Letter to Editor

A woman with late postpartum hemorrhage and placenta accrete

To the Editor

I read with interest the case reported by S. Akhavan published in the recent issue of the Journal of Research in Medical Sciences ¹. I would like to declare my opinion about it. For screening hemostasis disorders before invasive procedures or surgery, we usually test APTT, PT, INR, BT, and Platelet Count to detect relevant abnormalities. Although these tests may be sufficient for major hemostasis disorders, some hemostasis disorders such as factor XIII deficiency may be overlooked. The PT, APTT, and thrombin time are normal in factor XIII deficiency. In these patients, bleeding following trauma or surgery may be delayed for 12 to 36 hours, while in other patients immediate

bleeding occurs ².

Although the final diagnosis for the cited patient was placenta accrete, it is useful to consider factor XIII deficiency as a differential diagnosis in similar conditions. Factor XIII (fibrin stabilizing factor) catalyzes lysine bonds between various protein substrates such as fibrin monomers, a2-plasmin inhibitor, fibronectin, and collagen. These intermolecular cross-linking reactions between various plasma and extracellular matrix proteins contribute to hemostasis, wound healing, and maintenance of pregnancy ³. A screening test for the presence or absence of factor XIII consists of observing clot solubility or insolubility in 5 M urea or 1% monochloroacetic acid ⁴.

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In Response

Factor XIII is a transglutaminase that stabilizes fibrin clots by forming å-amino-ã glutamil cross-links between adjacent á and ã chains of fibrin. Factor XIII deficiency is an extremely rare inherited syndrome. Patients usually bleed in neonatal period from their umbilical stump or circumcision. In addition to hemorrhage, these patients may have poor wound healing, a high incidence of infertility among males and abortion among affected females, and a high incidence of intracerebral hemorrhage. These observations suggest that the enzyme may be important in other physiologic processes beyond hemostasis, including placental implantation, spermatogenesis, and wound healing. Several drugs including isoniazid, may bind to cross-linking sites on fibrinogen and mimic factor XIII deficiency by blocking enzyme activity. Normal hemostasis requires only 1% of normal enzyme activity; a single effusion of fresh frozen plasma or a purified factor XIII-rich product derived from human placenta called fibrogammin is effective.

The reported case was a woman with normal hemostasis in past medical history and her post-operative condition was normal.

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