Factor XIII

Factor XIII or fibrin stabilizing factor is an enzyme (EC 2.3.2.13) of the blood coagulation system that crosslinks fibrin. Deficiency of this factor (FXIIID) affects clot stability. FXIIID, while generally rare, does occur, with Iran having the highest global incidence of the disorder with 473 cases. The city of Khash, located in Sistan and Balochistan provinces, has the highest incidence in Iran, with a high rate of consanguineous marriage.

FXIII-A Val34Leu polymorphism is not associated with ICH risk in a Caucasian population. Further large and well-designed studies must be conducted to confirm this preliminary conclusion [1)].

Spontaneous subdural hematomas particularly with a decreased coagulation factor XIII activity require follow-ups of the neuroradiological diagnostic [2)].

A 68-year-old man presented with intracranial hemorrhage in the right frontal lobe, which rapidly increased the day after admission. Arishima et al. performed hematoma removal with a biopsy of the cortex around the hematoma. The day after the operation, a subcutaneous hematoma over the craniotomy appeared, and the computed tomography showed a recurrent hemorrhage with an acute subdural hematoma.

They were aware of a bleeding tendency, and a detailed hematologic examination by hematologists revealed autoimmune acquired factor XIII deficiency due to an antifactor XIII antibody. Specimens taken around the hematomas were pathologically diagnosed as cerebral amyloid angiopathy (CAA) on immunohistochemical examination.

They considered that acquired factor XIII deficiency had induced lobar hemorrhage in the frontal lobe affected with CAA, and the coagulation disorder induced postoperative rebleeding. The patient died from repeated lobar hemorrhage 3 years after the surgery. There is no routine screening coagulation test including the active partial thromboplastin time and the prothrombin time for factor XIII deficiency. It is important for neurologists and neurosurgeons to be aware of this rare disease in patients with a bleeding tendency [3)].

1)
Ye X, Ye B. Association between the Val34Leu polymorphism in blood coagulation factor XIII-A and intracerebral hemorrhage: a meta-analysis. Genet Mol Res. 2016 Jul 25;15(3). doi: 10.4238/gmr.15038327. PubMed PMID: 27525858.
2)
Bosche B, Kraus B, Molcanyi M. Spontaneous subdural hematomas particularly with a decreased coagulation factor XIII activity require follow-ups of the neuroradiological diagnostic. Neuroradiology. 2017 Apr;59(4):323-324. doi: 10.1007/s00234-017-1793-3. Epub 2017 Feb 24. PubMed PMID: 28236052.
3)
Arishima H, Neishi H, Kikuta KI, Morita M, Hosono N, Yamauchi T, Souri M, Ichinose A. Lobar Hemorrhage Induced by Acquired Factor XIII Deficiency in a Patient with Cerebral Amyloid Angiopathy. J Stroke Cerebrovasc Dis. 2017 Aug 8. pii: S1052-3057(17)30366-X. doi:

10.1016/j.jstrokecerebrovasdis.2017.07.009. [Epub ahead of print] PubMed PMID: 28801214.