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ABSTRACT N. PO78

ACQUIRED HEMOPHILIA A AS A PARANEOPLASTIC SYNDROME: TWO CASES REPORT ON A RARE COEXISTENCE.

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Background: Acquired hemophilia, meanwhile, is the most common acquired disease affecting clotting factors. Possible causes that have been empirically described include autoimmune disease (such as rheumatoid arthritis or systemic lupus erythematosus), malignancy (most frequently solid tumors), dermatologic disorders (such as psoriasis or pemphigus), postpartum changes or drug interactions. Acquired hemophilia occurs due to either acquired deficiency of factor VIII or development of factor VIII inhibitor in the body. Here, we describe a rare case of acquired hemophilia in a woman whit malignancy and in a man with pemphigus.

Cases repost: The first patient is a 75-year-old female, with a history with a history of breast cancer, currently in progression of the disease with pulmonary metastases. After the initiation of atezolizumab, the patient reported experiencing diarrhea and melanotic stool, without other episodes of bleeding, such as epistaxis, ecchymosis or hematemesis. A coagulation workup was done, revealing an elevated APTT-ratio of 4.10 (normal range: 0.85 to 1.20). Other lab studies included a complete blood count, revealing a hemoglobin of 8.4 g/dL and a platelet count of 274 platelets/mL. A subsequent mixing study did not normalize the PTT, suggesting the presence of an inhibitor. The titer of the inhibitor was elevated at 129 Bethesda units (normal range: less than 0.6 Bethesda Units). The second patient is a 76-years-old man with type pemphigus presented to the local hospital with progressively more extensive discoloration of the left thigh, accompanied by worsening thigh pain and limitation of limb movement, for over 5 days. He had a hemoglobin level of 9.9 g/dL with a platelet count of 315 x 103 /µL and WBC count of 7.3 x 103 /µL. His coagulation profile was normal except for isolated prolongation of APTT-ratio which was 2.89. Failure of correction of the prolonged APTT with mixing of pooled normal plasma suggested the presence of an inhibitor. Factor assay revealed a factor VIII level of 0.5% with normal factor IX and XI levels. The Bethesda assay to quantify inhibitory antibodies was done and the titre was 691 BU.

Conclusions: In these reports, two rare cases of acquired hemophilia in patients, who developed a factor VIII inhibitor as a paraneoplastic syndrome, are presented. Even though acquired haemophilia A is a rare entity, it should be considered as one of the causes of adult onset abnormal bleeding, especially in patients with underlying autoimmune disorder and malignancy. Early clinical suspicion, diagnosis and prompt management of the severe refractory form of acquired haemophilia A is lifesaving.

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