Symposium HAEMOPHILIA

THE IMPORTANCE OF MULTIDISCIPLINARY TREATMENT OF A SEVERE FORM OF HAEMOPHILIA A

Amalia Matei¹, Sorina Bădeliță¹, Manuela Crisan^{1,2}, Valentina Uscatescu¹, Inga Botnariuc¹, Razvan Mihalcea¹, Daniel Coriu^{1,2}

Haematology and Bone Marrow Transplant Department, Fundeni Clincal Institute, Bucharest ²"Carol Davila" Medicine and Pharmacy University, Bucharest

A 50 year old male known to have a severe form of haemophilia A, presents, after a fall, a bifocal fracture to the right femur followed in the next 48 hours by a superior digestive bleeding. The patient is transferred to Haematology department of Fundeni Clinical Institute for treatment with altered general status, severe pallor, nasogastric tube (1000 mls of fresh blood in 24 hours), multiple black stools and immobilized right leg.

After admittance, the OGD showed large gastric ulcer. The patient received Esomeprazole treatment as 80 mg iv bolus and continuous drip of 8 mg/hour without any improvement on the bleed. Due to low plasma level of Factor VIII (<1%) and high level of inhibitors of Factor VIII (50 BU), the patient received Feiba and Novoseven treatment without any improvement on the bleed followed by recombinant Factor VIII treatment associated with fresh frozen plasma and cryoprecipitate support which led to bleeding disappearance and reduced number of black stools. Due to altered parameters on coagulation tests (prolong aPTT and low plasma level of fibrinogen), it raised concerns on complications onset (infection or hematoma) of the right leg. The orthopaedic consult found a multifocal fracture of right femur and led to minimal invasive procedure with osteosinthesis with 3 Ender stems with good recovery and optimal plasma level of Factor VIII (19% ante- and 21% post-procedure). In the recovery period, the patient is complaining of sudden headache, emesis and becoming sleepy. The cerebral CT scan showed massive subdural hematoma of the right emisphere with signs of acute bleeding and cerebral oedema. The patient is transferred to Neurosurgeon Hospital where the hematoma was drained. The patient was discharged after the control OGD showed healed ulcer and no signs of acute bleeding.

For handing this case, a multidisciplinary team formed by Haematologists, Gastroenterologists, Orthopaedics, Neurologists and Neurosurgeons and ICU specialists was involved and a very large amount of blood, fresh frozen plasma and cryoprecipitate transfusions, 130.000 U recombinant factor VIII, 45 vials of Feiba and 50 vials of recombinant factor VIII were used.

The costs were covered mostly by the hospital and only a small amount was covered by Haemophilia National Programme.