中文題目：C 蛋白缺乏症之病人產生肝門腸繫膜靜脈栓塞之病例報告

英文題目：Portomesenteric Venous Thrombosis in a Patient with Protein C Deficiency

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Introduction: Mesenteric venous thrombosis (MVT) was first described as a cause of intestinal gangrene by Elliot in 1895 and further characterized by Warren and Eberhard in 1935. The incidence of MVT comprises 5% to 15% of all cases of acute mesenteric ischemia. Waren and Eberhard reported mortality of 34% in patients with venous mesenteric venous thrombosis after bowel resection in 1935. After merely 75 years, Brunaud et al quotes mortality rates of 29-39% for surgical treatment and 13-19% for “nonoperative management. The decline of mortality rate is contributed to earlier diagnosis with imaging studies and the use of anticoagulation. Mesenteric venous thrombosis can be classified into primary and secondary depends on its etiology. Primary thrombosis is mainly due to hereditary or acquired hypercoagulation disorder, such as deficinecy of protein C, protein S, antithrombin III, and factor V Leiden. However, factor V Leiden is rare in Chinese population. Secondary MVT may follow hypercoagulable states, portal venous stasis and hypertension, previous sclerotherapy for varices, intra-abdominal infection, inflammation or malignancy, use of oral contraceptive pills and splenectomy. We would like to present a patient who was previously diagnosed with protein C deficiency presents with portomesenteric venous thrombosis.

Case Presentation: A 50-year-old male with past medical history of left lower extremity deep vein thrombosis, pulmonary embolism, Protein C (22.5%) deficiencies with poor Warfarin compliance, presented with dull persistent abdominal pain and fullness started about 2 weeks before his admission. Initially patient described pain located at epigastric region that later shifted to the right upper quadrant. Lying on the right side worsened the pain. Pain improved slightly with lying on the left side. Pain was not associated with food intake. Patient denied pain radiation, fever, chill, nausea, vomiting, diarrhea, tarry stool or maroon colored stool. Patient initially went to other gastroenterologist for the abdominal pain. Duodenal ulcer was diagnosed after esophagoduodenoscopy (EGD) examination. Proton pump inhibitor was prescribed. However, abdominal pain was not relieved by the medication. Hence patient came to our emergency department to seek medical attention. On physical examination, patient had mild tenderness at epigastric region upon deep palpation. Abdomen was soft but distended. Hypoactive bowel sound was noted. Due to patient’s prior thormbophilic history, venous thrombosis was suspected. Contrast enhanced CT scan of abdomen revealed extensive thrombosis of the mesenteric branches and main trunk of the superior mesenteric vein, splenic, main and intrahepatic portal veins. Biochemical results and coagulation studies were within normal limits. Leukocytosis was noted. Patient was started on heparin. His abdominal pain improved on the second day of anticoagulant therapy. He had a complete recovery from abdominal pain after 11 days of treatment. Patient was started on the oral anticoagulant therapy and subsequently discharged.

Discussion: Diagnosis of MVT by history alone is difficult due to its vague abdominal symptoms. The most common clinical presentation was diffuse abdominal pain out of proportion to physical examination (83%-90%), followed by

nausea (54%) and vomiting (42%-77%), with the signs of peritonitis (32%), leukocytosis (49%), elevated serum lactate (28%), fever (25%), and sometimes upper (28%) or lower gastrointestinal bleeding (23%)

Its diagnosis requires high index of clinical suspicion and imaging CT is a good initial imaging study that has better sensitivity than ultrasound. Magnetic resonance angiography (MRA) is a nice adjunct because it does not require contrast load and it is especially useful when surgical clips or other artifacts obscure the field of interest.

MVT could be acute, subacute, or chronic. Acute thrombosis is associated with a definite risk of bowel infarction and peritonitis. In patients with subacute mesenteric venous thrombosis, abdominal pain is prominent, but neither bowel infarction nor variceal hemorrhage is likely. Patients with chronic mensenteric venous thrombosis do not have pain and have extensive venous collateral circulation.

Thrombophilic factors such as antithrombin, protein C and protein S have not been well studied in Asian population. A study conducted in a tertiary center in Taiwan evaluating prevalence of antithrombin III, protein C and protein S defieincies and factor V Leiden mutation in patients with venous thrombophilia. Total of 85 patients were evaluated. None of those patients have Factor V Leiden mutation. In this study, protein C and protein S deficiency rates of 17.2% and 33.6% were reported respectively. The authors concluded that protein C and S deficiencies were the most important risk factors for thrombosis in Chinese venous thrombophilic patients.

In our case, the patient most likely had subacute mesenteric venous thrombosis. Protein C deficiency predisposed him to the thrombosis event. The prompt diagnosis made in our emergency department was due to patient's prior history of deep vein thrombosis and pulmonary embolism and history or protein C deficiency leading to the high clinical suspicion of the mesenteric thrombosis. CT scan confirmed the diagnosis. Intravenous heparin was started immediately after diagnosis was established. Fortunately, this patient did not go down the route of bowel ischemia. However, life-long anticoagulation is warranted for this patient.