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Hemothorax and Bloody Ascites Caused by Vascular Ehlers-Danlos Syndrome

To the Editor:

We report a case of vascular Ehlers-Danlos syndrome (EDS) in a 29-year-old man without any underlying diseases, including atherosclerosis, and without any medical or family history. On admission, he had normal blood pressure (122/69 mm Hg) and was anemic (hemoglobin level of 5.9 g/dL). Contrast-enhanced computed tomography (CT) revealed hemothorax and a nodular lesion in the right lung, hemorrhagic ascites (Figure A), and aneurysms in the common hepatic artery and hepatic artery proper without extravasation (Figure B). The patient was admitted to an intensive care unit, and administration of an antihypertensive drug and transfusion of red blood cells were initiated. On the second day of hospitalization, his anemia was not improved despite the red blood cell transfusion. Contrast-enhanced CT, showed the deterioration of the bloody ascites. Transarterial embolization and pleural drainage were performed. On the seventh day of hospitalization, CT showed that the hemothorax and hemorrhagic ascites had been well absorbed. On the 10th day of hospitalization, brain magnetic resonance imaging revealed an aneurysm in the right internal carotid artery and right vertebral artery (Figure C). His blood pressure become stable (systolic blood pressure:120 mm Hg). He was discharged with oral antihypertensive medication on the 18th day of hospitalization. He had a *COL3A1* gene mutation, which is a pathological variant of vascular EDS.

VASCULAR EHLERS-DANLOS SYNDROME

Vascular EDS is a rare vascular abnormality in which aneurysms and raptures can occur. It is caused by mutations

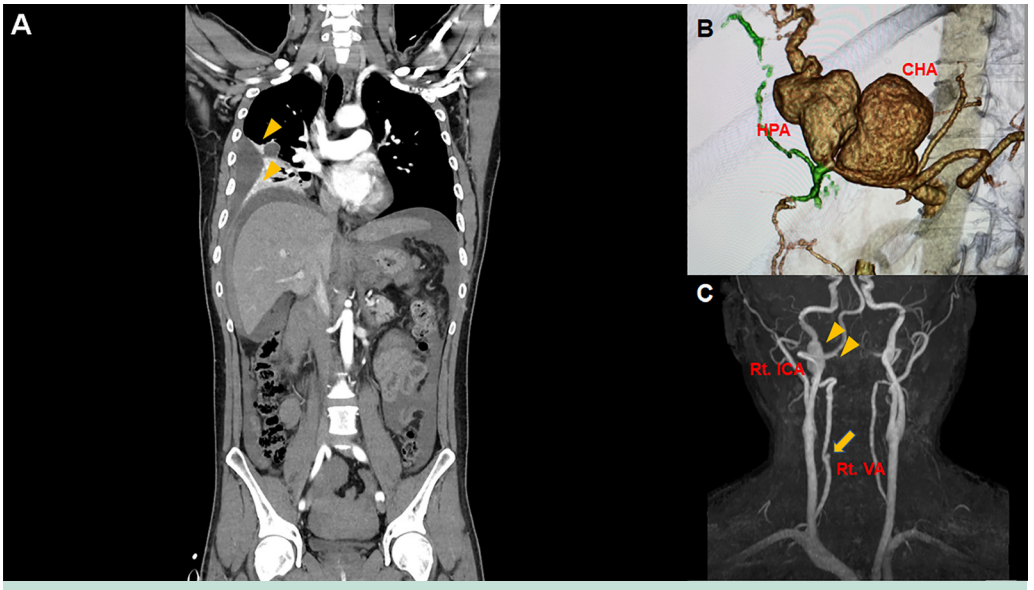

Figure **(A)** Right hemothorax (arrowhead) and bloody ascites shown by a contrast-enhanced computed tomography scan. **(B)** Aneurysm in the CHA and PHA. **(C)** Right ICA (arrowhead) and right VA (arrow) on plane magnetic resonance imaging. CHA = common hepatic artery; ICA = internal carotid artery; PHA = hepatic artery proper; VA = vertebral artery.

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in the gene for type III procollagen (*COL3A1*).[1] Up to 7% of patients with vascular EDS experience a first major event by the age of 20 years, and 40% of patients experience their first major complication by the age of 40 years.[2] The majority of affected arteries are medium-sized abdominal vessels, including renal, iliac, femoral, mesenteric, and hepatic arteries. The next most frequent locations of arterial abnormalities are the carotid, subclavian, ulnar, popliteal, and tibial arteries.[2] There is no established treatment for vascular EDS, and management includes maintaining blood pressure of less than 130/80 mm Hg[3] and managing for ruptures of aneurysms.[2] Clinical management of patients with vascular EDS remains challenging; however, it is important to follow patients in close collaboration with several specialists.

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References

1. Pepin M, Schwarze U, Superti-Furga A, Byers PH. Clinical and genetic features of Ehlers-Danlos syndrome type IV, the vascular type. *N Engl J Med* 2000;342(10):673–80. https://doi.org/10.1056/NEJM200003093421001.
2. Eagleton MJ. Arterial complications of vascular Ehlers-Danlos syndrome. *J Vasc Surg* 2016;64(6):1869–80. https://doi.org/10.1016/j.jvs.2016.06.120.
3. Erbel R, Aboyans V, Boileau C, et al. 2014 ESC guidelines on the diagnosis and treatment of aortic diseases. *Eur Heart J* 2014;35(41):2873–926. https://doi.org/10.1093/eurheartj/ehu281.