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Spontaneous Retroperitoneal Haematoma Unmasking Haemophilia a in a Young Adult

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Abstract
We report a 33-year-old man who presented to the Emergency Department with two brief syncope episodes and abdominal pain radiating to the hip after a 20-km motorcycle ride. He was tachycardic (165/min) and hypotensive (90/60 mmHg) with pallor; airway and breathing were intact. Initial investigations showed alkalemia with elevated lactate (pH 7.508, HCO_3^- 17.7 mmol/L, lactate 10.21 mmol/L), severe anaemia (Hb 6.22 g/dL), mild hyponatremia (Na 133 mmol/L) and hypokalaemia (K 3.2 mmol/L). ECG revealed sinus tachycardia with a short PR interval, and chest radiography was unremarkable. Contrast-enhanced CT of the abdomen identified a spontaneous retroperitoneal hematoma. Resuscitation included intravenous fluids, analgesia, vitamin K (10 mg), and transfusion of uncrossmatched packed red cells. Post-CT labs showed leucocytosis (31,600/μL), platelet count 4.57 lakh/μL, and markedly prolonged aPTT (72.7 s) with INR 1.20; repeat venous gas demonstrated acidosis and rising lactate (pH 7.251, lactate 12.20 mmol/L). Thromboelastography showed prolonged R (42.2 min) and K (9.8 min) with reduced α-angle (22.1°) and borderline MA (53.9 mm), consistent with a coagulation-factor deficiency phenotype. He underwent digital subtraction angiography, which demonstrated no aorto-iliac leak, and received intra-procedural blood products (packed red cells, fresh frozen plasma, cryoprecipitate). Coagulation assays confirmed haemophilia A with factor VIII activity of 4%; recombinant factor VIII was initiated (tapered over 3–6 June 2024), alongside cryoprecipitate support. Serial monitoring showed clinical stabilization, ultrasound evidence of hematoma reduction, and a rise in factor VIII to 73.9% at discharge. This case highlights spontaneous retroperitoneal haemorrhage as a sentinel presentation of previously unrecognized haemophilia A and underscores the value of early CT imaging, viscoelastic testing, targeted factor replacement, and selective angiography in management.
Keywords: *Spontaneous retroperitoneal haemorrhage, Haemophilia A, Factor VIII, Thromboelastography, Contrast-enhanced CT, Digital subtraction angiography*

INTRODUCTION

Spontaneous retroperitoneal haemorrhage (SRH) is an uncommon but potentially life-threatening entity that often presents with nonspecific abdominal, flank, or hip pain and hemodynamic instability; its deep anatomic location can delay recognition until substantial blood loss has occurred. Contrast-enhanced computed tomography (CECT) is the diagnostic modality of choice because it rapidly confirms the presence, extent, and – when present – active extravasation of retroperitoneal bleeding, thereby triaging patients for angiography or conservative management.(1) In contemporary practice, hemodynamically stable SRH is frequently managed non-operatively with resuscitation and correction of coagulopathy, whereas digital subtraction angiography with transarterial embolization is reserved for ongoing or severe haemorrhage and achieves high technical and clinical success in appropriately selected patients.(2, 3)
Coagulopathy is a major precipitant of SRH; among inherited causes, haemophilia A (factor VIII deficiency) is the most prevalent. Disease severity is defined by residual factor VIII activity – mild (5–40%), moderate (1–5%), and severe (<1%) – and correlates with bleeding phenotype. Deep-muscle and retroperitoneal bleeds are considered serious events requiring prompt factor replacement targeting high initial levels. The World Federation of Haemophilia (WFH) recommends aiming for ~80–100 IU/dL factor VIII for major internal bleeding, with subsequent maintenance at protective levels and close monitoring.(4) For adults with a new, isolated prolongation of activated partial thromboplastin time (aPTT) and soft-tissue bleeding, acquired haemophilia A (AHA) must also be considered. Best practice includes mixing studies and inhibitor quantification with a (Nijmegen-)Bethesda assay; management typically combines haemostatic therapy (bypassing agents or factor replacement when feasible) with immunosuppression to eradicate the inhibitor.(5)

Bedside viscoelastic testing can complement standard coagulation studies in bleeding patients. On thromboelastography (TEG), a prolonged R time indicates delayed clot initiation from coagulation-factor deficiency or anticoagulant effect, whereas a reduced α-angle and prolonged K time reflect impaired fibrin build-up/fibrinogen contribution; these patterns guide targeted transfusion and factor therapy.(6) We report a 33-year-old man who presented to the Emergency Department at SRIHER, Chennai, with syncope and abdominal/hip pain, found to have SRH on CECT and a factor-deficiency profile, culminating in a new diagnosis of haemophilia A – highlighting the importance of early CT imaging, structured haemostatic evaluation, and guideline-directed factor replacement in SRH.

Case report

A 33-year-old man presented with two episodes of syncope and abdominal discomfort radiating to the hip, which had begun one day earlier. The pain developed after he had travelled 20 km by motorcycle. Each syncopal spell lasted approximately 2–3 minutes. He denied any fall, headache, vomiting, seizure, or obstipation. He reported no significant past medical history, took no regular medications, and had no known drug allergies. His last oral intake occurred after the journey, about 12 hours before arrival at the emergency department. On presentation, his airway was patent, and he was able to vocalize. He was breathing spontaneously at 18 cycles per minute with an oxygen saturation of 99% on room air, and auscultation revealed bilateral vesicular breath sounds without added sounds. All peripheral pulses were palpable; the pulse rate was 165/min and blood pressure was 90/60 mmHg. Intravenous access was secured using an 18-gauge cannula. He was conscious and oriented with a Glasgow Coma Scale score of E4V5M6, and his pupils were equal and reactive to light. He was afebrile, and complete exposure for secondary survey did not reveal additional injuries or bleeding sources.

General examination showed pallor. Cardiovascular examination demonstrated normal S1 and S2 without murmurs. Respiratory examination confirmed good bilateral air entry with no added sounds. Neurological examination revealed no focal deficits. Abdominal inspection showed no visible swelling or scars and no visible peristalsis; hernial sites were free, and cough impulse was negative. On palpation, the abdomen was soft with diffuse tenderness, without rigidity or palpable masses; cough impulse remained negative. Percussion elicited tenderness in all quadrants with an overall resonant note, and liver dullness was present. Bowel sounds were audible on auscultation.

Initial investigations revealed a pH of 7.508, PCO_2 of 30.3 mmHg, PO_2 of 57.0 mmHg, bicarbonate of 17.7 mmol/L, and lactate of 10.21 mmol/L. Serum sodium measured 133 mmol/L, potassium 3.2 mmol/L, chloride 96.9 mmol/L, and glucose 121.0 mg/dL; haemoglobin was 6.22 g/dL. A 12-lead electrocardiogram showed sinus tachycardia with a short PR interval and was otherwise unremarkable. An anteroposterior chest radiograph did not demonstrate acute abnormalities; the lung fields appeared clear without focal consolidation, pleural effusion, or pneumothorax, and the cardiac silhouette was within expected limits. A provisional diagnosis of acute abdomen was considered in the emergency department. Initial management comprised intravenous normal saline with a 1-litre bolus followed by 100 ml/hour, analgesia with tramadol 50 mg diluted in 100 ml normal saline intravenously together with ondansetron 4 mg intravenously, and dispatch of routine laboratory tests including blood grouping and typing. A contrast-enhanced CT (CECT) of the abdomen was performed and demonstrated a spontaneous retroperitoneal haematoma.

Following the CECT, one unit of uncrossmatched packed red blood cells was transfused and vitamin K 10 mg was administered. A thromboelastogram (TEG) was obtained, vascular-surgery consultation was sought, and the patient was admitted under vascular surgery for digital subtraction angiography (DSA). Post-CECT laboratory evaluation showed haemoglobin 6.5 g/dl, total leucocyte count 31,600/cumm, platelet count 4.57 L, activated partial thromboplastin time (aPTT) 72.7 seconds (prolonged), prothrombin time 14.9 seconds with INR 1.20, blood urea nitrogen 28 mg/dl, and creatinine 0.9 mg/dl. A repeat venous blood gas revealed pH 7.251, haemoglobin 7.62 g/dl, and lactate 12.20 mmol/L. The TEG demonstrated marked coagulopathy: R time 42.2 minutes (elevated; normal 4–8), K time 9.8 minutes (elevated; normal 0–4), alpha angle 22.1° (reduced; normal 47–74), and maximum amplitude (MA) 53.9 mm (borderline low; normal 54–72). Additional indices showed G 5.8 K d/sc (low; normal 6.0–13.2), coagulation index −30.7 (low; normal −3 to 3), thrombodynamic potential index 5.9/sec (low; normal 32–527), and TMA 73.9 minutes. Overall, the tracing was consistent with deficiency of coagulation factors and impaired fibrin formation with borderline platelet contribution. The patient received cryoprecipitate transfusions (eight units administered periodically), and, following factor assays, recombinant factor VIII therapy was initiated for newly diagnosed haemophilia A. Coagulation testing

showed aPTT 60.1 seconds (reference 20.6–30.6) and a severely reduced factor VIII level of 4% (reference 70–150%), with other factors as follows: factor VII 106.9%, factor IX 209.7%, von Willebrand factor 201.5%, factor V 68.2%, factor II 127%, factor X 109%, factor XI 106%, factor XII 103.9%, factor XIII 103.9%, fibrinogen 407 mg/dl, and thrombin time 14.5 seconds (reference 16.4–18.8).
Vascular surgery proceeded to DSA with intraoperative transfusion of two units of packed red cells, eight units of fresh frozen plasma, and eight units of cryoprecipitate. The angiogram did not reveal any leak from the aorta or the iliac arteries. The post-procedure course in the ICU was uncomplicated; the patient was monitored for bleeding and continued on factor VIII replacement. The chronology of haemostatic management was as follows: the haematology service assumed care after DSA; on 01/06/2024 haemophilia A was diagnosed and plans were made for recombinant factor VIII replacement and extubation; on 03/06/2024 factor VIII was given intravenously every 12 hours for 48 hours; on 04/06/2024 three doses of 3,000 IU factor VIII were administered and the factor VIII level rose to 20.94%, after which the dose was reduced to 1,500 IU for one day; on 05/06/2024 abdominal ultrasonography showed reduction in the volume of the clot; and on 06/06/2024 the factor VIII dose was tapered to 500 IU per dose with further adjustments based on serial factor levels. The patient remained clinically stable and was discharged with advice for regular follow-up. Factor VIII measured 4% on 01/06/2024 and improved to 73.9% at discharge.

DISCUSSION

This case illustrates a rare but recognized presentation of SRH unmasking haemophilia A in an adult, with initial features of hypovolemia (tachycardia, hypotension, anaemia, high lactate) and abdominal/hip pain. SRH is frequently occult, may present late after substantial blood loss, and carries meaningful morbidity and mortality; timely recognition is essential. Contrast-enhanced CT is the preferred initial imaging because it rapidly localizes retroperitoneal blood and can demonstrate active extravasation, guiding the need for angiography or intervention.(7) The CECT in our patient demonstrated a retroperitoneal hematoma without an angiographic source on subsequent digital subtraction angiography. Contemporary series support a step-up strategy in SRH: hemodynamic resuscitation and correction of coagulopathy, observation when stable, and DSA with transarterial embolization for ongoing or hemodynamically significant bleeding. When pursued, embolization achieves high technical and clinical control rates and has largely supplanted open surgery.(8, 9) The diagnostic laboratory constellation – prolonged aPTT with normal PT/INR, markedly low factor VIII activity (4%), and a viscoelastic profile showing prolonged R and K with a reduced α-angle – was most consistent with a coagulation factor deficiency. On thromboelastography, a prolonged R time signifies delayed initiation of clotting (coagulation factor deficiency or anticoagulant effect), while prolonged K and a low α-angle point to impaired fibrin build-up/fibrinogen contribution; together they guide factor and cryofibrinogen replacement. The borderline-low MA suggested limited platelet contribution, but not to the degree typically seen with primary thrombocytopenia or profound dysfunction. These interpretations align with established TEG physiology and clinical guides.(6, 10, 11)
Factor assays confirmed haemophilia A. On phenotypic grounds, a factor VIII activity of ˜4% falls in the moderate range (moderate 1–5%; severe <1%; mild 5–40%). In patients with haemophilia, gastrointestinal and deep muscle/iliopsoas bleeds are considered serious or life-threatening events that warrant immediate factor replacement to high target levels. World Federation of Haemophilia (WFH) guidance recommends initial peak FVIII levels of 80–100 IU/dL for gastrointestinal haemorrhage (typically for 7–14 days) and for deep-muscle bleeds with neurovascular compromise or substantial blood loss, with subsequent maintenance at lower but protective levels.(4) Our management pathway – early cryoprecipitate, packed red cells, and initiation of recombinant FVIII – accords with these recommendations. Because this was a new adult presentation, acquired haemophilia A (AHA) – an autoantibody-mediated FVIII deficiency – was an important early differential. AHA typically presents with an isolated prolonged aPTT and soft-tissue or mucosal bleeds and requires confirmation with a mixing study and quantification of inhibitors by a (Nijmegen-)Bethesda assay. When inhibitors are present, first-line haemostasis often relies on bypassing agents or recombinant porcine FVIII, along with prompt immunosuppression to eradicate the inhibitor. In our case, the brisk rise of FVIII to 73.9% with standard recombinant FVIII replacement makes a high-titre inhibitor unlikely, but formal inhibitor testing remains best practice in new adult diagnoses.(12, 13)
Target levels and duration of factor replacement in major internal bleeds are critical. For severe or life-threatening haemorrhage (e.g., large soft-tissue or retroperitoneal bleeds), authoritative references advise

achieving FVIII levels of at least 50% and often 80–100% initially, then maintaining protective levels while the bleed stabilizes – principles reflected in WFH guidance and summarized in contemporary clinical resources.(4, 14) Finally, the negative DSA in this patient, coupled with correction of coagulopathy and targeted factor replacement, aligns with data showing that some SRH cases do not demonstrate an arterial 'bleeder' and can be managed non-operatively when haemodynamics and haemoglobin stabilize. Close monitoring in an ICU, serial labs (including factor levels), and imaging to document hematoma evolution are standard components of safe conservative care.(8)

Briefly, SRH should be considered in adults with syncope or shock and abdominal/hip pain, particularly when laboratory testing reveals an isolated aPTT prolongation. CT is first-line to confirm SRH and triage; TEG patterns can rapidly point to a factor-deficiency phenotype; and WFH-guided FVIII replacement to high initial targets is central to hemostasis. In new adult presentations of FVIII deficiency, rule out AHA with mixing and inhibitor assays, even when initial replacement appears effective.

CONCLUSION

This case underscored that spontaneous retroperitoneal haemorrhage may be the sentinel event revealing an underlying coagulation disorder, as in our patient with newly diagnosed haemophilia A. Early CECT imaging, bedside viscoelastic testing, and definitive factor assays enabled rapid haemostatic diagnosis and targeted therapy with recombinant factor VIII, achieving clinical stabilization without the need for embolization or surgery. Clinicians should maintain a high index of suspicion for factor deficiencies in adults presenting with abdominal/hip pain and isolated aPTT prolongation, ensuring prompt, guideline-directed replacement and close monitoring to optimize outcomes.

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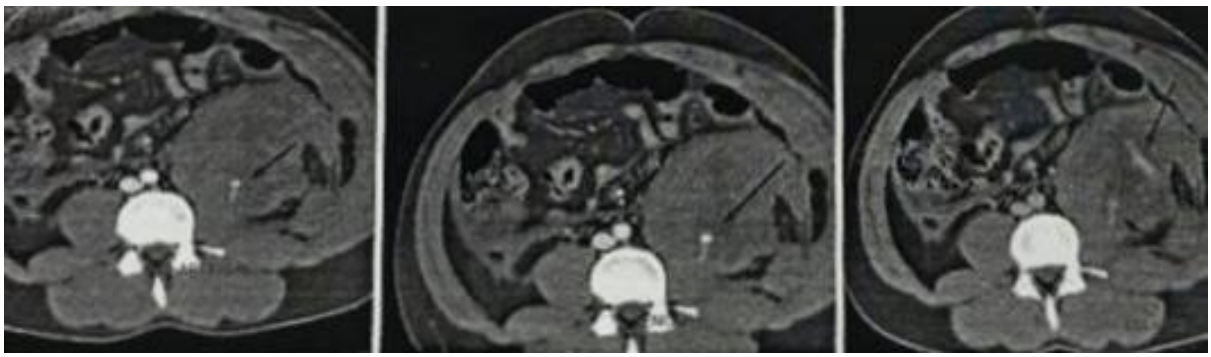

Figure 1: Computed tomography findings