Hematology Quiz

Maha Al- Yahyai, Arundathi Kurukulasuriya

Al-Yahyai M, *et al. OMJ.* 24, 145-146 (2009); doi:10.5001/omj.2009.32

A 2 year old baby awaiting surgery for repair of pyloric stenosis. His coagulation screen is as follows:
PT – 10.8s (9.3 – 11.7)
aPTT – 51s (27.2 – 39.1)
Fibrinogen – 3.0g/L (1.5 – 4.2)
Thrombin Time – 15s (12 - 16)

Question 1: The attending surgeon requests a hematology consultation before surgery regarding the prolonged aPTT. What is your response?

(Case continues):

The Corrected aPTT: 37.5s (Normal plasma is added to the test (patient's) plasma in a 50:50 mix and the aPTT is repeated)

Question 2: What can you infer from the results of the mixing test?

Question 3: Will you do any more laboratory test before responding to your colleague in surgery?

(Case continues):

The following are the results of intrinsic pathway coagulation factor assays:
Factor VIII – 89%
Factor IX – 78%
Factor XI – 72%
Factor XII- 15%

Question 4: Why did you carry out the intrinsic pathway factor assays?

Question 5: What advise would you give your surgical colleagues as a response to the hematology consultation?

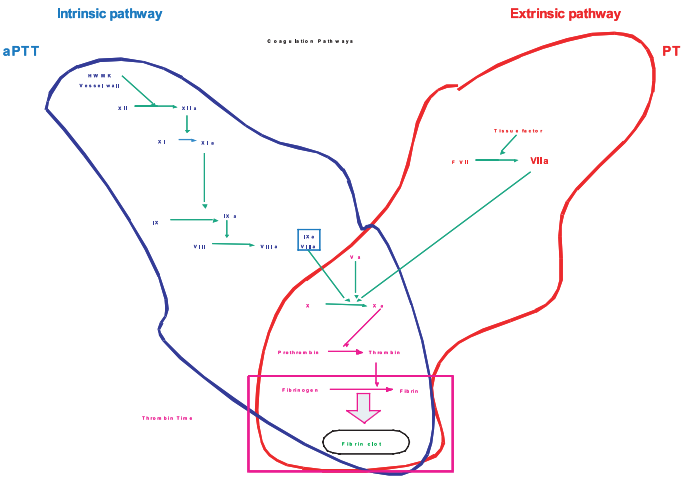
See answers overleaf

Answer 1:
I would repeat the aPTT to confirm that it is a true result and if still prolonged. I would also do a mixing tests. (correction tests)

Answer 2:
The prolongation of the aPTT was caused by a coagulation factor deficiency. On adding the normal plasma in a 50:50 volume mix and repeating the aPTT, the test is verified because the deficient coagulation factor level is brought up to more than 30%, which is adequate to normalize the aPTT[1]. If an inhibitor to a coagulation factor was present, the mixing tests will not correct the aPTT to the normal range as the inhibitor will prevent the action of whatever coagulation factor is added to the mixture. Hence, we know that the prolonged aPTT was caused by a coagulation factor deficiency and NOT an inhibitor.

Answer 3:
Yes, I would:
1) find out presence of a bleeding diathesis in the family
2) assay the coagulation factor levels in the intrinsic pathway.[2]

Answer 4: Deficiency of the intrinsic pathway factors prolong the aPTT[2] as shown in the diagram below.



The parts of the coagulation cascade tested by the Prothrombin time(PT) activated partial thromboplastin time (aPTT) and the thrombin time (TT) which are the coagulation screening tests used in clinical practice.

Answer 5:
The patient has a factor XII deficiency. He also has a family history of Factor XII deficiency. Three factors were considered to confirm the presence of a factor deficiency.

(1) An abnormal laboratory result, confirmed to be true after repetition of the test.
(2) A past family history of bleeding
(3) Bleeding after surgical challenge/child birth/tooth extraction.

The patient had a confirmed low factor XII. A positive family history and no past personal history of bleeding as Factor XII deficiency does not cause bleeding.
It is safe to proceed with the surgery. The patient does not need any blood products or factor concentrates to tide over surgical challenge as Factor XII deficiency is not known to cause bleeding.

Other causes of an isolated prolonged aPTT[3]
1. Lupus anticoagulant
2. von Willebrand disease
3. Inhibitors of the intrinsic pathway coagulation factors or an inhibitor to von Willebrand antigen

References

1. Lewis SM, Bain BJ, Bates I, Dacie and Lewis Practical Haematology; Churchill Livingston Elsevier Philadelphia, 10th ed. 2006, p406.
2. Roberts HR, Escobar MA. Less Common Congenital Disorders of Haemostasis: Consultative Haemostasis and Thrombosis, 2nd ed.2007, p72.
3 Lewis SM, Bain BJ, Bates I, Dacie and Lewis Practical Hematology; Churchill Livingston Elsevier Philadelphia, 10th ed. 2006, p406.

From the Department of Hematology and Blood Tranfusion, Royal Hospital, Muscat, Sultanate of Oman.

Received: 23 Dec 2008
Accepted: 10 Feb 2009

Address correspondence and reprint request to: Dr. Maha Said Al-Yahyai, Department of Hematology and Blood Tranfusion, Royal Hospital, Muscat, Oman.
E-mail: msay99@hotmail.com