International Surgery Journal
*Beedubailu R et al. Int Surg J. 2021 Jun;8(6):1913-1916*
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20212291

**Case Report**

# Large retroperitoneal hematoma in a patient with haemophilia B

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**Received:** 28 March 2021
**Revised:** 12 May 2021
**Accepted:** 14 May 2021

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## ABSTRACT

A spontaneous retroperitoneal hematoma is very rare and quite a challenging entity, more so when its presentation mimics other more common causes of acute abdomen, as in this interesting case. Here, patient presented with acute abdomen with polyarthralgia with flank ecchymosis (Grey Turner's sign) with flank mass which lead us in the direction of acute pancreatitis. However, past history of recurrent bleeding episodes and positive family history along with blood investigations revealing an isolated prolongation of activated partial thromboplastin time tickled our brains with the differentials of coagulation disorders. Computed tomography showed large retroperitoneal hematoma. After initial resuscitation in the emergency room he was managed conservatively with fresh frozen plasma, cryoprecipitate and blood transfusions. On detailed evaluation, coagulation workup showed deficient coagulation factor IX levels leading us to the diagnosis of haemophilia B. Now, the patient is doing good on regular follow up.

**Keywords:** Retroperitoneal hematoma, Spontaneous retroperitoneal hematoma, Acute pancreatitis, Bleeding disorders, Haemophilia B, Case report

## INTRODUCTION

Acute pancreatitis is an acute inflammatory process of the pancreas. Gallstone pancreatitis and ethanol abuse account for 70% to 80% of cases.[1] Abdominal blunt trauma, drugs like sulfonamides, metronidazole, erythromycin, tetracyclines, metabolic factors like hypertriglyceridemia, hypercalcaemia and procedures endoscopic retrograde cholangiopancreatography are other causes. Most patients with acute pancreatitis have acute onset of persistent, severe epigastric abdominal pain. The physical examination of the abdomen varies according to the severity. With mild pancreatitis, abdomen may be normal or reveal mild epigastric tenderness. Significant abdominal distension associated with generalised guarding is present in severe pancreatitis. Rare findings include flank and periumbilical ecchymosis (Grey Turner and Cullen signs).[2] The diagnosis is established with the help of clinical findings raised amylase and lipase levels. Regardless of the cause or severity, the cornerstone of the treatment is aggressive fluid resuscitation with isotonic crystalloid solution.[2]

Hemophilia B (also known as christmas disease) is a X-linked recessive disorder where there is deficiency of coagulation factor IX. Clinical manifestations of hemophilia relate to bleeding from impaired hemostasis, sequelae from bleeding, or complications of coagulation factor infusion. Patients can present with epistaxis, oral bleed, hemarthrosis, intramuscular hematomas, hematuria or rarely retroperitoneal hematomas usually following trauma.[3]

Retroperitoneal hematoma usually occurs after blunt or penetrating injuries, in the setting of abdominal aortic or visceral artery aneurysms, after acute or chronic

anticoagulation of fibrinolytic therapy or rarely in patients with bleeding diathesis.

The management ranges from observation to mandatory retroperitoneal exploration.[2] However, we managed this case of huge retroperitoneal hematoma with haemophilia B conservatively.

## CASE REPORT

A 31-years-old male presented with complaints of multiple joint pain since 7 days and pain abdomen since 5 days was referred from a hospital in Goa. The pain was pricking type, intermittent in nature localised in the right lumbar region radiating to back and relieving with analgesics.

The patient gave a past history of recurrent knee swellings, easy bruisability and nose bleeding episodes since childhood.

He had also undergone laparotomy following a road traffic accident 15 years ago.

Complaints of nose bleed and recurrent knee swelling were also there in many male members of his family on his maternal side; as depicted in the pedigree (Figure 1).

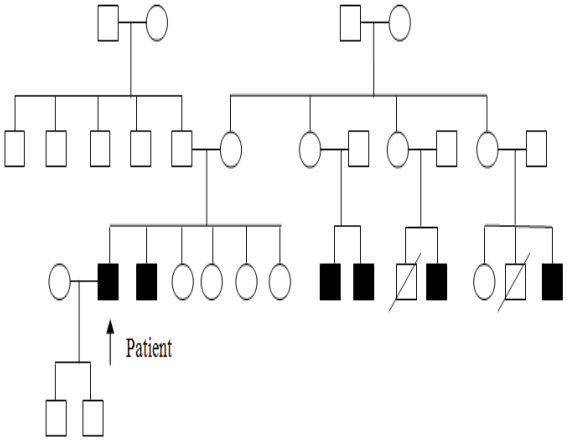


**Figure 1: Pedigree chart of the patient's family.**

On examination he had a GCS of 15/15, was pale, had tachycardia of 110 bpm and a BP of 90/60 mmHg. On abdominal examination he had a distended abdomen with bluish discolouration of skin over right side of the back extending into the right iliac and right lumbar regions and a midline vertical scar (Figure 2). On palpation abdomen was tense with tenderness in the right lumbar and umbilical regions with no guarding or rigidity.

Ill-defined firm to hard mass extending from right lumbar to iliac and umbilical regions not moving with respiration was also palpable.

He also had bilateral target knee joints (Figure 3). Blood investigation showed a low Hb of 6.9 g%, total leucocyte count of 13,600 cells/mm$^3$ and elevated activated partial thromboplastin time of 44.6 seconds with normal prothrombin time of 16.2 seconds and INR of 1.33. Serum amylase was 790 units/l and S. lipase of 9975 units/l.

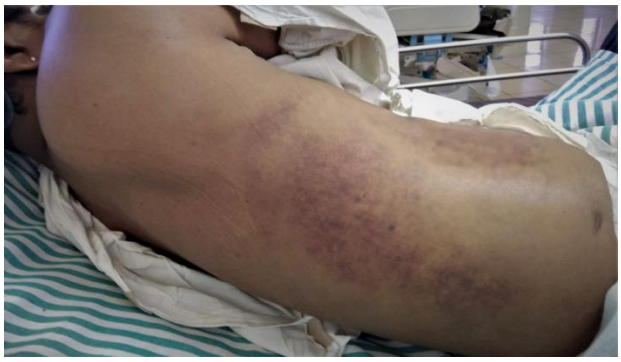

**Figure 2: Grey Turner's sign.**

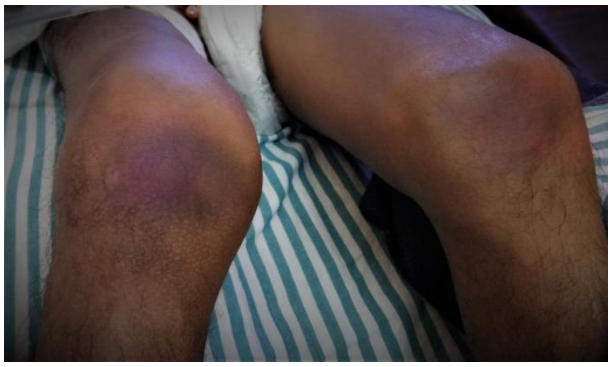

**Figure 3: Target joints.**

CT abdomen and pelvis was suggestive of a large retroperitoneal hematoma with hemoperitoneum and normal pancreas (Figure 4).

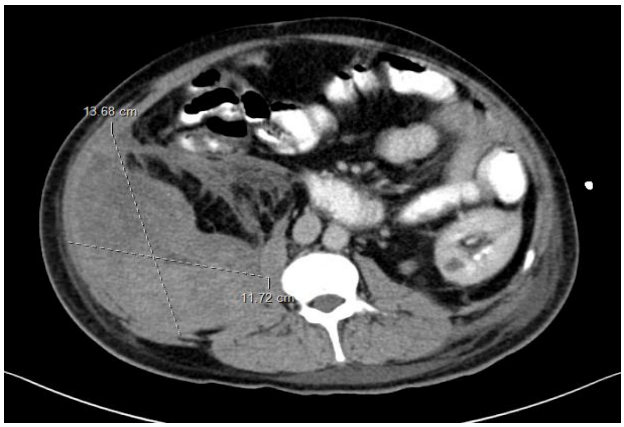


**Figure 4: CT abdomen showing retroperitoneal hematoma.**

On admission he was resuscitated in the emergency room. Hematologist's opinion sought and with a differential diagnosis of VWD and hemophilia, in order to maintain his hemodynamic stability 28 units of fresh frozen plasma, 6 units of cryoprecipitate and 2 units of pRBC transfused with serial APTT monitoring. He improved with conservative management with no further drop in hemoglobin and corrected coagulation profile.

Workup for coagulation disorder was done and factor IX levels was found to be 3.1% with normal factor 11, 12, vWF, fibrinogen, thrombin levels; hence diagnosed to have hemophilia B (christmas disease): moderate.

## DISCUSSION

Hemophilia is an inherited bleeding disorder due to deficiencies of coagulation factor VIII (haemophilia A), factor IX (haemophilia B) or factor XI (hemophilia C).

Hemophilia B (christmas disease) an X-linked recessive disorder is the second most common type of hemophilia where there is deficiency of factor IX [factor 9 (F9)]. It occurs in approximately 1 in 30,000 to 1 in 60,000 live male births, in India.[3] Approximately one-third to half have severe disease (ie, factor IX activity <1 percent of normal).

Mild hemophilia which is defined as a factor activity level >5 percent of normal and <40 percent of normal (≥0.05 and <0.40 IU/ml). Moderate hemophilia is defined as a factor activity level ≥1 percent of normal and ≤5 percent of normal (≥0.01 and ≤0.05 IU/ml). Severe hemophilia is defined as <1 percent factor activity, which corresponds to <0.01 IU/ml.[4]

Hemophilia is characterized by a prolonged activated partial thromboplastin time (aPTT). However, the aPTT may be normal in individuals with milder factor deficiencies (eg, factor activity level >15 percent), especially in hemophilia B (factor IX deficiency), where even individuals with moderate disease may have a normal aPTT. The platelet count and prothrombin time (PT) are normal in hemophilia.[3,4]

Measurement of the factor activity level (factor IX in hemophilia B) shows a reduced level compared with normal controls (generally <40 percent).[3]

Management depends on the severity of the clinical presentation. It includes intravenous infusion of factor IX and/or blood transfusions.[6]

The retroperitoneum is a large space bounded anteriorly by the posterior parietal peritoneum, posteriorly by the transversalis fascia, and superiorly by the diaphragm. Inferiorly, it extends to the level of the pelvic brim. Retroperitoneal hematoma is classified into three zones: (A) Central region (zone I): There is diaphragm on this region, medial borders of psoas muscles are at its sides and the pelvis is under it. Central retroperitoneal hematoma is caused by injuries of abdominal aorta, vena cava, superior mesenteric artery, renal vessels, common iliac vessels and veins, portal vein, pancreas and duodenum. (B) Lateral hematoma (zone II): it is a retroperitoneal hematoma region lying laterally to the psoas muscles, above the iliac wings and under the diaphragm and it is caused by the injury of kidney parenchyma, vessels feeding the left and the right colon and the injuries of the muscle vessel. (C) Pelvic hematoma (zone III): it is the presence of hematoma or fluid in the retroperitoneal region limited with the dome of the bladder at the front, sacral promontory at the rear and iliac wings at both sides.[7]

Spontaneous retroperitoneal hematoma (SRH) is a rare and life-threatening condition, owing to its anatomical location, leading to 10% rate of misdiagnosis.[8] It is defined as bleeding into the retroperitoneal space without associated trauma or iatrogenic manipulation.[11]The classic presentation of SRH is described by Lenk's triad: flank pain, palpable flank mass and signs of hypovolemia/shock. SRH has been associated with the use of anticoagulants and antiplatelets, however in up to a third of patients the bleeding is not related to an obvious or known anticoagulation state. Typically, in non-anticoagulated patients the etiology of the bleeding is secondary to tumors, vascular malformations, small aneurysms and vasculopathies in general.[8] The investigation of choice is contrast enhanced CT scan, which helps to know the site, size and helps rule out the causes.[7]

Management depends on the cause of hematoma. However, it starts with resuscitation aiming at restoration of circulating volume with packed red blood cells (PRBCs), fresh frozen plasma, units of platelets, colloids. Interventional radiology with embolisation of feeding vessel is an option in splanchnic aneurysms.[9] Surgery is reserved for those who do not respond to these and have concurrent surgical conditions. Up until the 1960s, the only surgeries individuals with hemophilia underwent were emergency procedures, due to the high mortality rates of 25-50%.[12] After development of plasma fractions and cryoprecipitates, the mortality rate fell to 10% and further to 1% after development of highly active factor VIII and IX concentrates.[12]

Presence of an expanding hematoma (usually post trauma), pulsatile mass, and uncontained abdominal mass indicate need for surgical exploration.

This study shows that with multidisciplinary approach and participation of experts, retroperitoneal hematoma in Haemophilia patients can be managed conservatively.

## CONCLUSION

Acute pancreatitis is one of the common diagnoses for acute abdomen in an adult in the emergency room.

Usually patients present with the history of alcohol consumption or history of biliary colic. But in this case, the patient presented to us with pain abdomen with palpable mass per abdomen similar to pancreatitis with pseudocyst formation. However, past history of easy bruisability and a family history of bleeding manifestations lead us to haematological evaluation, which helped us diagnose haemophilia type B. Patient was managed conservatively and discharged. Now the patient is doing good and on regular follow up.

## ACKNOWLEDGEMENTS

We want to thank our patient for consenting to the publication of the article. We would like to thank the ex-head of department, Department of General Surgery, Dr. Mallikarjun Desai, for leading and guiding the team in management of the case. We thank the present HOD, Dr. B. S. Pai for all the support and encouragement provided.

*Funding: No funding sources*
*Conflict of interest: None declared*
*Ethical approval: Not required*

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**Cite this article as:** Beedubailu R, Tubachi P. Large retroperitoneal hematoma in a patient with haemophilia B. Int Surg J 2021;8:1913-6.