

hepatic artery pseudo-aneurysm following trauma can be safely observed for 2 to 4 weeks and if there are no signs of thrombosis after 4 weeks, prophylactic embolisation may be considered.

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Idiopathic Spontaneous Intra-peritoneal Haemorrhage - a rare initial presentation of Haemophilia B

Introduction

We describe a case of idiopathic spontaneous intra-peritoneal haemorrhage (ISIH) as the initial presentation in a patient with Haemophilia B for highlighting the associated challenges in pre-operative evaluation and management of ISIH.

Case Report

A 39 year old male presented to our Emergency Department with a one-day history of acute abdominal pain and vomiting. Abdominal examination revealed generalised tenderness with guarding. An emergency laparotomy was performed on a clinical diagnosis of peritonitis and a three-litre haemoperitoneum was drained but no source of bleeding was evident. Baseline coagulation work up revealed a mildly deranged aPTT of 35.6 seconds (Normal: 25-34.8s). A CT angiogram done on the fourth post-operative day was normal. Post-operative recovery was uneventful and he was discharged on the eighth post-operative day. He presented once again to the Emergency department with a similar history on the sixteenth post-operative day. His PCV was 17% and his aPTT was 50.4 seconds; corrected to 30.4s with ½ patient and ½ control serum. CT angiogram revealed a haemoperitoneum with a large haematoma along the hepatico-duodenal ligament. A digital subtraction angiogram (DSA) showed no evidence of bleeding. Exploratory repeat laparotomy for the 1.3 litre haemoperitoneum once again did not reveal the source of the bleed. The post-operative period was uneventful. A haematology workup two weeks after operation revealed low factor IX levels (14.7%) suggestive of mild Haemophilia B.

Discussion

ISIH is an uncommon surgical emergency (previously known as abdominal apoplexy) with the commonest cause being trauma. Although it was first described by Barber

in 1909¹, Green and Powers² coined the term “abdominal apoplexy” in 1931 for describing haemorrhage arising from the smaller abdominal arteries or veins. After extensive evaluation of up to 30% of patients with ISIH, no cause was identifiable.³

The aetio-pathogenesis of ISIH remains obscure. Arterial bleeds often arise from small aneurysms especially from the branching points of the abdominal aorta and from its smaller branches.³ Venous bleeds in the setting of portal hypertension present as an ooze from retroperitoneal veins.^{3,4} Histopathological analysis of post-mortem specimens have revealed disruption of the layers of the arterial wall leading to its weakening and rupture during times of rise in intra arterial pressure.⁵

ISIH is more likely in the fifth to sixth decades of life with a male preponderance.³ The clinical presentation of intra peritoneal haemorrhage is variable⁶ ranging from a mild initial phase with abdominal pain to a final phase of haemoperitoneum and hypovolemic shock. The diagnosis is usually made intra-operatively in an unstable patient. Contrast enhanced computerised tomography (CECT) scan of the abdomen is the investigation of choice in stable patients. The ultrasonography (USG) of the abdomen is useful in an unstable patient and when a pelvic source of bleeding is suspected, but is unreliable in detecting solid-organ-bleeds.^{7,8} The diagnostic role of magnetic resonance imaging (MRI) and DSA in ISIH is unclear. In the stable patient, DSA is indeed a therapeutic option as the bleeding vessel may be amenable to embolization. In the unstable patient, surgical management is the treatment of choice.

To summarise, ISIH is a poorly understood, diagnosis of exclusion and encompasses all spontaneous intra-abdominal haemorrhage of unknown etiology, presenting diagnostic and therapeutic challenges. Early aetio-pathogenic diagnosis and appropriate management helps avoid mortality, which otherwise approaches 100%.

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