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Case Report

Making Diagnosis in a Bleeder Case with Impaired Coagulation Study: a Case Report

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Abstract

A 14 year old boy with diagnosis of right leg-calve-par thesis had been admitted to orthopedic surgery department in our hospital. He had prothrombin time: 13 second, activated partial thromboplastin time: 33 second and platelet count; 262 x10^9/µl. The patient had undergone femoral endarterectomy release surgery successfully, but had hemorrhage at site of incision. The bleeding had continued in 3 subsequent days despite daily infusion of 2 bags of fresh frozen plasma. The patient had received 5 bags of packed cell and 16 bags of fresh frozen plasma during subsequent 2 weeks.

The results of coagulation study were as follow: factors VIII: 38%, IX: 12.9%, V: 5.6%. In re-interview with the patient, it was revealed that he had history of a severe epistaxis and bleeding after circumcision. The interview with the family directed us to a known case of mild hemophilia B. The treatment changed from plasma to factor IX concentrate with target of reaching to plasma level of 80%. The bleeding at site of incision began to reduce and stopped in second day of infusion of factor IX concentrate and continued till 4 days.

After a month the factor IX level was 6%. The second surgery was programmed for 2 months later with infusion of 3000 unit factor IX concentrate before and after surgery that kept on to 5 days later, then tapered to 1500 U until 5 others days. He did not experience any bleeding at site of surgery except epistaxis at 9th and 10th days of hospitalization.

Keywords: Hemophilia B; Diagnosis; Coagulation assay; Factor IX; Medical history; Intervie

Case Presentation

A 14 year old boy with main chief complaint of pain and claudicating of right foot from last year had been referred to our hospital. His symptoms had been aggravated during last year. He had admitted to orthopedic surgery department with primary diagnosis of right leg-calve-par thesis. His physical examination revealed no history of trauma and fever. The biochemical and hematological parameters had been reported normal including Prothrombin Time (PT): 13s (second) (normal range: 12.5s), Activated Partial Thromboplastin Time (APTT): 33s (normal range: 25-35s) and platelet count; 262 x 109/µl. The patient had undergone femoral endarterectomy release surgery successfully, but had been experienced severe hemorrhage at site of incision. The bleeding was continued in 3 subsequent days despite infusion of 2 bags of Fresh Frozen Plasma (FFP) daily. The patient had been undergone of infusion of 5 bags of packed cell and 16 bags of FFP during subsequent 2 weeks without any recovery. Also primary coagulation study had requested that showed PT: 23s and APTT: 70s.

A consultation on current case was requested of hematology department. At this stage the patient was stable with active bleeding, according to impaired APTT test. Owing to normal APPT before the surgery a panel of coagulation factor assay requested as follow: factor XIII, I, V, VIII, and IX. The results of reported factors VIII: 38%, IX: 12.9%, V: 5.6% and the others (I & XIII) were in normal range. Again a detailed past medical history asked of the patient and it was revealed that he experienced a severe epistaxis and bleeding after circumcision. A survey and making history in other member of the family directed us to a registered case of mild hemophilia B and the other suspected brother with history of vast ecchymosis, bleeding after tooth extraction, epistaxis and post circumcision bleeding. The latest brother underwent coagulation assay and revealed mild hemophilia B too (factor IX: 6%). The therapeutic regimen of the patient changed from FFP to factor IX concentrate with target of reaching to plasma level of 80%. The bleeding at site of incision began to reduce and stopped in second day of infusion of factor IX concentrate and continued till 7 days.

After a month, the factor IX level was assayed for the patient that showed mild hemophilia B with factor IX level: 6%. The patient and his new diagnosed brother refereed to hemophilia center for registration in database.

Owing to complete his surgical procedure he needed a complementary and second orthopedic surgery, so the patient admitted 2 months later and underwent implant arthroplasty with infusion of 3000 U (unit) factor IX concentrate before the surgery and after surgery and continued to 5 days later and then tapered to 1500 U until 5 others days. He did not experience any bleeding at site of surgery, but an episode of epistaxis occurred at 9th and 10th days of hospitalization. The consultation with department of ear, nose, and throat was done and they packed up tampons in nose and after its removal usage of vitamin A cream 3 times a day prescribed and the patient discharged after stabilization.

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Discussion

Hemophilia B is an X-linked hemorrhagic disorder due to abnormality in the factor IX gene. Its frequency is 1/3 of hemophilia A, but has similar sub grouping [1]. Hemophilia B is nearly similar in clinical picture and type of inheritance with hemophilia A too.

A simple and strict interview with the patient and his/her family may be useful screening toll in detection of diathesis to hemorrhagic disorders [2] and would be first step of evaluation of a suspected case. In the case of neonate and children, interview should be done with parents and also it would be better to cover time line from delivery time to now. The age of suspected case would be considered in interview due to some bleeding manifestation are impossible in childhood [3]. The babies with hemophilia who had been delivered using forceps or vacuum extraction (ventouse) may show symptoms such as vast ecchymosis or even intracranial bleeding in first day of life. While it is not complete known which question in medical interview of bleeder one may be more helpful in making diagnosis [2] and there are no consensus among experts about value questions interview on hemorrhagic diathesis [4], it seems a complete questions set that cover both primary and secondary hemostasis abnormalities may be informative.

The net diagnosis in bleeding disorders is necessary to avoid mismanagement and prevention of aggravated consequences [1]. In current case, the primary interview with the patient was done by a general physician student and it was written on medical recorded that the patient has no important past medical history. The missing of past bleeding manifestation resulted in long hospitalization and usage of untargeted drug as FFP in current case.

Also coagulation study in current case was unravel of net diagnosis owing to it showed multiple coagulation factors defect and was not instructive. In the active bleeding phase, several of coagulation factors may be reduced secondary to continuous consumption and so study of coagulation may not helpful to make diagnosis. Returning to primary stage and checking medical past history using interview with the patient and his family was complete enlightening and terminated to correct diagnosis.

Conclusion

Some kites of APPT assays are not complete sensitive enough for screening mild type of hemophilia B. Although hemostasis tests are key step in diagnosis, however are nearly expensive and time wasting so their request would be complete targeted. Making a complete medical interview will lead physician to targeted requested hemostasis test(s), sooner making diagnosis, saving budgetary and time. In pre-operative evaluation of the patients complete asking past medical history (both positive and negative results) in association with results of primary screening tests of hemostasis should be considered especially if they are even about 2-3 s above reference values.

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