
بسم الله الرحمن الرحيم

ACQUIRED HEMOPHILIA A WITH INTRAMUSCULAR HEMATOMA AT AN UNUSUAL AGE

FROM CASE REPORT TO PRACTICAL, GUIDELINE-BASED MANAGEMENT

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WELCOME AUDIENCE. INTRODUCE TOPIC: AHA IS RARE, LIFE-THREATENING, AND DIAGNOSTIC DELAYS ARE COMMON, ESPECIALLY AT UNUSUAL AGES. PRESENTING CLINICAL STORY, DIAGNOSTICS, AND PRACTICAL TREATMENT

PATIENT STORY

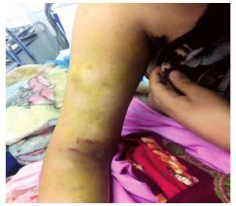
Previously healthy young patient
No personal/family bleeding history
Sudden painful intramuscular hematoma
Progressive anemia, no trauma or anticoagulant use
Speaker Notes:
Start with patient story to engage audience. Highlight absence of bleeding history. Mention timeline from symptom onset to lab testing.

Case report

A 13-year-old girl *presented to the Emergency Department of Imam Khomeini Hospital, Tehran, Iran with painless swelling, ecchymosis, and pain in her right arm (Figure). She had a two-week history of frequent epistaxis that would take a long time to cease. Her medical history was not significant. She had not experienced trauma, fever, or surgery within the month before her admission. She did not have any personal or familial history of bleeding disorders. The patient had been experiencing frequent unusual epistaxis and diffuse ecchymosis in the dorsal side of her left leg associated with swelling and pain since two weeks before her admission. On admission, she had a blood pressure of 110/80 mmHg, a heart rate of 106 beats/minute, and a body temperature of 36.8 °C. On physical examination, there were antecubital fossa ecchymosis (size: 5 × 4 cm) and right axillary fossa ecchymosis (size: 10 × 6 cm). The non-pitting swelling of her arm had disseminated to the right hemi-thorax without any evidence of swelling in the hand and forearm. The radial and ulnar pulses were normal. There was no pain or abnormal neurologic findings in the distal of upper limbs. Evaluation with ultrasonography reported compressible jugular and proximal subclavian veins with normal flow. Doppler sonography of the veins and arteries of the right upper limb was normal, but there was a hematoma in the soft tissue. Disseminated superficial ecchymosis was determined following the above evaluations. The patient's coagulation test, factor assay, and laboratory test results are shown in Table. These results confirmed the diagnosis of acquired hemophilia A. After the initial evaluations, underlying inhibitory or immunologic etiologies were presumed to be the cause of the disease. In addition, due to the severity of the hematoma, the patient was given one dose (4.8 mg) of factor VII (90 μg/kg) immediately after the initial evaluations as factor VII could alleviate the symptoms. In the first days of admission and after laboratory confirmation of antibody-involved hemophilia, intravenous immunoglobulin (IVIG) (1 g/kg) was prescribed. Symptoms and the color of skin lesions alleviated in response to treatment after a few days. Consequently, the patient was discharged from the hospital after her condition became relatively stable. She was prescribed cortisone acetate (1 g/kg per day) for one month and azathioprine (50 mg per day) for three months. She was regularly followed up with prothrombin time (PT), activated partial prothrombin time (aPTT), international normalized ration (INR), and anti-FVIII antibody tests. After one month of treatment initiation, cortisone acetate was tapered and stopped within two weeks. In about two months, antibody level decreased to the normal upper limit level. After three months, the levels of PT, PTT, INR, and antibody were normal, and the patient was in a good physical condition. Subsequently, azathioprine was tapered (after three months of treatment).*

Discussion

In this study, we reported a novel case of acquired hemophilia in a 13-year-old female. Acquired hemophilia A is very uncommon in children. The incidence of acquired hemophilia has been reported to be 1 in 1.48 million/year the United Kingdom [1]. A study reported six cases of hemophilia A in children in the U.S. and a literature review revealed another eight presumed or definite cases [8]. In addition, a large retrospective study reported another six cases [9].



Massive hematoma and swelling of the patient's arm and forearm after obtaining a blood sample

The results of the patients' coagulation test, factor assay, and laboratory tests

Laboratory test	Laboratory result
Hemoglobin	5.6 g/dL (normal: 12–16 g/dL)
Hematocrit	16.1 % (normal: 36–48 %)
Von Willebrand factor activity	93 % (normal: 50–160 %)
aPTT	104.6 seconds (normal: 32–40 seconds)
Mixed aPTT	52 seconds (high–not corrected)
PT	13.4 seconds (normal: 12.3–14.5 seconds)
Bleeding time	3 minutes (normal: 2–7 minutes)
Factor 8 inhibitor	16.8 Bethesda Units (< 0.4 none)
Platelet count	Normal
Liver function	Normal
Activity of factors 2, 5, 7, 9, 10, 11	Normal
Anti-thyroid peroxidase (anti-TPO)	1.8 (negative)
Antinuclear antibodies (ANA)	2.3 (negative < 10)
Anti-double-stranded DNA (anti-dsDNA)	0.6 (normal < 100)
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However, studies on acquired hemophilia A in Asian countries are rare. To our knowledge, the present report is one of the few reported cases of an Asian patient that was diagnosed with acquired hemophilia A at a young age. A study reported that the weighted mean (SD) age at diagnosis of acquired hemophilia was 58.10 (16.96) years in Asian countries compared to 75.70 (14.47) years in an European series (with an absolute difference of 17.6 years) [10]. There seems to be no significant difference in the occurrence of acquired hemophilia in the two genders. However, younger age (< 50 years) and female gender have been reported to be associated with more risks for AHA [5]. AHA usually occurs in women in the postpartum period, in patients with connective tissue disease, paraneoplastic

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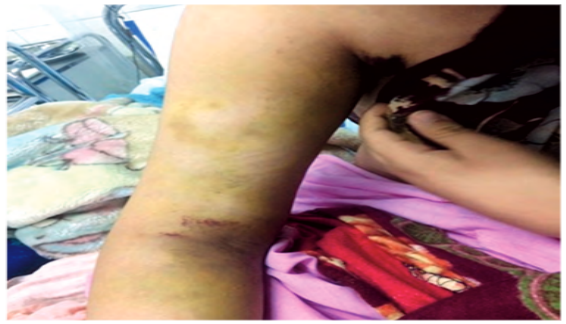

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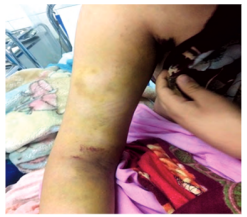

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INITIAL CLINICAL IMPRESSION & RED FLAGS

- Isolated prolonged aPTT → major red flag
- Sudden soft tissue/muscle bleeding without trauma
- No history of anticoagulant use
- Consider acquired factor inhibitor early
- Speaker Notes:
- Stress early suspicion for AHA to prevent delays. Red flags guide lab testing and management

TIMELINE OF CLINICAL EVENTS

- ***Day 0: Hematoma o***
- ***Day 1–2: Hemoglobin drop***
- ***Day 3: Isolated prolonged aPTT detected***
- ***Day 4: Mixing study performed***
- ***Day 5: FVIII inhibitor confirmed***

INITIAL DIFFERENTIAL DIAGNOSIS

- ***Congenital hemophilia***
- ***Lupus anticoagulant (usually no bleeding)***
- ***Anticoagulant/medication-induced bleeding***
- ***Other rare acquired factor inhibitors***

- Discuss rationale for each differential. Age and spontaneous bleeding are clues for AHA.

LABORATORY INVESTIGATIONS

- ***Hemoglobin*** ***8.9 g/dL***
- ***WBC*** ***$7.4 \times 10^9/L$***
- ***Platelets*** ***250/000***

- *PT* *12.0 sec* *(11–14 sec)*
- *INR* *1.0*
- *aPTT* *72 sec* *(25–35 sec)*
- *Mixing study* *Incomplete correctio*

- *FVIII activity 5% (50–150%)*

(Severely reduced)

Suggests inhibitor

- *Bethesda assay 8 BU (<0.6 BU)*
- *(Positive inhibitor)*

aPTT prolonged → suspect FVIII deficiency
Mixing study incomplete → inhibitor present
FVIII 5% + Bethesda 8 BU → definitive diagnosis
Autoimmune / organ function tests normal
Speaker Notes:
Links lab data to pathophysiology. Shows reasoning from labs → diagnosis.

AUTOIMMUNE SCREEN

- ***ANA*** → ***negative***
- ***ds DNA*** → ***negative***
- ***No systemic autoimmune***
- ***Renal & Liver function*** → ***Normal***

- ***No organ dysfunction***

- Highlight isolated prolonged a PTT. Mixing study + Bethesda assay confirm inhibitor

DETAILED LABORATORY INTERPRETATION

- ***Hemoglobin: acute blood loss***
- ***Normal platelets/PT***
- ***aPTT prolonged → suspect FVIII deficiency***
- ***Mixing study incomplete → inhibitor present***
- ***FVIII 5% + Bethesda 8 BU → definitive diagnosis***
- ***Autoimmune/organ function tests normal***

DIAGNOSTIC APPROACH

- ***Isolated prolonged aPTT → red flag***
- ***Mixing study → distinguishes inhibitor vs factor deficiency***
- ***Factor VIII activity measurement***
- ***Bethesda assay → confirms inhibitor***
- ***Exclude lupus anticoagulant***

- Early suspicion is critical. Mixing study guides interpretation; FVIII + Bethesda gives definitive diagnosis.

KEY DIFFERENTIALS EXPLAINED

- Congenital hemophilia → lifelong/family history
- Lupus anticoagulant → usually no bleeding
- Medication-induced coagulopathy → check drugs
- Other rare inhibitors → post-partum/autoimmune

- Explain why each differential considered. Age + spontaneous bleeding clues for AHA.

TREATMENT PRINCIPLES

- ***Control acute bleeding (haemostatic therapy)***
- ***Eradicate inhibitor (immunosuppressive therapy)***
- ***Monitor response & side effects***

- Both goals are parallel. Haemostatic therapy alone → recurrence. Early intervention saves lives.

HEMOSTATIC THERAPY OPTIONS

- ***Bypassing agents: rFVIIa / aPCC***
- ***FVIII concentrates: usually ineffective if high titer***
- ***Choice guided by bleeding severity & inhibitor titer***

- ***Start hemostatic therapy immediately in severe bleeding. Discuss local dosing guidelines.***

CHOICE OF HEMOSTATIC AGENT (PRACTICAL TIPS)

- ***rFVIIa → preferred for high titer***
- ***aPCC → alternative***
- ***Monitor thrombosis in adults***
- ***Pediatric: lowest effective dose***

- Age-specific considerations. Monitor bleeding, FVIII, side effects.

INHIBITOR ERADICATION THERAPY

- ***First-line: corticosteroids 1 mg/kg/day***
- ***Add cyclophosphamide or rituximab if needed***
- ***Combination → higher remission***
- ***Duration: 4–6 weeks, taper steroid***

- ***Early initiation critical. Age-specific considerations: fertility, toxicity, thrombosis risk. Monitor labs weekly***

IMMUNOSUPPRESSIVE REGIMEN DETAILS

- ***Prednisone: 1 mg/kg/day, taper 4–6 wks***
- ***Cyclophosphamide: 1–2 mg/kg/day if combined***
- ***Rituximab: 375 mg/m² weekly × 4 doses***
- ***Monitor: FVIII activity, inhibitor titer, CBC, liver/renal, infection***

- Practical guidance. Monitor for side effects, especially in pediatric patients. Adjust therapy per response.

ADULT VS PEDIATRIC CONSIDERATIONS

- ***Adults: higher thrombosis risk → careful with bypassing agents***
- ***Pediatrics: minimize cytotoxic exposure, growth/fertility considerations***
- ***Principles similar, risk–benefit differs***
- ***Adjust immunosuppressive regimens based on age/ comorbidities***

- Adult → bypassing ± steroid ± cyclophosphamide
- Pediatric → consider rituximab earlier, minimize cytotoxic exposure
- Monitor labs, side effects, growth/fertility

CLINICAL OUTCOME

- ***Bleeding controlled in X days***
- ***Inhibitor eradicated in Y weeks***
- ***No thrombotic complications***
- ***Patient monitored for relapse***

- Guideline-based therapy → excellent outcome even at unusual age.

FOLLOW-UP & MONITORING

- ***Weekly FVIII activity & inhibitor titer until normalized***
- ***Monitor bleeding episodes***
- ***Watch steroid/cytotoxic complications***
- ***Long-term follow-up for relapse***

- Structured follow-up crucial. Age-specific considerations

TREATMENT ALGORITHM (ADULT VS PEDIATRIC)

- ***Step 1 – Diagnosis: aPTT → mixing study → Bethesda assay***
- ***Step 2 – Acute Bleeding: Severe → rFVIIa / aPCC; Mild → FVIII concentrate***
- ***Step 3 – Inhibitor Eradication: Steroids ± cyclophosphamide/rituximab***
- ***Step 4 – Follow-Up: Weekly FVIII & inhibitor; monitor bleeding, thrombosis***
- ***Step 5 – Age-Specific: Adult → thrombosis risk; Pediatric → minimize cyto***

- Walk audience stepwise. Emphasize simultaneous management and age-specific modifications.

TAKE-HOME MESSAGES

- ***Suspect AHA in spontaneous bleeding + isolated prolonged aPTT***
- ***Early hemostatic therapy + inhibitor eradication is life-saving***
- ***Unusual age → vigilance required***
- ***Pediatric vs adult → individualized therapy***

- Practical, guideline-based decision making. Apply lessons from this case

PUBLISHED CASE REPORT

- ***Title: Acquired hemophilia A with intramuscular hematoma at an unusual age: a case report***

- ***Journal: [Journal Name], 2023; Volume:Pages***
- ***Key Point: Rare age, diagnostic challenge; guideline-based therapy successful***

- Shows credibility. Peer-reviewed. Rare age emphasizes novelty.