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Acquired hemophilia A: the cause of life-threatening hemorrhage in elderly

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Acquired hemophilia A (AHA) is a rare bleeding disorder with a poor prognosis if the diagnosis and effective treatment are delayed. While the overall incidence is approximately one case per million per year, it is underestimated because diagnosis is often missed in routine practice as its symptoms differ from its congenital counterpart. We describe the case of a 72-year-old female presented with spontaneous intramuscular bleeding in the left thigh, with no evidence of fracture. Initial laboratory study revealed the activated partial thromboplastin time(aPTT) was prolonged at 108 seconds (normal 28-45 seconds). Her intramuscular bleeding was well controlled without complication and she left the hospital without being diagnosed. Two weeks later, she was hospitalized with her intramuscular hemorrhage getting worse. A 1:1 mixing assay showed a partially corrected aPTT of 44.6 seconds. We assayed for clotting factors specific to the intrinsic pathway and determined that the patient had a factor VIII deficiency 1% (normal 60-140%). Further assays revealed factor VIII inhibitors in the patient's serum. AHA diagnosis was established and patient was started treatment with desmopressin, and prednisolone, cyclophophamide for immunosuppression. Despite of improvement in the laboratory values she continued to have progression of her bleeding. Excessive hemorrhage led to disseminated intravascular coagulation(DIC), and patient died eleven days after the diagnosis. The failure to early recognize this condition leads to delays in the initiation of effective treatment, and leads to a fatal outcome. AHA should be suspected as the cause of intramuscular or subcutaneous hemorrhage in elderly with no previous history of bleeding presents with bleeding and an unexplained prolonged aPTT. Rapid and accurate diagnosis, effective hemostatic therapy, and timely treatment are important in the management of acquired hemophilia.

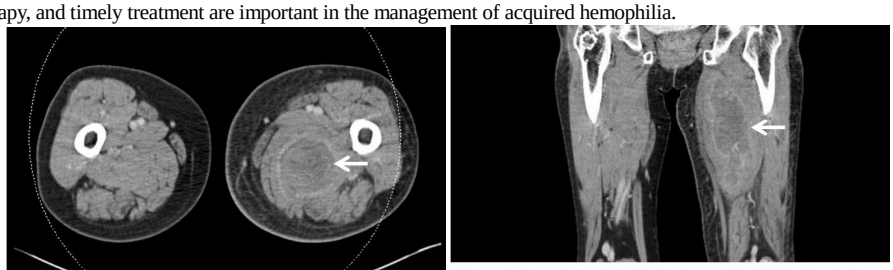

Figure 1. Acute hematoma in the Lt. proximal thigh, just below ischial tuberosity, about 5*6*15 cm in size, associated diffuse soft tissue edema in the Lt. thigh subcutaneous fat layer, muscles, intermuscular septum. No gross fracture in the Lt. ischial tuberosity & Lt. femur.

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Prognostic Significance of Serum Ferritin in Patients with Acute Myeloid Leukemia

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Background/Aims: Acute myeloid leukemia (AML) is a clonal malignant disease of hematopoietic tissues, leading to a premature arrest of the normal differentiation of stem cells. Elevated serum ferritin (SF), is often observed in non-transfused AML patients, however, the prognostic value of the baseline SF is unclear. We evaluated the clinical significance of baseline SF levels in non-transfused AML patients. **Methods:** The retrospective study cohort included 40 patients who were newly diagnosed with AML from January 2010 to December 2017 and received standard induction chemotherapy. Allogenic hematopoietic stem cell transplantation was performed in 10 patients. **Results:** The patients with AML were predominantly male (75%, 30/40). Median age at diagnosis with AML was 62 years (range, 21 – 79 years), and most of them (45%, 18/40) received high dose chemotherapy only, 25% (10/40) of patients underwent allogenic peripheral blood stem cell transplantation.(Table1) About 40% (16/40) patients were died during follow-up period. The median overall survival (OS) and relapse-free survival (RFS) were 26.2 months and 13.3 months, respectively. The median SF value at diagnosis of AML was 614.3mg/dL. When patients were divided into the low baseline SF group (≤ 1000 mg/dL) and high baseline SF group (≥ 1000 mg/dL), particularly striking was the finding that the median RFS of former group was superior to the latter group (14.9 vs 9.3 months, p<0.001). However, there was no difference in OS between two groups (26.2 vs 18.7 months, p=0.653). **Conclusions:** We have demonstrated that SF level ≥1000 mg/dL was associated with worse RFS. In conclusion, we believe that baseline SF level could be a prognostic factor for RFS in AML patients.

Variables (Total N=40)	
Median Age	62 (range, 21 – 79)
Gender	
Male	75% (30/40)
Female	25% (10/40)
Treatment	
Standard chemotherapy only	50% (20/40)
Standard chemotherapy followed by allogenic peripheral blood stem cell transplantation	25% (10/40)
Palliative chemotherapy only	25% (10/40)
Median baseline Ferritin (mg/dl)	613.9 (range, 42.81~2000)
High ferritin (>1000mg/dl)	25% (10/40)
Low ferritin (≤1000mg/dl)	75% (30/40)
Using deferasirox	45% (18/40)