aspects. There was no communication between the gastric fundic varix and the splenic vein (Fig. 1C).

Technetium-99m (^{99m}Tc)-labeled phytate scintigraphy showed two focal radiotracer uptakes in the LUQ, corresponding to the previously CT-observed wedge-shaped spleen and mass, confirming the duplicated spleen (Fig. 1D).

Doppler US also revealed a duplicated spleen and a 6 mm diameter vein between the spleen and the gastric fundus, suspected to be a gastric fundic varix. The portal vein (PV) was found to be normal, as evidenced by a resistive index of the hepatic artery above 0.5, a systolic acceleration time below 80 ms, hepatopetal flow in the PV, and a peak flow velocity in the hepatic vein (HV) over 20 cm/s and a venous pulsatility index exceeding 0.2 (Fig. 1E).

A splenectomy was performed due to the risk of life-threatening upper GI bleeding associated with untreated gastric varices.

Histological examination of one spleen, possibly the round shaped spleen in the image, revealed areas of dense fibrosis encircling chunky, yellow-to-brown iron and calcium deposits, identified as Gamna-Gandy bodies (GGBs) (Fig. 1F) (4).

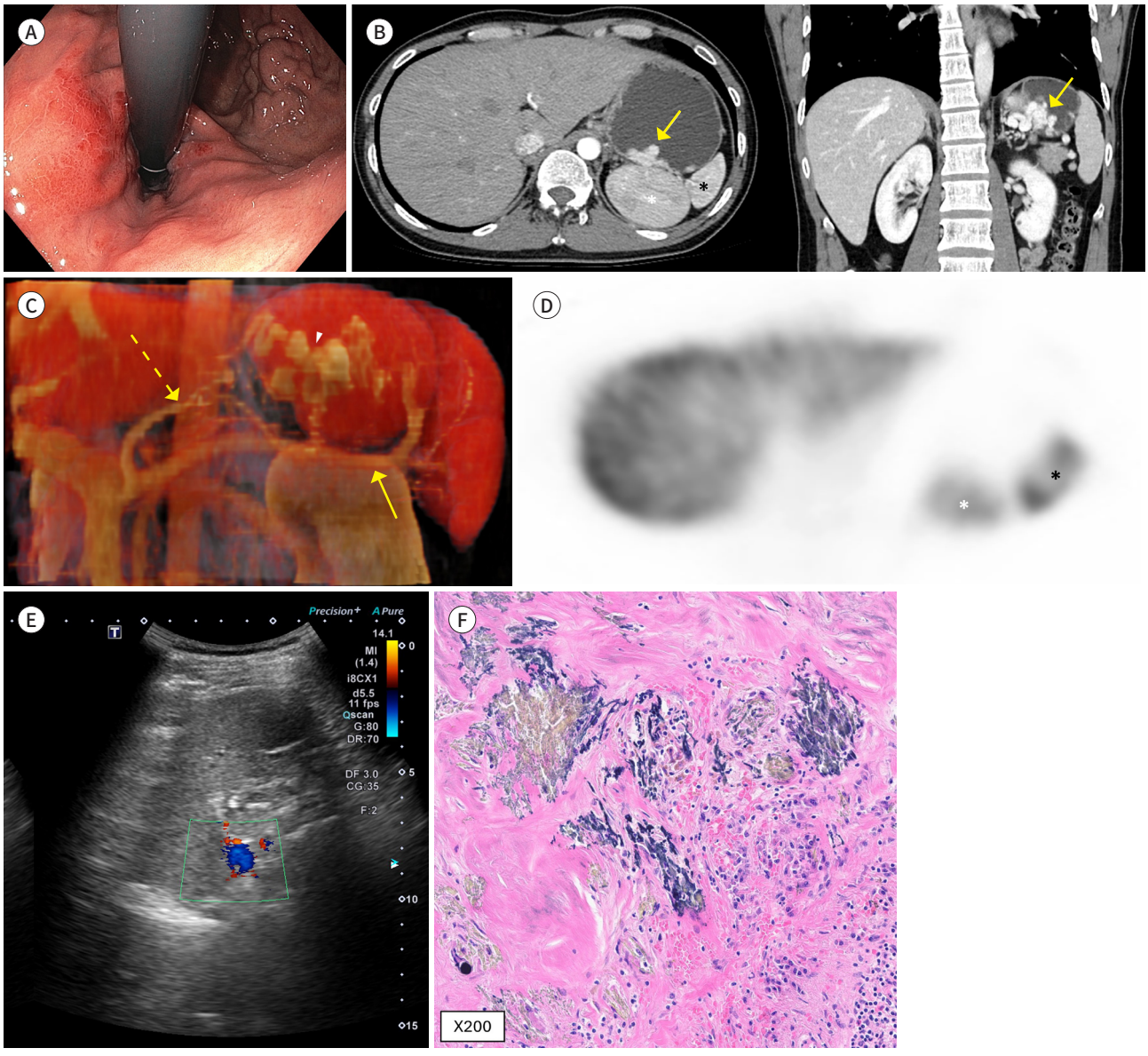
This study complies with the Declaration of Helsinki and written informed consent was obtained from the patient.

DISCUSSION

The spleen, a mesodermal derivative, initially appears as a cluster of mesenchymal cells within the dorsal mesogastrium toward the end of the fourth week of embryonic development. In its early form, mesenchymal cells differentiate into capsules, connective tissue frameworks, and splenic parenchyma. Improper fusion of these cells can result in congenital splenic anomalies, including polysplenia, accessory spleen, and splenic duplication. Other congenital splenic abnormalities encompass wandering spleen, splenic clefts, and lobulation (1, 2).

Polysplenia syndrome, also known as left isomerism, should be considered in patients with multiple spleens, such as our patient. It is typically characterized by equally sized and round spleens. Patients with polysplenia syndrome often have associated conditions such as congenital heart disease, shortened pancreas, unusual positioning of abdominal organs, or abnormalities in the inferior vena cava (IVC) (1, 2).

Fig. 1. A 40-year-old male with gastric varices secondary to splenic duplication.
A. Upper gastrointestinal endoscopy shows IGVs (IGV1, large, RCS0) without esophageal varices or active bleeding.
B. Contrast-enhanced CT. Axial and coronal images show a 10 cm-sized wedge-shaped spleen (black asterisk) and a 7 cm round-shaped mass (white asterisk) with similar enhancement characteristics, including a zebra pattern of enhancement. These structures are located adjacent to each other in the left upper quadrant of the abdomen. The round-shaped mass receives its blood supply from splenic artery branches. It forms an engorged draining vein that extends to the posterior wall of the stomach, leading to the development of gastric fundic varices (arrows).
C. A cinematic volume rendering technology image provides a clearer visualization of the anatomical structures of the duplicated spleen and their associated vessels, including the gastric fundic varix (arrowhead), left gastric vein (dashed arrow), and splenic vein (arrow). There is no communication between the gastric fundic varix and the splenic vein.
D. ^{99m}Tc-labeled denatured phytate spleen SPECT images. Axial images show focal uptake in two areas in the left upper quadrant of the abdomen, which correlate with the observed spleen (black asterisk) and nearby mass (white asterisk) located in the gastric fundus area on prior CT, confirming the presence of a duplicated spleen.
E. Hepatic Doppler US. Color Doppler shows a dilated vascular structure between the round-shaped spleen and gastric fundus with splenofugal flow, suspected to be the gastric fundic varix draining the round-shaped spleen.
F. Histologic examination of one spleen, hematoxylin-eosin staining possibly the round-shaped spleen seen in the image, shows areas of dense fibrosis encircling chunky, yellow-to-brown iron and calcium deposits, leading to a diagnosis of GGBs. The pathological presence of GGBs suggests organized hemorrhage, indicating regional portal hypertension.
GGBs = Gamna-Gandy bodies, IGV = isolated gastric fundus varices, RCS = red color signs, ^{99m}Tc = Technetium-99m



However, in our case, one of the two spleens was wedge-shaped rather than round. Moreover, our patient exhibited no congenital anomalies of other organs, and distinct branches of the splenic artery were observed entering each splenic hilum separately, suggesting splenic duplication without association with polysplenia syndrome (1, 2).

Previous reports have described the imaging findings of splenic duplication as two adjacent spleens with a similar enhancement pattern, including a zebra pattern and separate vascular structures, consistent with our observations (1, 2). The wedge-shaped spleen drains normally into the portal system via the splenic vein, while the round spleen drains through the gastric fundus varices into the portal system.

^{99m}Tc -labeled denatured red blood cell (RBC) scans can accurately identify splenic tissue, with studies demonstrating their high sensitivity and specificity (5). In this case, phytate was used instead of RBCs as a slightly less sensitive but still valid diagnostic method for identifying the spleen. Phytate successfully identified the two spleens, confirming splenic duplication.

IGVs are rare and typically occur secondary to regional, left-sided, or sinistral portal hypertension, often due to splenic vein obstruction. An altered pressure in the left portal system can cause venous flow reversal, resulting in variceal formation and potential GI bleeding. Left-sided portal hypertension is primarily caused by splenic vein thrombosis due to pancreatitis, with rare instances linked to regional inflammation, hepatopancreatobiliary cancer, or surgical splenic vein damage (6).

In this patient, a round, duplicated spleen developed with a draining vein that was not connected to the splenic vein, leading to partial left-sided portal hypertension.

This is supported by three key findings. First, there was no draining vessel observed from the round-shaped spleen to the splenic vein, but a vessel draining from it connected to the gastric fundic varix, with engorgement of the left gastric vein. The reflux of venous blood through the short gastric veins likely elevated intravenous pressure, leading to the development of gastric varices in the fundus. Morphological changes such as a rounded shape and blunted edges in this patient may be indicative of venous congestion associated with this condition. Second, the pathological presence of GGBs suggests organized splenic hemorrhage, indicating regional portal hypertension (4). Lastly, the complete resolution of the varix after splenectomy further supports this conclusion of partial left-sided portal hypertension.

A previous study suggested that increased total splenic volume might lead to increased blood flow and potentially cause gastric varices (2). In this case, CT volumetry revealed that the two spleens had volumes of 113.20 mL and 97.78 mL, respectively, totaling 210.98 mL. This is slightly larger than the average spleen volume of 178.14 mL for a Korean male in his 40s (7). Therefore, it seems unlikely that increased splenic volume alone was responsible for the gastric varices observed in this patient.

Given the rarity of splenic duplication as a cause of IGVs, it is essential to rule out more common etiologies, such as liver cirrhosis (LC), before attributing varices to this rare condition. LC and portal hypertension were excluded due to normal liver US, CT, FibroScan, liver function tests, and normal PV Doppler findings (8). Despite the potential for portosinusoidal vascular disease to cause portal hypertension with normal FibroScan results, it was ruled out due to the absence of characteristic findings such as liver remodeling, splenomegaly, focal nodular hyperplasia-like nodules, other varices or shunts, and PV thrombosis (9). Budd-Chiari syndrome,

characterized by the obstruction of hepatic venous outflow, leading to elevated portal pressure and varices, was excluded due to patent HVs and IVC on CT and Doppler US.

Furthermore, there were no signs or history suggestive of other rare causes of the varices, such as sarcoidosis, congenital hepatic fibrosis, or sinusoidal obstruction syndrome. As there were no other clear causes of the gastric varices, it could be concluded that the condition in this patient was primarily due to splenic issues.

The optimal treatment for gastric varices remains controversial (2, 10). In this case, given the patient's relatively young age and good health, a splenectomy was performed to address the root cause. Interventional radiology techniques are becoming increasingly important for the treatment of splenic diseases that can lead to gastric varices. However, in this case, balloon-occluded retrograde transvenous obliteration was not feasible due to the absence of a gastrorenal or gastrocaval shunt, a prerequisite for this procedure. Embolization was also considered but deemed unsuitable due to the risk of splenic infarction. Band ligation was not recommended because of the location of the IGV in the fundus, greater curvature, and submucosa, which made it difficult to completely ligate the varix and opposing wall, potentially leading to continued blood flow and massive bleeding (2, 6, 10).

In conclusion, we described a rare case of gastric varices secondary to splenic duplication, a seldom observed cause. A preoperative diagnosis is possible using various imaging modalities. This study contributes to the limited documentation of this condition in medical literature.

Author Contributions

Conceptualization, L.T.Y.; investigation, all authors; supervision, L.T.Y.; writing—original draft, all authors; and writing—review & editing, all authors.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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비장 중복, 위 정맥류의 드문 원인: 증례 보고

김슬기[1] · 이태영[1,2*]

비장 중복은 종종 다비 증후군이라고도 불리며, 비장이 비슷한 크기의 조각들로 나뉘어 있는 경우로, 가끔씩 관찰된다. 하지만, 중복된 비장에 의해 발생하는 위저부 정맥류는 극히 드물며, 이러한 의학적 이상은 문헌에서 거의 보고된 바 없다. 이 증례 보고는 비장 중복으로 인한 위저부 정맥류의 드문 사례를 보이는 40세 남성 환자를 제시한다. 내시경, CT, 도플러 초음파 및 핵의학 비장 스캔을 포함한 포괄적인 영상 검사를 통해 진단을 했다. 이 증례는 의학 문헌에 귀중한 정보를 제공하여, 거의 논의된적 없는 질환에 대한 이해를 돕고 있다.

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