# Recurrent Spontaneous Subdural Hematoma in a Patient with Severe Hemophilia A: A Case Report and Clinical Reflections from a Resource-Limited Setting


## Abstract

Background: Intracranial hemorrhage (ICH) is one of the gravest complications in hemophilia, often leading to significant neurological morbidity and mortality. Spontaneous subdural hematomas (SDHs) are especially rare and pose diagnostic and therapeutic dilemmas in patients with severe coagulation defects.

Case Presentation: We report a 31-year-old male with genetically confirmed severe Hemophilia A who developed recurrent spontaneous SDHs without antecedent trauma. The patient was managed conservatively with recombinant factor VIII concentrate and neurocritical care monitoring, resulting in clinical stabilization and radiological improvement.

Conclusion: This case underscores the critical need for high clinical vigilance, early imaging, and aggressive factor replacement in preventing progression of hemorrhagic events in hemophilia. Structured hemophilia care networks and access to prophylactic therapy remain indispensable in resource-limited countries such as India.


Keywords: Hemophilia A, Subdural hematoma, Intracranial hemorrhage, Coagulopathy, Factor VIII, Resource-limited settings.

## Introduction

Hemophilia A, an X-linked recessive disorder, results from mutations in the F8 gene encoding coagulation factor VIII, leading to varying degrees of bleeding diathesis. Severe hemophilia, characterized by factor VIII activity <1%, predisposes to spontaneous

hemorrhages into joints, muscles, and vital organs including the brain. Intracranial hemorrhage (ICH) remains a life-threatening event and has been reported in up to 10% of patients with severe hemophilia, particularly in those not receiving primary prophylaxis. [1,2]

Among the various types of ICH, spontaneous subdural hematomas (SDHs) are infrequent and generally follow trauma. However, spontaneous SDHs have been reported even in the absence of trauma, likely due to vascular fragility and defective secondary hemostasis. The limited literature on recurrent SDH in hemophilia underscores the need for awareness and timely intervention. [3,4]

This case from a tertiary care center in India highlights the presentation and conservative management of spontaneous recurrent SDH in a patient with severe Hemophilia A and reflects on the broader implications of hemophilia care in low- and middle-income countries.

**Case Report**

A 31-year-old male, diagnosed with severe Hemophilia A in childhood, presented with a two-day history of progressive frontal headache and multiple episodes of vomiting. There was no history of trauma, loss of consciousness, focal neurological deficits, or seizures. His medical history included a prior conservatively managed SDH, completed therapy for pulmonary tuberculosis, and prior thrombophlebitis of the lower limb. He was not on regular prophylactic factor VIII due to financial constraints. There was no history suggestive of inhibitor development, and a recent Bethesda assay was negative.

On examination, he was afebrile, alert, and oriented with stable vital signs. Neurological examination was unremarkable, with no papilledema or motor deficits. Routine blood investigations were within normal limits except for an activated partial thromboplastin time (aPTT) >300 seconds and mildly elevated CRP. CT imaging of the brain revealed an acute

right fronto-temporo-parietal SDH measuring 10.2 mm in thickness, with a 3.6 mm midline shift and early ventricular effacement as shown in Image 1.

The patient was managed in a high-dependency setting. Recombinant factor VIII was administered to maintain trough levels above 50 IU/dL for five days, followed by tapering. Neuroprotective measures such as head-end elevation, mannitol, and normovolemia were implemented. Neurosurgical consultation was sought, but given the absence of neurological deterioration, conservative management was favored. Serial imaging showed gradual resolution of the hematoma and midline shift.

**Discussion**

ICH in hemophilia represents a critical emergency. Studies have shown that without early intervention, mortality can reach as high as 34%, and survivors often suffer long-term neurocognitive deficits. [5,6]The pathophysiology of SDH in hemophilia is multifactorial—ranging from venous rupture secondary to minor unnoticed trauma to spontaneous vessel rupture in the absence of structural anomalies. Vascular fragility due to repeated subclinical bleeds, inflammatory changes, and prolonged deficiency of coagulation factors are implicated.[7]

While the majority of ICH in hemophilia involves intraparenchymal or intraventricular spaces, SDH presents a unique management challenge. A conservative approach is often effective in hemodynamically and neurologically stable patients, particularly when adequate factor replacement is available. However, rapid neurosurgical intervention becomes necessary in cases of raised intracranial pressure or clinical worsening. [8,9]

The WFH recommends immediate replacement of factor VIII at doses of 50–60 IU/kg during acute ICH, followed by sustained levels >50 IU/dL for at least 7–14 days depending on resolution and clinical course. [10] This approach was mirrored in our patient, with gradual tapering post-clinical stabilization. In the presence of inhibitors, bypassing agents such as recombinant activated factor VII (rFVIIa) or activated prothrombin complex concentrate (aPCC) are preferred, although they carry a risk of thrombosis. [11]

Emerging therapies, including the bispecific antibody emicizumab, offer a promising alternative for prophylaxis, especially in inhibitor-positive patients. Its subcutaneous administration and prolonged half-life improve compliance and quality of life. [12] In India, while state-led programs have made recombinant factor concentrates more accessible, a universal prophylactic program is yet to be implemented. Kerala's model of comprehensive hemophilia care offers a scalable framework. [13]

Interestingly, our patient had a history of thromboembolic events. While hemophilia is traditionally considered protective against thrombosis, literature reports rare thrombotic episodes in hemophiliacs, especially in the setting of immobility, central venous access, or use of procoagulant therapies. [14] These events demand individualized decision-making and close interdisciplinary collaboration.

## Conclusion

Recurrent spontaneous SDH is a rare but serious manifestation of severe Hemophilia A. Early diagnosis through neuroimaging, prompt factor replacement therapy, and close monitoring can avert the need for surgical intervention. Structured hemophilia care, including access to prophylaxis and comprehensive follow-up, remains the cornerstone for preventing ICH recurrence. This case reinforces the importance of resource-sensitive care models in improving outcomes for hemophiliacs in developing countries.

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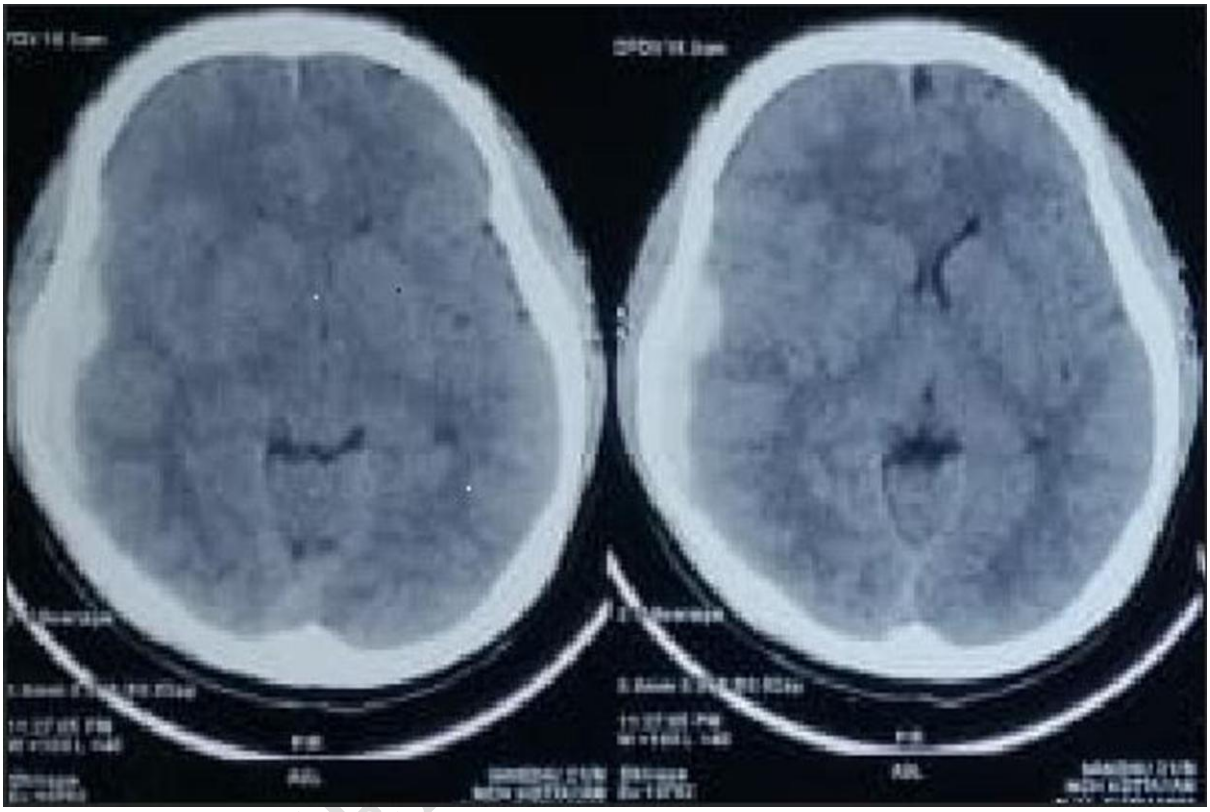

Image 1- Acute Subdural Hematoma with 3.6 mm midline shift