Case Report

Quite a backup: pericardial varices in a patient with hereditary antithrombin deficiency

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Introduction

A 39 year-old female with known hypertension, gastro-esophageal reflux disease, and hereditary anti-thrombin deficiency (hATD) on life-long anti-coagulation presented to our emergency room with acute atypical chest and epigastric pain. Her hATD was diagnosed five years prior when repeated deep vein thromboses were discovered. At that time, the patient was started on life-long anticoagulation with warfarin and was able to consistently achieve therapeutic levels.

Case report

Physical examination of the patient was unremarkable except for mild tenderness to palpation in the epigastric region. Chest radiograph was significant for a circumscribed opacity in the left cardiophrenic angle, isodense to the heart (*Figure 1*). Contrast-enhanced computed tomography (CT) of her chest, abdomen and pelvis demonstrated evidence of portal thrombosis with cavernous transformation and extensive varices with extension to the left pericardium (*Figures 2-4*).

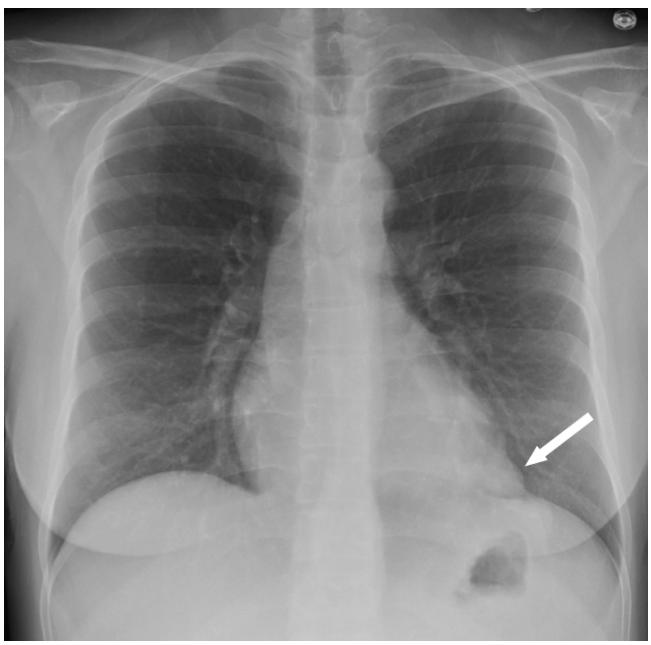

Figure 1 A frontal chest radiograph demonstrated a circumscribed bulbous opacity (arrow) in the left cardiophrenic angle, isodense to the heart

Discussion

Anti-thrombin (AT) is a vitamin-K independent glycoprotein that is endogenously produced by the liver and inhibits thrombin, factor Xa, and factor IXa (1).

While rare, hATD, the first identified hereditary thrombophilia, has an autosomal dominant pattern with variable penetrance (2). Individuals with hATD, like other inherited thrombophilias, have a genetic predisposition for forming venous thromboembolisms. The initial thrombotic event in 40% of people with hATD occur spontaneously while the remaining 60% occur in association with other pro-thrombotic risk scenarios such as pregnancy, oral contraceptive use, surgery, and trauma. In a large Spanish study approximately 13% of venous thromboembolisms were caused by inherited thrombophilias (3). The most

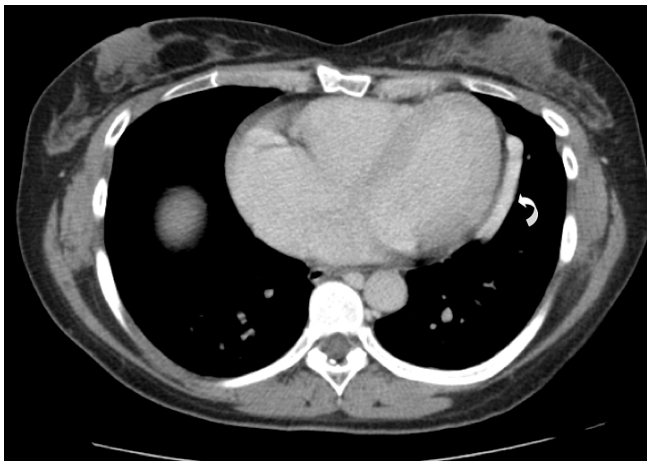

Figure 2 Axial CT image obtained after intravenous (IV) contrast injection demonstrated a large enhancing paracardiac varix (curved arrow) correlating with the radiographically seen opacity

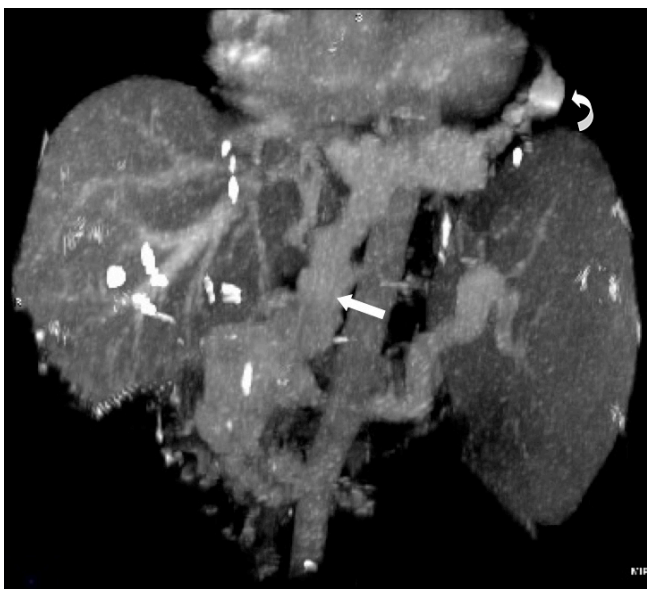

Figure 4 A thick maximum intensity projection (MIP) image better defined the course of the massive abdominal varix (straight arrow) and communication with the paracardiac varix (curved arrow)

common anatomical sites include deep veins of the lower extremities and mesenteric veins with up to 60% of patients developing recurrent thrombotic events (4).

While the incidence and prevalence of cardiac varices in patients with inherited thrombophilias remains unknown, 20% of patients with portal hypertension appear to have cardiac varices with most occurring on the right (90%)

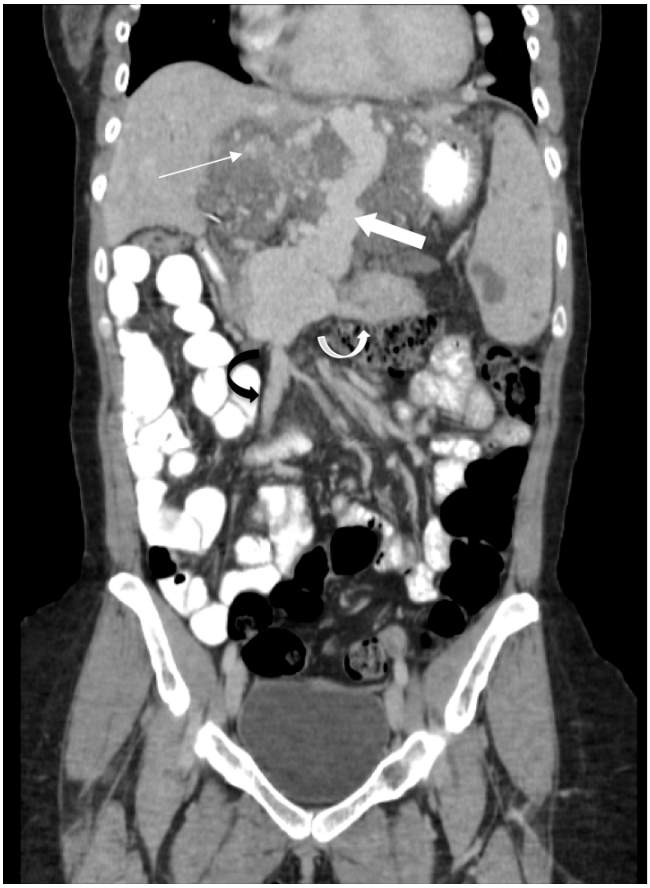

Figure 3 Coronal CT image obtained after contrast injection showed a tortuous massive varix (large white arrow) formed by the confluence of the superior mesenteric (black curved arrow) and splenic (white curved arrow) veins. The varix extended in the direction of the diaphragm. Multiple smaller varices (small white arrow) branch off the main varix towards the porta hepatis

compared to the left (5).

Currently, patients with hATD and one or more spontaneous thrombosis are recommended to start indefinite anti-coagulation (6). In the setting of acute thrombosis in patients with hATD, heparin should be used with caution as it typically quantitatively lowers AT levels by up to 30% (7). A pooled concentration of AT has been used, and is recommended, for patients with hATD and recurrent or severe thrombosis despite anti-coagulation or with difficulty achieving therapeutic levels of anti-coagulation (8).

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