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Case Report

Spontaneous Subcutaneous Extracranial Bleed in Cirrhosis- A Rare Phenomenon

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Abstract

Case Report: A Sixty five year old female, a known case of Chronic liver disease with portal vein thrombosis, decompensated, non bleeder on regular follow up presented with sudden onset headache and subcutaneous swelling on left forehead and around left eye and over occipital area. The systemic examination was essentially normal except Per abdominal examination revealed gross splenomegaly. The complete haemogram showed hemoglobin of 14 g/dL, white blood cell count 29, 200/L, microcytic hypochromic anemia with no malaria parasite. The liver function test (LFT) revealed serum bilirubin of 2.6 gm% with conjugated and unconjugated being 1.1 gm% and 1.5 gm% respectively. The transaminases were mildly elevated i.e. AST and ALT were 88 and 62 I.U. respectively. The International normalized ratio (INR) was significant ally deranged to 2.44. The ultrasound abdomen revealed features of Chronic liver disease, Cholelithiasis and Chronic cholecystitis. The Contrast enhanced computed tomography (CECT) scan brain revealed two subcutaneous bleed i.e. in left fronto-parietal area and left occipital area. The MRI abdomen revealed altered liver texture, splenomegaly with portal vein thrombosis. On upper gastro-intestinal endoscopy showed grade -2 esophageal varices for which tablet carvidelol was started. The Fibroscan score was suggestive of cirrhotic pattern i.e. 14 Kpa. The patient was treated with broad spectrum Intravenous antibiotics, vitamin K, Fresh frozen plasma and other supportive therapy. On treatment, patient showed symptomatic improvement, subcutaneous swelling decreased, headache subsided, leucocytosis normalized and INR decreased to 1.3.

Conclusion: In cirrhotic patients, there can be both spontaneous as well as traumatic bleed at various sites of human body. There are typical sites of bleed in cirrhotic patients like Gastro-esophageal, dental area, intracranial but one should also remain vigil regarding atypical sites like extracranial, as in our case.

Keywords: *Chronic Liver Disease; Extracranial Bleed; Coagulopathy; Vitamin K; Fresh Frozen Plasma*

Introduction

Cirrhotic have changed haemostatic's with procoagulant and anticoagulant mechanisms, balance of which could be tilted toward either side, depending upon conditions at different point of time of illness [1]. The coagulation Factors V, VII, IX, X, XI, prothrombin, protein C, and protein S are decreased with simultaneous increase in Factor VIII and von Willebrand factor (vWF) level [2]. Low platelet count, increased nitric oxide and prostacyclin inhibit platelet function (PF), and higher vWF and Factor VIII activity cause platelet aggregation [3]. Low platelet count is caused by splenic sequestration in portal hypertension, decreased hepatic thrombopoietin synthesis, and immune-mediated platelet destruction [4]. Patients with acute liver failure (ALF) have prolonged INR but preserved thrombin generation potential, thus they usually do not bleed. In cirrhosis, there is mild increase in INR and it depicts short-term patient mortality, but not

bleeding risk, due to adequate thrombin generation potential [5]. Acute-on-chronic liver failure (ACLF) has significant mortality rate in view of associated organ failure and progressive clinical course [6]. In ACLF, the dynamics shift from procoagulant to an anticoagulant due to systemic inflammation and endothelial activation [7-9]. The Standard coagulation tests (SCTs) do not exactly measure the risk for bleeding in liver disease [3], leading to incorrect use of blood products which is often ineffective and causes volume overload and transfusion-related acute lung injury (TRALI) [10,11].

Case Report

A sixty five year old female, a known case of chronic liver disease with portal vein thrombosis, decompensated, non bleeder on regular follow up presented with sudden onset headache and subcutaneous swelling on left forehead and around left eye. The swelling around left eye subsided in four days but forehead swelling persisted. There was no recent history of trauma, alternative medications intake, fever, gastrointestinal bleed or Icterus. She was initially treated at private set up and her baseline blood parameters at that point of time revealed leucocytosis (TLC-29,200) but platelets and hemoglobin levels were normal. Later on, after seven days, she reported at our department. Patient was conscious, afebrile, mildly icteric and pedal edema was absent. There was single, soft and compressible subcutaneous swelling present over left forehead of 3 x 2 cm. The systemic examination was normal except per abdomen examination which revealed splenomegaly. The patient hemoglobin was 14 g/dL, total leukocyte count 22,300/L and platelet levels were in normal range. The serum bilirubin was 2.6 gm% with conjugated and unconjugated being 1.1 gm% and 1.5 gm% respectively. The transaminases were mildly elevated i.e. AST and ALT were 88 and 62 I.U. respectively. The International normalized ratio (INR) was increased to 2.44. The ultrasound abdomen revealed features of Chronic liver disease, Cholelithiasis and Chronic cholecystitis. The kidney function test, blood sugar, serum electrolytes, thyroid and lipid profile, viral screen, autoimmune profile, serum copper and ceruloplasmin level were normal. The Contrast enhanced computed tomography (CECT) scan brain revealed two Subcutaneous bleed i.e. in left fronto-parietal area and left occipital area. The MRI abdomen revealed altered liver texture, splenomegaly with portal vein thrombosis. On upper gastro-intestinal endoscopy, grade -2 esophageal varices were seen and for it tablet carvidelol was started. The Fibroscan score was 14 Kpa i.e. suggestive of cirrhotic pattern. The patient was treated with antibiotics, Vitamin K, Fresh frozen plasma and other supportive therapy. On treatment, patient showed symptomatic improvement, subcutaneous swelling and headache slowly decreased and ultimately got resolved, leucocytosis normalized and INR decreased to 1.3.

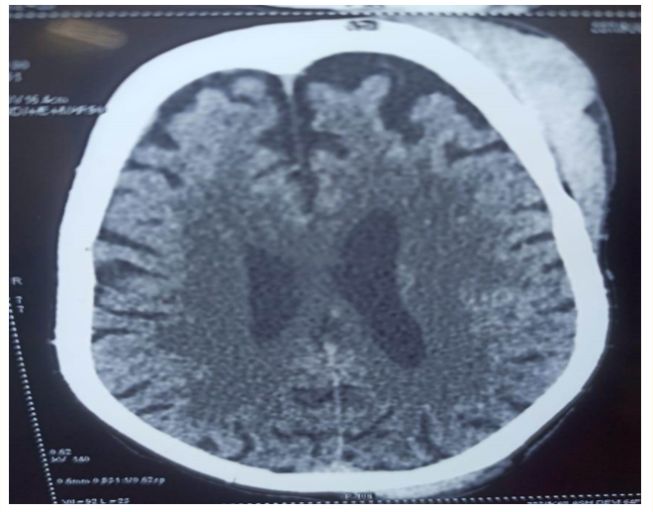

Figure 1: *CECT Scan Brain Showing Fronto-Parietal and Occipital Bleed.*

Figure 2: *Patient Photograph Showing Subcutaneous Fronto-Parietal Bleed.*

Discussion

Many studies in past have clearly highlighted role of liver disease in causing Intracerebral hemorrhage [12-18] but there is dirth of reports on spontaneous extracranial bleed in cirrhotic patients. Gastroesophageal variceal bleeding is life threatening complication of cirrhosis, thus various preventive and treatment modalities have been developed for the same. In end-stage liver disease, other bleeding sites include gums and nose but soft tissue bleeding and hemartrosis are rare. In cirrhotic, most common thrombotic complication is portal vein thrombosis followed by venous thromboembolism, especially in decompensated stage. The factors II, IX, XI, and XII, are reduced and their level depend upon severity of liver disease. Many studies have shown that hepatic injury increases plasminogen activator inhibitor (PAI-1) expression [19,20]. In comparison to PAI-1, tissue plasminogen activator is increased both due to reduced hepatic clearance and to enhanced release [21], leading to hyperfibrinolytic state in cirrhosis [22]. Some studies have shown that actually fibrinolysis is not increased in cirrhotic, with a balanced reduction of both pro- as well as anti-fibrinolytic agents [23], and a lack of association between TAFI reduction and actual hyperfibrinolysis [24,25]. The increased levels of D-dimer are consequence of the activation of the coagulation cascade, which might accumulate in the presence of diminished hepatic clearance [26-28]. The Factor XIII levels of < 50% significantly correlated with an increased risk of severe upper gastrointestinal bleeding and mortality [29]. The reduced FXIII activity in addition to multiple coagulation and hemostatic defects, increase the risk of hemorrhage [29,30]. Regarding primary hemostasis, chronic liver disease is characterized by a variable degree of thrombocytopenia due to increased platelet destruction, increased splenic and/or hepatic sequestration, and to reduced levels of thrombopoietin. Both platelet number and platelet function are deranged due to defective thromboxane A2 synthesis, storage pool deficiency and abnormalities of the platelet glycoprotein Ib [31-34]. In cirrhosis, minor bleeding occurs due to platelet defects and not due to deranged coagulation parameters. In variceal bleeding, local factors, portal pressure and severity of liver disease play important role, whereas hemostasis defects cause recurrent epistaxis, gingivorrhagia, purpuric skin lesions, menometrorrhagia, and excessive bleeding after dental extractions. The prevalence of coagulation-related complications in cirrhotic like Intracerebral bleeding, deep muscle bleeding, and hemartrosis is same as in general population.

In our patient, the presentation was of acute onset and was associated with seepage of blood subcutaneously over bilateral eyes and whole face also which was frightening for patient as well as family members. We were able to pinpoint main reason for above symptoms,

coagulopathy as evidenced by significant ally deranged INR and complete recovery occurred on correction of coagulopathy. The MRI abdomen never revealed any evidence of hepatocellular carcinoma. Whether this coagulopathy was precipitated due to Cholelithiasis induced cholecystitis, is matter for further research and cannot be ruled out. Once a cirrhotic patient presents with bleed at any site, then treating team should remain cautious for bleed from other sites of body.

Conclusion

In cirrhotic patients, there can be both spontaneous as well as traumatic bleed at various sites of human body. There are typical sites of bleed in cirrhotic patients like Gastro-esophageal, dental area, intracranial but one should also be vigilant regarding atypical sites like extracranial one, as in present case.

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