A case of celiac artery and inferior mesenteric artery thrombosis in a patient with protein C & S deficiency

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ABSTRACT

Thrombosis of the celiac artery along with inferior mesenteric artery causing mesenteric ischemia is an emergency surgical condition with high mortality and morbidity when there is delay in diagnosis and treatment. Protein C deficiency is associated with an increased incidence of venous thromboembolism, whereas no association with arterial thrombotic disease has been found. Here we report a case of a young patient with celiac artery and inferior mesenteric artery thrombosis presented as colonic gangrene with peritonitis secondary to protein C & S deficiency, hyperhomocysteinemia and thrombocytosis which was managed with anticoagulant therapy and minimal invasive surgery.

Key words: Celiac artery thrombosis, Hyperhomocysteinemia, Inferior mesenteric artery thrombosis, Protein C & S deficiency, Thrombophilia, Thrombocytosis.

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INTRODUCTION

Thrombosis of the celiac artery along with inferior mesenteric artery causing mesenteric ischemia is an emergency surgical condition with high mortality and morbidity when there is delay in diagnosis and treatment[1]. Protein C deficiency is associated with an increased incidence of venous thromboembolism, whereas no association with arterial thrombotic disease has been found[2].To our knowledge this is the first case report of celiac artery and inferior mesenteric artery thrombosis with colonic gangrene secondary to protein C & S deficiency, hyperhomocysteinemia and thrombocytosis which was managed with anticoagulant therapy and minimal invasive surgery.

CASE HISTORY

A 33 years old gentleman presented in our emergency department with history of post prandial abdominal pain since 6months with increased pain

since 10 days associated with loose stools and vomiting for 2days along with malena for 2 days. He had a history of weight loss of 20kgs in 6months and was smoking 20 cigarettes per day since 10years. On examination his pulse rate was 100/min, Blood pressure was 90/60mm of Hg, Temperature was 36.9C .On examination patient had umbilical, supra pubic and left iliac fossa tenderness without rebound tenderness.

Laboratory investigations showed haemoglobin of 9.1gm/dl, total counts of 15,280 cells/mm^3 with neutrophils 87%, lymphocytes of 8% with raised platelet count to 5,12,000/mm^3, CRP-120. Repeated platelet count was 7,00,000/mm^3 the next day. CT-scan abdomen revealed ischemic necrosis of descending colon with near total occlusion of celiac axis with collateral circulation established through gastro duodenal and gastro epiploic arteries filling distal celiac artery (Figure 1, 2)

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Figure 1: CT scan of abdomen showing descending colon necrosis.

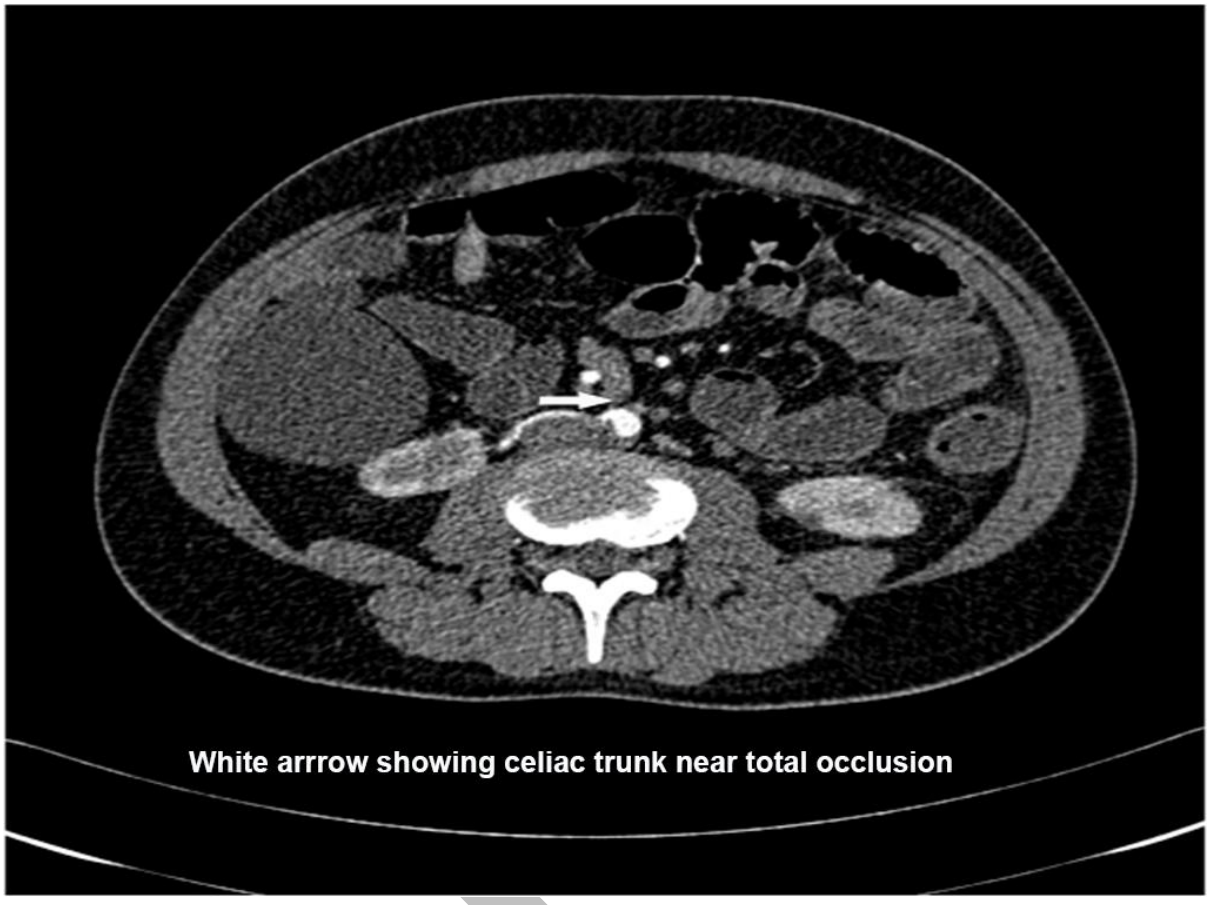

Figure 2: CT scan abdomen showing near total occlusion of the Celiac trunk.

There was occlusion of inferior mesenteric artery from its origin with bilateral renal and splenic infarcts (Figure 3).

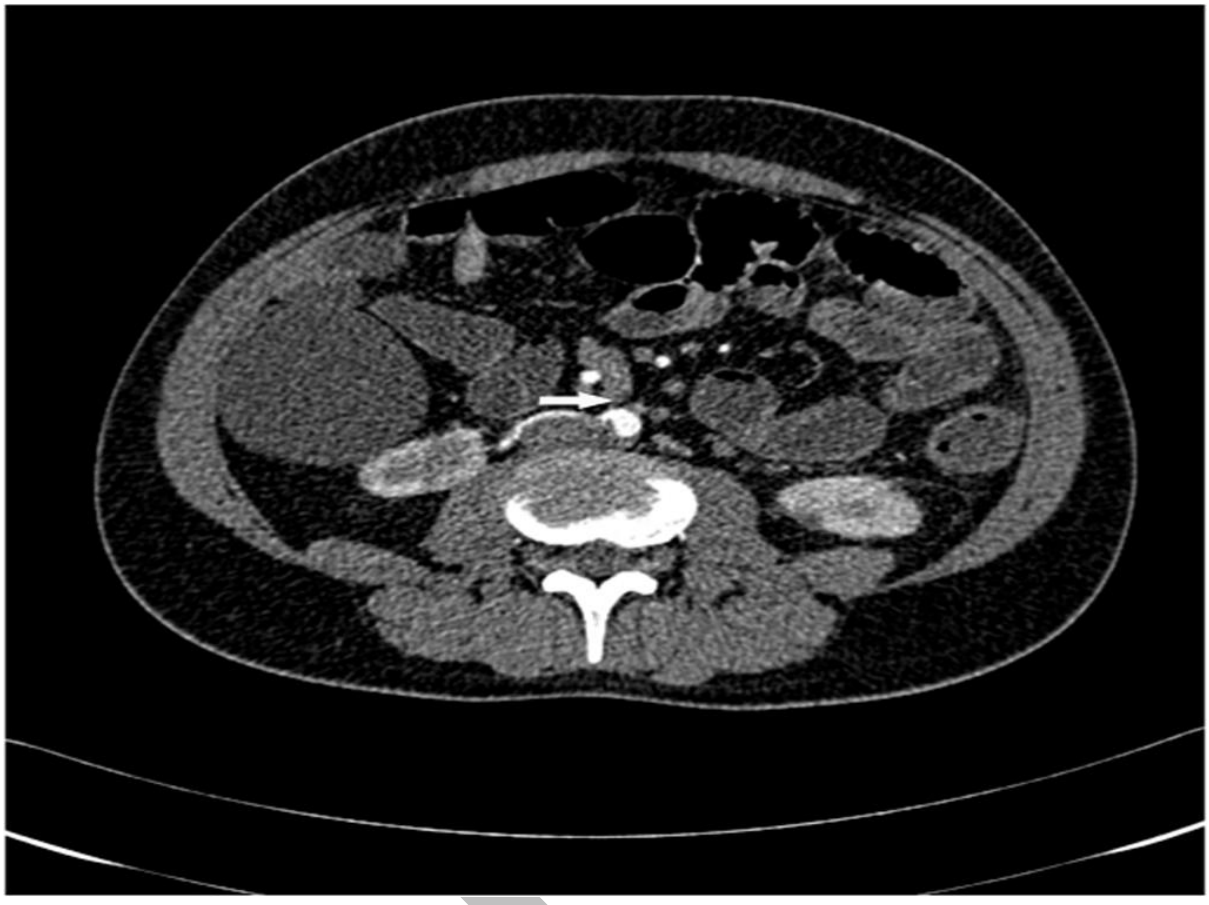

Figure 3: CT scan abdomen showing Inferior mesenteric artery thrombosis at the origin.

Autoimmune profile had done revealed Lupus anticoagulant positive with ANA, ANCA C, ANCA P, Anti phospholipid antibodies and anti cardiolipin antibodies being negative. Thrombophilia profile showed deficient Protein S antigen (free) 39.2 % (72.2% - 149.8%), low Protein C antigen 1.35mg/L (normal range-1.67-3.16mg/L) with low Protein S activity 48% (77%-143%). Anti thrombin III antigen and activity were normal with no Activated Protein C resistance (APCR). Patient's serum homocysteine value was very high 50 micro mol/L (5.46-16.2 micro mol/L).

Patient was resuscitated with crytalloids, intravenous antibiotics and was started on low molecular weight heparin with nasogastric aspiration. Diagnostic laparoscopy was performed which showed gangrenous colon with multiple perforations extending from lateral one third of transverse colon upto rectosigmoid junction (Figure 4).

Figure 4: Descending colon gangrene with mucosal sloughing

Patient underwent laparoscopic left extended hemi colectomy with transverse colon end colostomy and distal rectal stump as mucus fistula. Patient had uneventful post operative recovery and was discharged after 7 days. Patient was taken up for laparoscopic resortoration of colon after 3 months. Patient is presently on long term oral dicoumarol (warfarin) treatment.

DISCUSSION

Thrombophilia predisposing to venous thromboembolism of mesenteric venous circulation leads to mesenteric ischemia with high mortality and morbidity [1]. Occlusion of celiac, superior mesenteric, inferior mesenteric artery was reported by Cakmak et al in a 48year old patient with peripheral thromboangitis obliterans (Beurger's disease)[3]. Deficiencies in protein S, protein C, and antithrombin account for venous thromboembolism, while rare causes include the dysfibrinogenemias [4, 5].

Hasan et al have reported celiac artery thrombosis and superior mesenteric artery stenosis in a patient with essential thrombocythemia [6]. Patients usually develop venous thrombosis with protein C and S deficiency [2]. C. W. Kim and J. W.

Kim reported a case of celiac artery thrombosis and splenic infarction which was related to protein S deficiency [7]. Romano et al reported a case of superior mesenteric artery thrombosis in a 39yrs old woman with protein S deficiency [8].

In our patient thrombophilia work up showed protein C & S deficiency with platelet count of 7,00,000/mm^3 along with hyperhomocysteinemia, and lupus coagulant positive predisposing patient to arterial thrombosis. Even though there was near total occlusion of celiac trunk at the origin with complete occlusion of inferior mesenteric artery, because of the collateral circulation between Celiac artery and superior mesenteric artery the gangrene was confined to left colon.

This is a rare case of protein C and S deficiency along with hyperhomocysteinemia and thrombocytosis which was managed successfully with early diagnosis and minimal invasive surgical intervention. We recommend a thrombophilic work up in a young patient with arterial splanchnic thrombosis.

CONCLUSION

Patients with protein C and S deficiency who usually develop venous thrombosis can present with multiple splanchnic arterial thrombosis. Thrombophilic work up is recommended in young patients who develop arterial splanchnic thrombosis.

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