coagulation disorders, and patients on anticoagulants and antiplatelets. Etiologies proposed for this uncommon condition include suboptimal hemostasis, emetogenic, traumatic, aortic disease related and idiopathic. 36% patients present with the triad of retrosternal chest pain (due to esophageal distension by expanding hematoma), hematemesis and dysphagia or odynophagia, with 80% reporting at least 2 of these symptoms.[3] Dyspnoea is unusual but may occur from tracheal compression by the hematoma. In our case, the use of anticoagulants was the predisposing factor, with hematoma occurring either spontaneously or secondary to nasogastric tube insertion. Repeated suctioning of nasogastric tube may produce negative pressure to the esophageal mucosa, leading to submucosal injury and consequent hematoma.[3]

The most common site for hematoma is the distal esophagus (83%) followed by the middle esophagus (78%) and proximal esophagus (27%). Contrast swallow (barium or gastrograffin) typically shows a "double barrel esophagus" or "mucosal stripe" sign, and CT scan with intravenous contrast shows thickened esophageal wall with a non-enhancing, high-attenuation intramural mass causing luminal compromise.[5] Endoscopy typically shows an intraluminal protruded mass covered by fluctuant purple or dark-colored mucosa running along the long axis of esophagus, that may be intact or show a laceration. Endoscopic ultrasound shows a uniform low echo mass in the submucosal layer, and can also be useful to assess other submucosal lesions and mediastinal structures.Treatment is mainly conservative with reversal of anticoagulation, parenteral nutrition and analgesics. 96% hematomas resolve spontaneously over a period of 1 to 3 weeks1, with only 2.3% deaths reported.

Hence, intramural hematoma of the esophagus is an unusual condition resulting from heamorrhage within the esophageal wall with a threatening presentation. It is important to differentiate it from other esophageal conditions (particularly malignancies) due to its benign course and lack of need of therapeutic intervention. Recognition of this condition by clinical presentation, imaging and endoscopic finding should prevent clinicians from inappropriate diagnosis and treatment.

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Acute Cytopenias after Liver Transplantation: A Case of Thrombotic Thrombocytopenic Purpura

Thrombotic thrombocytopenic purpura (TTP) is a rare disease and diagnosis requires high index of suspicion. Timely diagnosis is very important as mortality is very

high in untreated cases. We describe a case of living donor liver transplantation that had TTP, probably related to cytomegalovirus virus infection or tacrolimus.

Case Report

A 33 year old male underwent living donor liver transplantation (LDLT) for decompensated alcoholic cirrhosis, Child's score 13/15, MELD score 32 before liver transplantation. He was diagnosed to have cytomegalovirus (CMV) viremia (1890 copies/ml) on day 21 after LDLT. He complained odynophagia, a gastroscopy was done which revealed esophageal ulcers, biopsy and immunohistochemistry was positive for CMV. He received injection ganciclovir initially followed by oral valganciclovir. Initially he was given tacrolimus, mycophenolate and steroids, it was modified in view of acute kidney injury to low dose tacrolimus (target trough level 4-6) and everolimus was added. He was admitted on post op day 100 with complaints of abdominal pain. His investigations revealed stable liver function tests; however, he had progressive thrombocytopenia **(Figure 1)** with anemia. A hematology opinion was taken. His work-up revealed normal iron profile, B12 and folate levels, raised lactate dehydrogenase (4232 U/L, normal up to 618), reticulocyte count 2.39%, peripheral smear showed 3.2% schistocytes and platelet count of 18000 cu/mm, serum ADAMTS13 activity was below normal. A diagnosis of thrombotic thrombocytopenic purpura (TTP) was made and tacrolimus was stopped. He was started on plasma exchange and his platelet count started to increase as shown in figure 1. He also had psychosis, which was improved with plasma exchange. He needed a total of 9 sessions of plasma exchange, mean plasma volume processed was 5.8 L (1.6 plasma volume), mean 13.7 fresh frozen plasma units used, average duration of plasma exchange was 183 minutes. At present his platelet count is 329000/cmm after 48 days of last session of plasmapheresies.

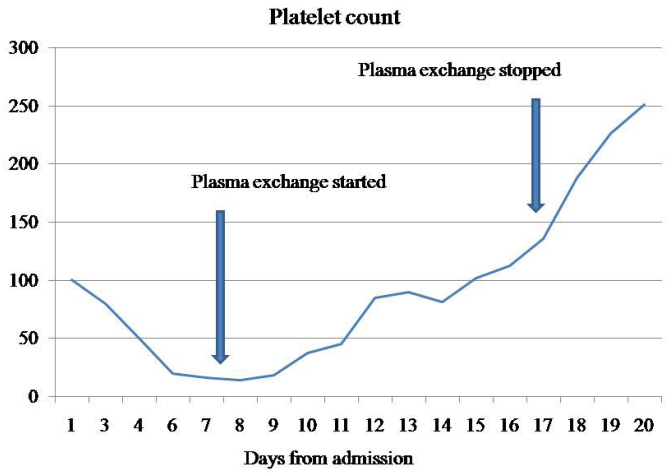

Figure 1: Platelet counts before and after plasma exchange.

Discussion

Thrombotic microangiopathies are several diseases characterized by microangiopathic hemolytic anemia, thrombocytopenia, and variable organ involvement. Common thrombotic microangiopathies are TTP, Hemolytic-uremic syndrome and pregnancy related HELLP (pre-eclampsia/hemolysis, elevated liver enzyme levels, and low platelet) syndrome.[1] TTP is very rare (< six cases per million per year) and timely diagnosis is important as mortality is 90% in untreated cases.[2] Differential diagnosis include autoimmune hemolysis/ Evans syndrome, disseminated intravascular coagulation, pregnancy-associated disorders (HELLP and eclampsia), hemolytic uremic syndrome, certain drugs and infections (generally viral including CMV).[1] TTP is caused by deficiency of von Willebrand factor (vWF) cleaving protein ADAMTS13. This deficiency may be congenital or acquired (by antibodies against ADAMTS13). Acute idiopathic TTP is the most common form of TTP.1 The ultra-large multimers of vWF released from endothelium are not cleaved in absence of ADAMTS13 and it causes spontaneous platelet aggregates at places of high shear (like microvasculature of the brain, heart and kidneys) which results in ischemia and thrombocytopenia due to consumption of platelets in platelet-rich thrombi. TTP should be treated as medical emergency as early death may occur after diagnosis.[1] In a retrospective series of 18 thrombotic microangipathy (number of TTP not given), the authors found that overall rate of thrombotic microangiopathy after LDLT was 3.34 cases per 100 patient-years. Eight of these 18 patients (44%) died before improvement with plasmapheresis.[3] Treatment consists of

plasma exchange. CMV and tacrolimus have been shown to be associated of TTP.[4,5] TTP should be suspected in presence of microangiopathic hemolytic anemia and thrombocytopenia in absence of other identifiable causes and a high index of suspicion is necessary for timely diagnosis and management.

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Double Papilla of Vater

Abnormal rotation and recanalization during embryologic development can lead to variation in pancreatico-biliary ductal anatomy. Usually, the bile duct and pancreatic duct join together at the level of duodenum to form papilla of Vater. During embryogenesis, complete non-union of distal bile duct and ventral pancreatic duct give rise to separate orifice of these two ducts as double papilla. It is a very rare condition and only few cases have been reported in literature.[1-4]

Case Report

A 60 years female presented with right upper abdominal pain 9 months back and on evaluation was found to have a large hydatid cyst. She underwent exploratory laparotomy and enucleation of the hydatid cyst in another hospital. Post operatively, patient developed bilioma which was drained by percutaneous catheter drainage (PCD). Patient presented to us with persistent drainage of bile through the PCD. On evaluation hemogram and biochemical parameters were as follows: hemoglobin-12.2g/dl, total leucocyte count (TLC) - 9900/mm^3, platelet count-173x10^3/mm^3, bilirubin-0.7mg/dl, AST-21 U/L, ALT-15 U/L, APL-182 U/L. Her renal function and coagulogram were normal. In view of persistent bile leak, the patient was taken up for endoscopic retrograde cholangiopancreatography (ERCP), and during the procedure two separate papillary openings were identified **(Figure 1)**. The cranial orifice was located at 11 o'clock position and the caudal orifice was at 7 o'clock position. Selective cannulation of the cranial orifice (biliary orifice) was done **(Figure 2)** and contrast injected. Following that, only common bile duct was opacified and no opacification of the main pancreatic duct was observed. Cholangiogram revealed leakage of the contrast from the right ductal system **(Figure 3)**. A 7 Fr 7 cm double pigtailstent was placed in the biliary system and following that patient had marked improvement in her symptoms with reduction in the drain output.

Discussion

The apertures of pancreatic and bile ducts opening into the duodenum is called the ampulla of Vater. It is