Hemophilia A Presenting as Hemosuccus Pancreaticus

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Abstract

Hemosuccus pancreaticus (HP) is a rare cause of upper gastrointestinal bleeding. Case of haemophilia A presenting as HP has not been reported. We describe the case of a 50 year old patient, who presented with complaints of melena, he was initially diagnosed as HP on upper gastrointestinal (GI) endoscopy and following extensive workup patient also diagnosed with mild hemophilia A. Factor VIII injections failed to control the bleeding, which was treated by angiographic embolization of superior pancreatic duodenal artery (SDPA) and posterior inferior pancreatic duodenal artery (IPDA).

ABBREVIATIONS

HP: Hemosuccus Pancreaticus; GI: Gastrointestinal; SDPA: Superior Pancreaticoduodenal Artery; IDPA: Inferior Pancreaticoduodenal Artery; CECT: Contrast Enhanced Computed Tomography

INTRODUCTION

Hemophilia A is an X-linked recessive hemorrhagic disease due to mutations in the F8 gene. It is classified as severe (<1%), moderate (1-5%), mild (6-30%), based on the residual activity of Factor VIII levels [1]. The disease is characterized by bleeding into the joints, soft tissues, and muscles after minor trauma or even spontaneously. HP is a rare and potentially life-threatening clinical entity and is described as bleeding from the pancreatic duct into the gastrointestinal tract via ampulla of Vater. It is most often caused by chronic pancreatitis, pancreatic pseudo cysts and pancreatic tumors due to rupture of a pseudo aneurysm of the peripancreatic vessel into the pancreatic duct. We present a rare case of hemophilia A presenting as HP.

CASE PRESENTATION

A 50-year-old male patient presented with a history of melena of 3 days duration to the medicine casualty. There was no history of pain abdomen and hematemesis. He had significant past history of bleeding episodes and blood transfusions, during tooth extraction and surgery for fracture neck of the right femur. There was no past history of melena and pain abdomen. There was a family history of hemophilia in younger brother. He was not an alcoholic and never consumed tobacco in any forms. There was a family history of hemophilia in younger brother. He was not an alcoholic and never consumed tobacco in any forms. On examination, he had pallor, tachycardia of 130 per min and blood pressure of 100/60 mmHg. His abdominal examination did not reveal any abnormal findings. Nasogastric aspiration showed altered blood. His hemoglobin had dropped to 4.8g/dl.

His bleeding time was 03 mins 15 secs, clotting time was 04 mins 51 secs, prothrombin time 12.50 secs, INR 0.85 and activated partial thromboplastin time 28.3 secs. Factor VIII activity was 22.5%. An emergency upper GI endoscopy was performed which revealed altered blood in the stomach, fresh blood in the duodenum and a large adherent clot at the ampulla of Vater which were features suggestive of HP. His serum amylase was 122 IU/L (reference range: 25-140 IU/L) and serum lipase was 36.06 IU/L (reference range: 0-60 IU/L). A contrast enhanced computerized tomography scan (CECT) of the abdomen revealed normal study. Initially, the patient was managed conservatively with factor VIII infusions to maintain levels more than 80% and packed cell transfusions. In spite of this, the patient’s hemoglobin was not increasing and sustained tachycardia was present which were suggestive of ongoing blood loss. Then the patient was shifted to angiography and access was gained through the right femoral artery, aortogram and selective injection of the celiac artery and superior mesenteric artery done. An irregular caliber of SDPA and posterior IPDA noted. Selective cannulation and embolization of SDPA and posterior IPDA performed and the procedure was uneventful. A total of 14 units packed cell transfusions were done. In spite of successful embolization Factor VIII injections were continued for 7 days post procedure followed by maintenance dose for another week. Melena resolved 1-week post procedure and patient’s hemoglobin was improving. He was discharged after 1 week with advice for regular follow up.

One week later the patient presented again with melena of one-day duration. His hemoglobin had dropped from 9 g/dl to 7 g/dl. This bleeding was less compared to the first episode. All investigations were repeated, including upper GI endoscopy and it was normal. A CT abdominal angiography scan doesn’t reveal any active bleeding site. The patient recovered from the symptom...
of melena after 3 days of factor VIII injections and 2 units packed cell transfusion. During this episode also factor VIII injections were given for 14 days.

A CECT abdomen with angiography was repeated after 1 month of follow-up which revealed poorly defined heterogeneously enhancing a hypodense area of size 21*43mm in the neck and proximal body of the pancreas suggestive of post pancreatitis changes. Hyper dense contents in the SPDA and IPDA seen likely secondary to embolization. The patient is on regular follow-up and symptom-free for the past 12 months.

DISCUSSION

Gastrointestinal bleeding in hemophilia patients may appear in between 10% and 25% of the cases, and up to 85% of them are due to peptic ulcers [2,3] (Figure 1,2). These Figures have grown over the last years due to the introduction of nonsteroidal anti-inflammatory drugs in the treatment of chronic hemophilic arthropathy [4]. Cases of spontaneous hemobilia have been reported in hemophilia patients [5].

HP is the least frequent cause of upper GI (1/1500) bleeding and is most often caused by chronic pancreatitis, pancreatic pseudo cysts, or pancreatic tumors [6]. In 80% of cases, HP is a complication of pancreatitis, typically from bleeding pseudoaneurysms into a pancreatic pseudo cyst which communicates with the main pancreatic duct [7]. It presents as intermittent episodes of upper GI bleeding and epigastric pain. Amylase levels were also raised during the acute episode in most of the patients.

The first step in the management was an assessment of severity of blood loss and making the patient hemodynamically stable. The initial approach was same as for the other cases of upper GI bleeding, beginning with upper GI endoscopy. Active bleeding can be visualized through upper GI endoscopy in only 30% of the cases of HP making the diagnosis difficult and it is also helpful to rule out the other possible causes like peptic ulcer, gastritis etc [8]. CT scan is an important diagnostic modality which may help for early therapeutic intervention [9] and can also demonstrate any pancreatic pathology and pseudo aneurysms. Angiography remains the gold standard for diagnosis and therapy. It identifies the causative artery and helps to delineate arterial anatomy for therapeutic intervention. The sensitivity of angiography is usually greater than 90% [9]. If the source of hemorrhage is found by angiography then interventional radiographic procedures are the first choice for initial management with immediate good results in 79-100% of the cases and an overall success rate of 67% [9].

The recurrence rate of bleeding after angiographic embolization is around 30% [9]. Surgery is advised following failed angiographic intervention procedure. The surgical procedure was deferred in this patient because of the absence of definitive arterial abnormality.

CONCLUSION

In conclusion coagulation disorders may also present as HP in the absence of pancreatic abnormalities and pseudo aneurysms of peripancreatic arteries.
REFERENCES


