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Case Report

Internal Medicine Section

Multiple Abdominal Veins Thrombosis Secondary to Protein S Deficiency - A Case Report

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ABSTRACT

Abdominal venous thrombosis may present either as Budd-Chiari syndrome (BCS) caused by hepatic vein or proximal inferior vena cava (IVC) obstruction or as an extra hepatic portal obstruction (EHPVO) caused by Portal vein thrombosis or mesenteric vein thrombosis, but a mixed involvement is uncommon. Multiple abdominal venous obstructions presenting with thrombosis of hepatic vein, IVC, portal vein and renal vein are very rarely seen. We are reporting a rare case with thrombosis of IVC, hepatic vein, portal vein and renal vein, with protein S and protein C deficiencies, which was managed by giving anticoagulant therapy.

Keywords: Hepatic vein thrombosis, Inferior vena cava (IVC) thrombosis, Renal and portal vein thrombosis

CASE REPORT

A 38-year-old female presented to General Medicine Department with complaint of localized, beaded swellings of both lower limbs which had occurred two years ago, which had gradually increased in size since their onset. Two months ago, she had fever for 10 days without chills and later, she had developed localized beaded swellings in left flank and then around umbilicus [Table/Fig-1]. They had gradually increased in size. Patient had no history of melena, haematemesis, jaundice, abdominal distension and abdominal pain. She also had no history of previous usage of oral contraceptive pills, trauma, prolonged bed rest or major abdominal operations. In the past, she had one episode of induced abortion which was medically not significant. She had not conceived later. Her siblings were normal. Her family or relatives had no similar complaints.

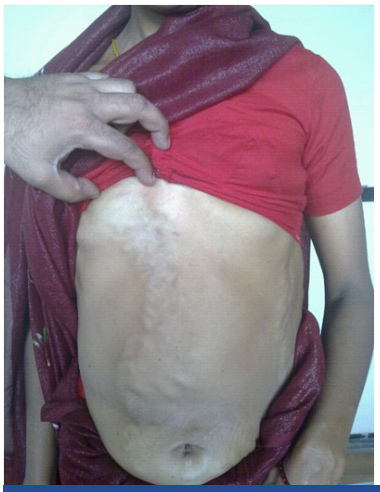
On examination, she was found to be moderately built. In her lower limbs, varicose veins with perforator incompetence pattern, without deep vein thrombosis pattern were seen. They extended from medial malleoli to sapheno-femoral junction [Table/Fig-2]. On abdomen, in the left flank, collaterals were found to be present, which had flow in IVC pattern. On anterior wall, around umbilicus also, IVC pattern was found to be present [Table/Fig-1]. On back also, dilated but untortuous veins were prominently seen [Table/Fig-3]. All other systems were essentially normal, except for mild splenomegaly.

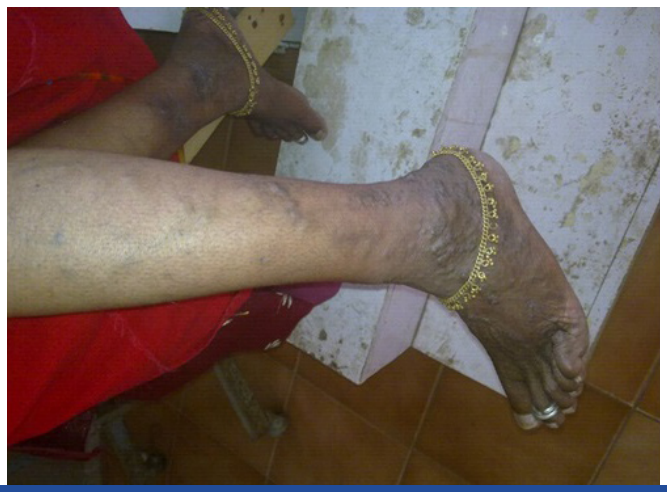
All routine investigations, antiphospholipid antibody, homocysteine and antithrombin-III (AT III) were within normal limits. Protein C level was 56.5% (normal range -70%-140%), which was just below normal limits, but protein S levels had grossly decreased, with a value of 13.1% (normal range -72%-106%). D-dimer values were more than 200 micro gm/ml. USG of abdomen and CECT of abdomen had shown caudate lobe hypertrophy, a mildly enlarged spleen, portal vein narrowing (? secondary to fibrosis of thrombus) from hepatic to renal vein, IVC thrombosis, renal vein thrombosis and extensive collaterals [Table/Fig-4a,4b,5]. Lower limb Colour Doppler studies had shown perforator incompetence without deep vein thrombosis. Upper gastrointestinal endoscopy revealed grade 2 oesophageal varices. 2D–ECHO and CT of brain showed normal results.

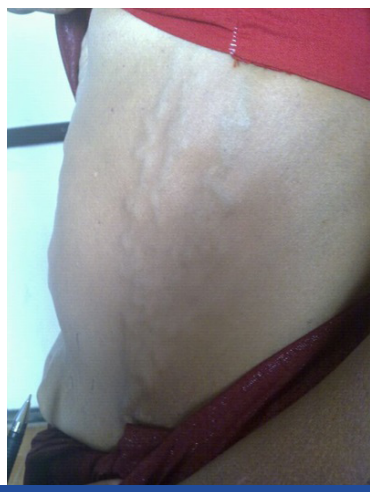
We started treatment with low molecular weight heparin which was given for week, followed by warfarin and finally, she was discharged on 2 mg warfarin with a prothrombin time/ international normalized ratio (PT/INR) of 2.75.

DISCUSSION

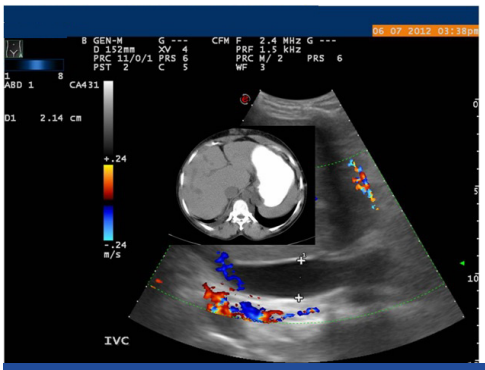
Abdominal venous thrombosis is a rare disorder. It presents either as Budd-Chiari syndrome (BCS) or splanchnic vein thrombosis (SVT), but a mixed involvement is uncommon. Venous thromboembolism (VTE) is a common event, which is often precipitated by surgery,

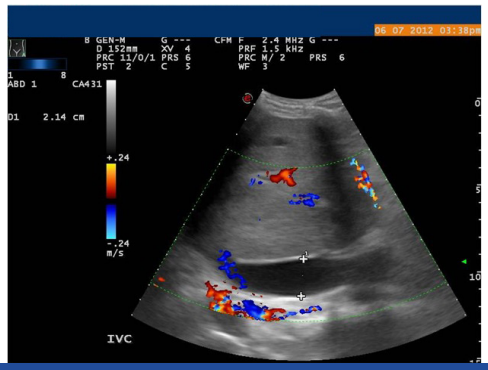


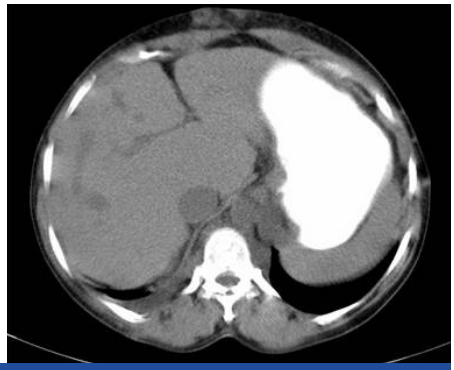




[Table/Fig-1]: Collaterals on anterior wall around umbilicus and on left flank
[Table/Fig-2]: Lower limbs varicose veins
[Table/Fig-3]: Dilated but untortuous veins on back







[Table/Fig-4a]: Doppler showing decreased color flow in portal vein
[Table/Fig-4b]: Doppler showing dilated IVC with no color filling s/o thrombus
[Table/Fig-5]: Image showing IVC thrombosis

immobility or active malignancy [1]. The diagnostic work up done for the patients includes testing for inherited and acquired hypercoagulable conditions, which usually includes functional quantitative assays for proteins C and S and antithrombin, as well as testing for lupus anticoagulant, antiphospholipid antibodies, activated protein C resistance (with or without genetic testing for Factor V Leiden) and determination of the G20210A Prothrombin gene mutation [2]. Among hereditary risk factors, Factor V Leiden mutation is common in BCS and protein C deficiency is common in portal vein thrombosis (PVT) [3]. Deficiency of protein C is more common than protein S deficiency in both BCS and PVT [4]. The level of protein S was low and that of protein C was just below the normal level in the present case.

Inherited thrombophilia is found in patients with SVT, although making diagnoses of inherited deficiencies of antithrombin, protein C, and protein S is difficult in the presence of liver impairment, which causes a reduced synthesis of the natural anticoagulant proteins [5-8]. This difficulty is not seen with the Factor V Leiden and prothrombin G20210A mutations. Interestingly, a high prevalence of prothrombin G20210A mutation has been consistently reported in various series of patients with EHPVO [6-8], whereas Factor V Leiden appears to be more common in BCS patients [8]. A 4-fold increased risk for prothrombin G20210A has been reported in EHPVO patients and a 2-fold increased risk for factor V Leiden has been reported in BCS patients [8]. D-dimer is considered as a marker of hypercoagulable state, besides endogenous fibrinolysis and so, increased D-dimer levels are detectable in patients who are affected by arterial and/or venous thrombosis, as it was seen in the present case [9]. It was reported that PVT which was provoked by protein C and S deficiencies could be recanalized by giving short-term low molecular heparin plus oral warfarin therapies [10]. In the present case, patient was relieved of varicose veins by giving low molecular weight heparin, followed by oral warfarin for four weeks and she was discharged on 2 mg warfarin. She was advised to come for follow-ups . The present case of multiple abdominal vein thrombosis is a rare case with protein S and protein C deficiencies.

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