Saving Life if Recognized, Causing Mortality if not Recognized: Acquired Hemophilia A

Tanınırsa Hayat Kurtarır, Tanı Konulamazsa Yüksek Mortalite: Edinsel Hemofili A

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ABSTRACT

Acquired hemophilia A is a bleeding disorder caused by antibodies to factor VIII. Patients typically present with an isolated prolonged activated partial thromboplastin time due to FVIII deficiency. In the control of acquired hemophilia, it is important the diagnosis time. In this case, I presented a case of acquired hemophilia A, which I diagnosed after massive bleeding after pregnancy.

Keywords: Acquired hemophilia A, pregnancy, inhibitor

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ÖZET

Edinilmiş hemofili A, faktör VIII'e karşı antikorların neden olduğu bir kanama bozukluğudur. Hastalar tipik olarak FVIII eksikliğine bağlı olarak izole uzamış aktive parsiyel tromboplastin zamanı ile başvururlar. Edinsel hemofili kontrolünde tanı zamanı önemlidir. Bu olguda gebelik sonrası masif kanama sonrası teşhis ettiğim bir edinsel hemofili A olgusunu sundum.

Anahtar Sözcükler: Edinsel hemofili A, gebelik, inhibitör

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To the editor,

Acquired hemophilia A is a bleeding disorder caused by antibodies to factor VIII (FVIII). These antibodies—called inhibitors—interfere with normal hemostasis, leading to potentially catastrophic bleeding. Patients typically present with an isolated prolonged activated partial thromboplastin time (aPTT) due to FVIII deficiency (1). It occurs in 1/1000000 cases and there are no randomized controlled trials for definitive treatment. In 50% of the acquired hemophilia cases, an underlying cause cannot be identified and the etiology of the remaining ones is solid tumors, hematological malignancies, rheumatic diseases, pregnancy, and drugs (2-3). In the control of acquired hemophilia, It is important the diagnosis time. Early and accurate diagnosis is directly related to morbidity and mortality (4).

A 25-year-old female patient had her first vaginal delivery in August 2021. The patient, who had no history of bleeding diathesis and had normal prenatal prothrombin time (PT) and activated partial thromboplastin time (aPTT) levels, were admitted to the emergency department on the postpartum 40th day for nausea, vomiting, and flank pain. Left ureter cystoscopic biopsy was performed in the patient with ureteronephrosis by urologist after evaluation. During the operation, 3 units of erythrocyte suspension and 2 units of fresh frozen plasma were given. After the biopsy result was reported as papillary urothelial neoplasia, a left nephrectomy was performed at the end of November, although the preoperative aPTT was 96 sec [normal range: 21-35 sec]. In the patient who was admitted to the gynecology clinic with hypovolemic shock in the 10 days following the operation, massive transfusions were performed and extensive intra-abdominal hematoma areas were detected in the previous operation site. At the same time, no evidence of malignancy was found in the pathological evaluation of the nephrectomy material. The patient was consulted at our clinic. Her aPTT: 60 sec, PT: 13 sec, fibrinogen 6.46 g/L, platelet count 336x10^9/L, hemoglobin 5.9 g/dl were detected and aPTT did not correct in the immediate and incubated mixing study. FVIII level of the patient was 3%, and FVIII antibody titer was detected as 3 Bethesda unit (BU). The patient was started on rFVIIa at a dose of 90 mcg/kg with 2 doses and methylprednisolone at a dose of 1 mg/kg. The bleeding was brought under control and the patient was discharged with the continuation of corticosteroid therapy. In the next 1-month follow-up, aPTT was 34 sec, factor VIII level was 15% and inhibitor level was 1 BU.

Acquired hemophilia, although it is very rare, is a fatal disease in which ecchymosis, hematoma, severe mucosal hemorrhages are seen, and severe bleeding cases. If the underlying cause can/can not be found, it is essential for the treatment of inhibitor elimination with steroid and/or immunosuppressive drugs in addition to the underlying cause therapy.

In this case, a patient with very massive bleeding after delivery and a critical clinical course was presented. Our experience with this case suggests that all physicians should be cautiously interpreted clotting tests in the preoperative period. If the physician is detected the prolonged clotting tests, they should be requested the mixing tests. Acquired hemophilia is a hematological emergency. Physicians should be aware of this life-threatening condition for saving patients' life on time.

Conflict of interest

No conflict of interest was declared by the author.

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