

Portal, Splenic and Mesenteric Thrombosis in Hypereosinophilic Syndrome: A Case Report¹

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Idiopathic hypereosinophilic syndrome is a spectrum of diseases characterized by prominent peripheral eosinophilic leukocytosis without an identifiable cause. Several reports have described hepatic involvement as depicted on sonography and CT imaging in patients with hypereosinophilic syndrome. However, thrombosis of the portal, splenic and mesenteric veins in hypereosinophilic syndrome has been rarely reported. We present here a case of portal, splenic and mesenteric thrombosis in a 33-year-old man with hypereosinophilic syndrome.

Index words : Thrombosis

Portal vein

Mesenteric veins

Eosinophilia

Liver

Tomography, X-ray computed

Hypereosinophilic syndrome is a spectrum of diseases characterized by prominent peripheral eosinophilic leukocytosis without an identifiable cause (1, 2). Eosinophilic infiltration occurs in various systems and organs such as the nervous system, heart, hematopoietic system, liver, spleen, bowel and lung.

The diagnostic criteria of idiopathic hypereosinophilic syndrome includes peripheral eosinophilia of more than 1500 eosinophils/mm³ for more than six months, absence of parasitic, allergic or other causes of eosinophilia and evidence of organ involvement (3). Several reports have described the sonographic and computed tomography (CT) imaging findings in patients with hepatic in-

volvement (4, 5); however, portal, splenic and mesenteric thrombosis in hypereosinophilic syndrome has been rarely reported (6). We report a case of portal, splenic and mesenteric thrombosis in a 33-year-old man with hypereosinophilic syndrome.

Case Report

A 33-year-old man was admitted for evaluation of dyspnea, dizziness and abdominal distention. On a physical examination, a large amount of ascites was suggested. Based on laboratory results, the WBC was 15,300/mm³ with 53% of eosinophils. The patient had no history of any allergic disease or ingestion of raw animal meat or liver. Stool, skin and ELISA tests for parasites such as *Paragonimus westermani*, *Clonorchis sinensis*, *Sparganosis* and *Toxocara canis* were negative. The patient underwent a high-resolution chest CT examination for evaluation of dyspnea and a multiphase abdominal CT exami-

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nation for evaluation of ascites. An unenhanced abdominal CT scan was performed and arterial, portal venous phase scans were performed after intravenous contrast administration. A chest CT image showed multifocal patchy consolidations and ground-glass opacities with mainly peripheral distribution in both lungs and bilateral pleural effusion (Fig. 1A). An unenhanced abdominal CT image showed the presence of a hyperdense thrombus in the portal vein and the splenic vein (Fig. 1B, C). A contrast-enhanced abdominal CT image obtained during

the portal venous phase showed an enlarged liver with diffuse heterogeneous low attenuated areas, splenomegaly and a large amount of ascites (Fig. 1D). After the initial CT examinations, an abdominal CT scan was performed three days later for evaluation of newly developed acute severe left upper abdominal pain. A contrast-enhanced abdominal CT image obtained during the portal venous phase showed progression of thrombosis in the splenic vein, acute splenic infarction and newly developed thrombosis in the superior mesenteric

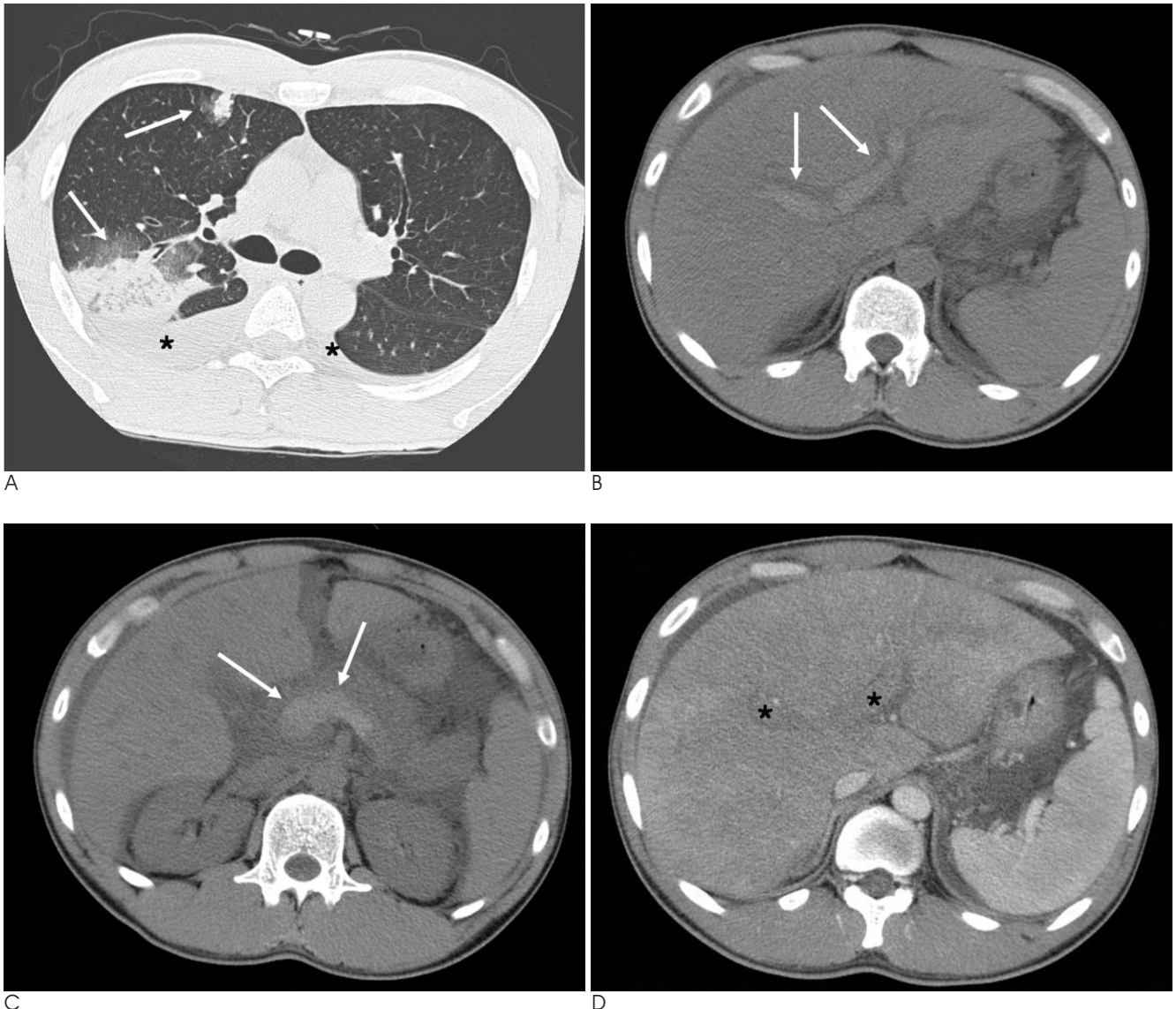


Fig. 1. Imaging findings are presented for a 33-year-old man with hypereosinophilic syndrome combined with venous thrombosis in the portal, splenic and superior mesenteric veins.

A. An axial chest CT image with the lung window setting shows two areas of patchy consolidations (arrows), surrounded by zones of ground glass attenuation in the right upper lobe and a small amount of bilateral pleural effusion (asterisks).

B. An axial unenhanced CT image shows the presence of a hyperdense thrombus in the right and left portal veins (arrows).

C. A CT image obtained just inferiorly shows the hyperdense thrombus in the main portal vein and splenic vein (arrows).

D. An axial contrast-enhanced CT image obtained during the portal venous phase shows diffuse low attenuation of the enlarged liver with heterogeneity, splenomegaly and ascites, in addition to portal vein thrombosis (asterisks).

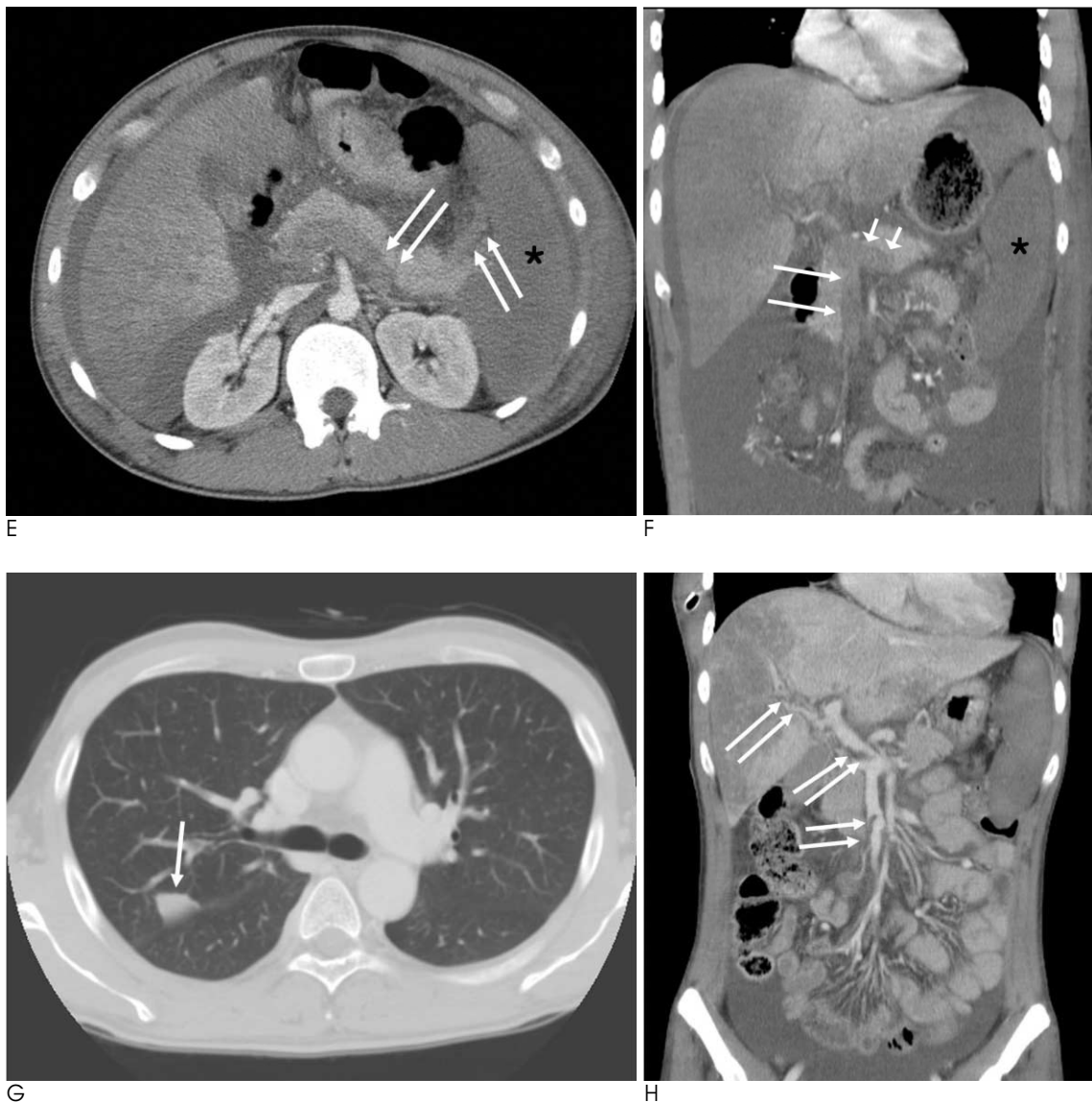


Fig. 1. E. A follow-up CT image obtained three days after the initial CT examinations demonstrates the progression of the splenic venous thrombus (arrows) and acute splenic infarction (asterisk). F. On a coronal CT image obtained during the portal venous phase, superior mesenteric vein thrombosis (long arrows) is demonstrated beside the splenic vein thrombosis (short arrows) and splenic infarction (asterisk). The CT image also shows heterogeneous low attenuation of the liver and a large amount of ascites. G. A follow-up CT image obtained four weeks after the initial steroid therapy reveals disappearance of patchy consolidations and ground glass attenuation, except for a small focal consolidation (arrow) in the right upper lobe. H. On a coronal CT image, thrombi in the portal, splenic and mesenteric veins have disappeared, except for small thrombi (arrows) in the right portal vein and superior mesenteric vein. The CT image also shows improvement of heterogeneous low attenuation of the liver and ascites.

vein (Fig. 1E, F). Under the clinical impression of hyper-eosinophilic syndrome, the patient underwent systemic corticosteroid therapy for one month. As determined from follow-up laboratory results, the WBC was $7,400/\text{mm}^3$ with 0.7% of eosinophils. Follow-up chest and abdominal CT examinations were performed after the corticosteroid therapy. A chest CT image demonstrated improvement of the multifocal patchy areas of

consolidations and ground-glass opacities, except for a small consolidation in the right upper lung (Fig. 1G). On an abdominal CT image, portal, splenic and mesenteric thrombosis disappeared except for the presence of small thrombi in the main portal vein, the right portal vein and superior mesenteric vein (Fig. 1H). With the findings of hypereosinophilia in the peripheral blood, evidence of organ involvement and the response to cor-

ticosteroid therapy, idiopathic hypereosinophilic syndrome was diagnosed.

Discussion

Hepatic involvement occurs in 40–90% of patients with hypereosinophilic syndrome, and usually presents with hepatomegaly and abnormal liver function (1, 2). Previous studies about the CT findings of hepatic involvement in hypereosinophilic syndrome have demonstrated the presence of small low attenuation lesions that are scattered throughout the liver, especially in areas adjacent to the portal veins, narrowing of the intrahepatic portal veins and lobar or segmental low-attenuated lesions (4, 5). However, portal, splenic and mesenteric thrombosis in hypereosinophilic syndrome has rarely been reported (6). The most frequent causes of portal vein thrombosis are a myeloproliferative disorder, liver cirrhosis with portal hypertension, deficiency of natural anticoagulant proteins, gene mutations and hepatocellular carcinoma (7). Hypercoagulable syndrome can lead to portomesenteric and splenic vein thrombosis and can be complicated with acute or subacute intestinal angina (8). Thus, early diagnosis and treatment of portomesenteric and splenic vein thrombosis in hypercoagulable syndrome is important to avoid complications such as intestinal angina (8). One of the causes of hypercoagulability is hypereosinophilic syndrome (6, 9, 10).

Hypereosinophilic syndrome has been associated with various thrombotic manifestations such as deep venous thrombosis, cerebral venous thrombosis, hepatic veno-occlusive disease or the presence of intracardiac thrombi (9, 10). Activated eosinophils and granular proteins including eosinophil cationin protein and major basic protein are thought to modify coagulation and fibrinolysis in eosinophilia, resulting in thrombosis (6, 9, 10). Thus, venous thrombosis with hypereosinophilic syndrome

can progress rapidly, and corticosteroid and anti-coagulant therapy should be considered to avoid complications (6, 9, 10).

In summary, we experienced a case of rapidly progressing portal, splenic and mesenteric thrombosis in a patient with hypereosinophilic syndrome. The splenic infarction was complicated during disease progression. When portal, splenic and mesenteric thrombosis is associated with eosinophilia and pulmonary consolidation, hypereosinophilic syndrome should be considered in the differential diagnosis to avoid complications due to rapid progression of venous thrombosis.

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호산구증가증 환자에서 나타난 간문맥과 비장정맥 및 장간막정맥 혈전증: 증례 보고¹

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황수연 · 장경미 · 김민정 · 이관섭 · 고성혜 · 전의용 · 이 현 · 최주현 · 이미연

특발성 호산구증가증은 밝혀진 원인 없이 말초혈액의 호산구가 증가하는 질환이다. 호산구증가증에서 다양한 장기에 다량의 호산구들이 침착하여 다양한 영상 소견을 보일 수 있다. 주로 간을 침범한 환자에서 초음파 소견과 CT 소견에 대해 보고되었으나, 간문맥과 비장정맥 및 장간막정맥을 침범한 혈전증은 잘 알려져 있지 않다. 이에 본 저자들은 특발성 호산구증가증에 동반된 간문맥과 비장정맥 및 장간막정맥을 침범한 혈전증에 대한 증례를 보고하고자 한다.