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A Bruise Like a Butterfly:
When Hemostasis Goes Awry



by
Sheri L. Boyce
Department of Biological Sciences
Messiah University, Mechanicsburg, PA

Part I – Blood on the Pillow

"Mommy, that looks like a butterfly!" Tessa watched as her four-year-old daughter Macy traced the edges of a large bruise on Tessa's arm. It did look a little like a butterfly, if she tilted her head and squinted. Tessa had no idea how she had gotten the ugly purple bruise, just like she couldn't explain recent bruises on her shin and shoulder.

Caring for an inquisitive preschooler occupied Tessa for the rest of the day, and she forgot about the bruising until that night. She was exhausted and quickly brushed her teeth, only to be startled at the amount of bright red blood when she spit into the sink. Looking into the mirror, Tessa discovered her gums were bleeding again, and she groaned as she recalled her dentist's warnings to floss more regularly.

Tessa fell asleep instantly but was awakened in the middle of the night with a nosebleed that soaked her pillowcase and streaked her T-shirt. It took almost 15 minutes for the bleeding to stop, long enough for Tessa's husband Dan to wonder if they needed to go to the ER. The next morning, he suggested Tessa call the doctor. "That's the third bad nosebleed in the last ten days, and those bruises look pretty bad. Maybe it's more than the dry winter air." Tessa reluctantly agreed.

Several days later, Tessa described her symptoms to Dr. Ramos. She had also noticed heavier menstrual periods, being winded with even mild exertion, and crushing fatigue.

"But I have a four-year-old, so fatigue is normal, right?" Tessa laughed. Dr. Ramos smiled, but then explained all of Tessa's symptoms taken together could indicate a bleeding disorder. He ordered blood tests to confirm his suspicion, and then listened to her lungs again.

"The shortness of breath and fatigue could be related to a possible bleeding disorder, and I don't like the way your lungs sound. Even though you haven't felt sick, let's get a chest X-ray too, just to rule out bronchitis or pneumonia."

Questions

1. Based on the steps of hemostasis, make some predictions (in general terms) regarding possible causes of Tessa's frequent and prolonged bleeding from her nose and gums.

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2. Bruises occur when trauma ruptures a blood vessel in the skin and blood escapes into the surrounding tissue. As the hemoglobin is degraded, the color progresses from purple to green to yellow. Explain how Tessa's severe bruises support the doctor's concern about a bleeding disorder.

3. Dr. Ramos ordered the following blood tests. Based on Tessa's signs and symptoms, predict whether her results might be elevated, decreased, or not affected. Explain your answers.
   a) Hemoglobin and hematocrit:

   b) Platelet count:

   c) Activated partial thromboplastin time (APTT)(measures the function of the contact activation (intrinsic) and common pathways of coagulation):

   d) Prothrombin time (PT)(measures the function of the tissue factor (extrinsic) and common pathways in coagulation):

4. Explain how Tessa's fatigue and shortness of breath could be related to a bleeding disorder.

Part II – Off to the Hospital

A few hours after having her blood drawn and the chest X-ray taken, Tessa saw the results were uploaded in her patient portal. She noticed several numbers that were clearly not normal.

Table 1. Blood test results.

Test	*Tessa's results*	*Normal range*
Hemoglobin (g/dL)	10.8	12–16
Hematocrit (%)	34	37–47
Platelet count (per μl)	< 7000	150,000-400,000
APTT time (sec)	31	25–35
PT time (sec)	11	9–13

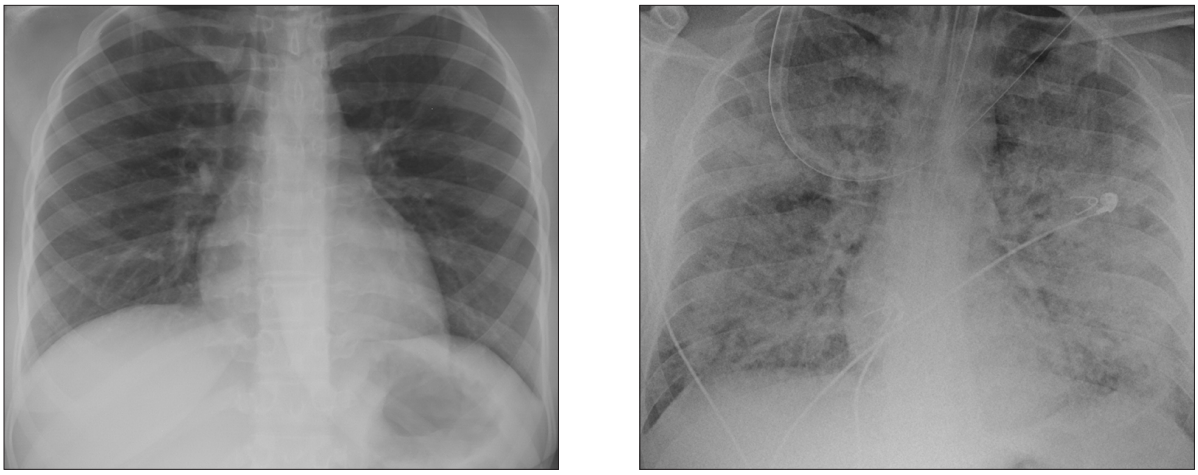


Figure 1. Chest X-rays.

Left: An image of healthy lungs. The black indicates the presence of air in the alveoli. *Image credit:* Normal chest x-ray, Dr. Yale Rosen Atlas of Pulmonary Pathology | Flickr, CC BY-SA 2.0.

Right: Tessa's chest X-ray. The cloudy appearance may indicate the presence of something other than air (e.g., fluid) in the alveoli. *Image credit:* Ground glass opacity, James Heilman | Wikimedia Commons, CC BY-SA 4.0.

Before Tessa could google low platelets, Dr. Ramos himself called.

"Your platelet count is critically low," he explained. "Since platelets are necessary for bleeding to stop, this explains why even minor bumps cause significant bruising and normal brushing makes your gums bleed. In addition, your chest X-ray showed what is called ground glass opacity. Due to your low platelet count, I'm concerned that you may have bleeding in your lungs. I'd like you to go to the hospital as soon as possible to receive a platelet transfusion; that will hopefully prevent catastrophic bleeding and buy us some time to figure out what is causing the low platelet count."

Stunned, Tessa called Dan and made arrangements for Macy to stay with friends. When they arrived at the hospital, the staff quickly began a transfusion of platelets to Tessa, and then sent her for a CT of her lungs.

Dr. Chen, a pulmonary specialist, noted the CT confirmed the ground glass opacity and explained that Tessa needed a bronchial lavage. "The appearance of your lungs indicates the small sacs where gas exchange takes place (the alveoli) are filled with something other than air; in your case, it could be blood. We'll sedate you, and then 'rinse' your airways several times with saline."

Dan tried to lighten the mood. "So kind of like rinsing out my coffee mug?"

Tessa was not amused but Dr. Chen smiled. "Yes. Putting fluid into your lungs sounds like a strange thing to do. However, it's a relatively small amount and when we suck it back out, whatever is filling up your lungs comes out with it. Then we'll know why it's difficult for you to breathe."

The results of the lavage did indeed show that Tessa had bleeding in her alveoli. Dr. Chen noted that this was an unusual complication of bleeding disorders, but it should resolve once they determined the cause and initiated appropriate treatment. "Until then, we'll keep you comfortable and provide additional transfusions if needed."

Questions

5. Dr. Ramos concluded that Tessa's bleeding is likely due to a problem with her platelets rather than a clotting disorder. How do the results of her blood tests support his conclusion? Your answer should reference more than one test.

6. Briefly describe the production, typical lifespan, and destruction of platelets.

7. Based on your answer to Question 6, predict several possible causes (in general terms) of Tessa's low platelet count.

8. What is the clinical term for a low platelet count?

Part III – A Case of Friendly Fire

Additional testing showed that Tessa has a form of thrombocytopenia known as immune thrombocytopenia (ITP). This is an autoimmune disorder in which the immune system generates antibodies that coat the body's own platelets and mark them for destruction in the spleen.

Question

9. Based on Tessa's diagnosis and what you know about the life cycle of platelets, what are some avenues for possible treatment? Answer in general terms.