Case of the Week

ACUTE MYOCARDIAL INFARCTION IN A PATIENT WITH MODERATE HEMOPHILIA-A

Managed at **Ashoka Medicover Hospital, Nashik**

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Introduction

Acute Myocardial Infarction (AMI) in patients with Hemophilia A is an uncommon but increasingly recognized condition as life expectancy improves. Hemophilia A, a congenital bleeding disorder caused by Factor VIII deficiency, predisposes to hemorrhagic events rather than thrombosis. However, with advancing age and the presence of cardiovascular risk factors, atherosclerotic heart disease and MI can occur.

This case highlights the clinical and therapeutic challenges of managing myocardial infarction in a moderate Hemophilia A patient, requiring multidisciplinary coordination to balance thrombosis prevention and bleeding control.

Pathophysiology

In Hemophilia A, Factor VIII deficiency results in defective thrombin generation and impaired fibrin clot formation.

Despite this hypocoagulable state, coronary thrombosis can still occur due to plaque rupture and platelet aggregation.

<u>Common predisposing risk factors include:</u>
- Hypertension
- Diabetes mellitus
- Dyslipidemia
- Smoking and sedentary lifestyle

Treatment challenges arise as antithrombotic therapy, though lifesaving in MI, carries a high risk of major bleeding unless Factor VIII activity is adequately corrected.

Clinical Presentation

A middle-aged male, known case of moderate Hemophilia-A (Factor VIII activity ~3%), presented with:
- Sudden-onset retrosternal chest pain radiating to the left arm
- Associated sweating and breathlessness
- No trauma or active bleeding

On evaluation:
- ECG: ST-segment elevation in anterior leads (suggestive of STEMI)
- 2D Echo: LAD territory severe hypokinesia, LVEF 30%, severe LV systolic dysfunction
- Troponin I: Positive
- Baseline Factor VIII activity: 3%

Clinical Summary
The patient was diagnosed with Acute Anterior Wall Myocardial Infarction with severe LV systolic dysfunction and cardiogenic shock in the background of moderate Hemophilia A.
A multidisciplinary approach involving Cardiology and Hematology was adopted.

On arrival in ER, stat long-acting Factor VIII replacement was administered to raise activity to 100% before initiating antithrombotic therapy

Dual antiplatelet therapy and anticoagulation were started under close monitoring

Coronary angiography revealed LMCA involvement with critical triple vessel disease (TVD) and proximal LAD 100% occlusion (culprit lesion)

Primary PCI to LAD was performed with drug-eluting stent placement under Factor VIII coverage

As the patient was in cardiogenic shock, he was managed with Noradrenaline infusion and supportive therapy

Gradual hemodynamic improvement noted; vasopressor support tapered off by POD-1

Patient mobilized comfortably by POD-2 and shifted to ward in stable condition

Long-acting Factor VIII replacement was continued every 3rd day during recovery to maintain adequate hemostatic levels and prevent delayed bleeding

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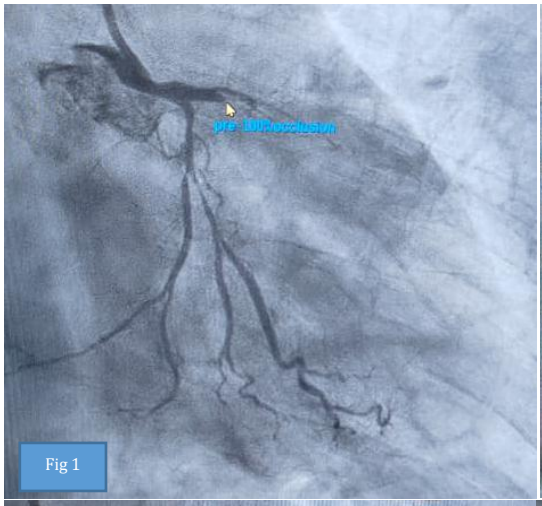

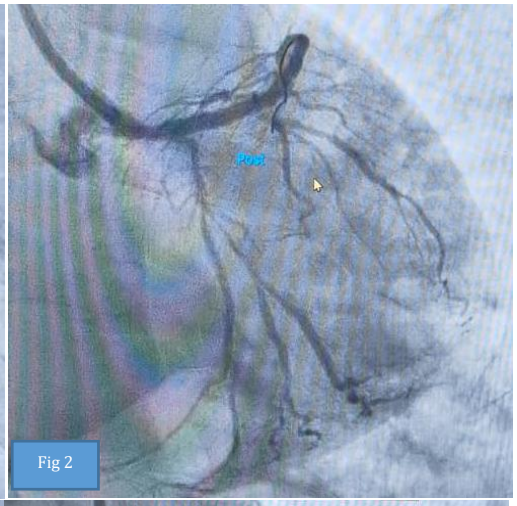

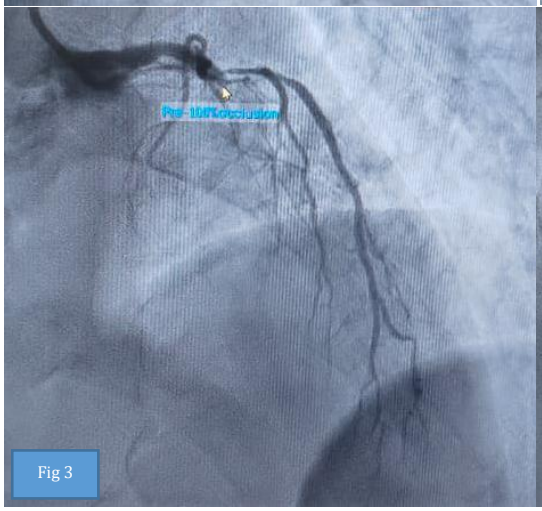

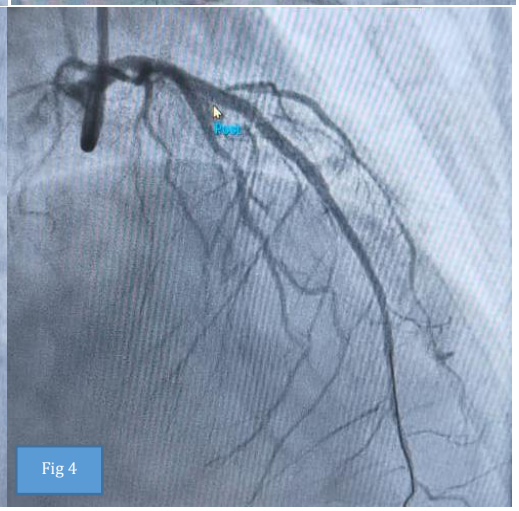

Figure 1 & 3: Coronary angiogram showing proximal LAD 100% occlusion (Pre-PCI)

Figure 2 & 4: Post-PCI angiogram showing restored LAD flow (TIMI 3)

Management

Admission to Cardiac Intensive Care Unit (CICU) under multidisciplinary supervision

Stat long-acting Factor VIII replacement followed by maintenance every 3rd day to sustain hemostatic levels (>50%) throughout the intervention and recovery phase

Dual antiplatelet therapy and controlled anticoagulation initiated after hemostatic correction

Primary PCI to LAD performed under appropriate factor coverage

Cardiogenic shock and LV failure managed with vasopressors, diuretics, and oxygen therapy

Serial monitoring of CBC, coagulation profile, and Factor VIII activity

Continuous coordination between Cardiology and Hematology teams for post-PCI care

Detailed family counselling regarding the diagnosis, treatment protocol, and follow-up requirements

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Discharge

The patient showed progressive clinical and hemodynamic improvement:
- No active cardiac complaints
- Mobilized comfortably
- Noradrenaline tapered and discontinued
- No bleeding or ischemic recurrence
- Platelet count and coagulation parameters remained stable
- Tolerating oral cardiac medications well

Discharged in stable condition with regular follow-up planned with Cardiology and Hematology.

Final Diagnosis

· Anterior Wall Myocardial Infarction with Cardiogenic Shock
· CAG - LMCA + Triple Vessel Disease
· Primary PCI to LAD (culprit lesion)
· Moderate Hemophilia A (Factor VIII Deficiency)

Why Timely Treatment Matters

<u>With Treatment:</u>
-Timely revascularization restored myocardial perfusion
-Early reversal of cardiogenic shock
-Prevention of secondary organ failure
-No bleeding complications due to appropriate Factor VIII correction

<u>Without Treatment:</u>
-High risk of extensive myocardial necrosis and mortality
-Possibility of fatal arrhythmias or cardiac arrest
-Severe bleeding if thrombolytics or antithrombotics used without correction
-Poor recovery and long-term cardiac dysfunction

Revascularization Strategy and Protocol

As thrombolysis is absolutely contraindicated in Hemophilia patients, revascularization was undertaken via Primary Percutaneous Coronary Intervention (PCI) under the Institutional STEMI Management Pathway, consistent with the European Society of Cardiology (ESC) STEMI Guidelines 2023 and the AHA/ACC STEMI Management Protocol 2021.

Given the hospital's tertiary cardiac care infrastructure with cath-lab facilities, Primary PCI was chosen as the definitive revascularization strategy, ensuring rapid myocardial salvage with optimal hemostatic control and multidisciplinary oversight.

Key Points

- Myocardial infarction can occur even in patients with moderate Hemophilia A due to atherosclerotic risk factors
- Factor VIII replacement is essential before any invasive or antithrombotic therapy
- Maintenance long-acting Factor VIII should be administered every 3rd day during the recovery phase to sustain adequate hemostasis
- Factor VIII correction to ≥80–100% is mandatory before PCI or initiation of antithrombotics
- Primary PCI is the preferred revascularization strategy in Hemophilia, as thrombolysis is contraindicated
- Multidisciplinary coordination between Cardiology and Hematology ensures optimal safety and outcomes
- Close post-procedure monitoring and structured follow-up are vital for recurrence prevention and complete recovery.

Reference

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