Aplastic anemia: the correct nomenclature matters

Aplastic anemia is defined as pancytopenia with hypocellular bone marrow in the absence of an abnormal infiltrate and with no increase in reticulin.[1] Dr. Paul Ehrlich, who treated a young woman who died following an illness characterized by bleeding, severe anemia, and high fevers, first described the term in 1888. Her bone marrow was analyzed and labeled as "strikingly hypocellular." It was the French internist, Dr. Anatole Chauffard, who finally introduced the name "Aplastic Anemia" in 1904.

More than 100 years after Ehrlich's discovery, we continue to diagnose and treat the same disease. Hematology has shown that the bone marrow is not only responsible for erythropoiesis as it was believed, but also for the complex production and maturation of leukocytes and platelets. Now we know of diseases that only affect mature neutrophils, lymphocytes, or thrombocytes and, therefore, we are capable of directing management at each particular cell type. Using the correct nomenclature is critical in this process, which is why it should always be precise. Aplastic Anemia is an exception to the rule. Years have passed without the proper modification to reflect its true characteristics. We understand by consensus that "Aplastic Anemia" is a total aplasia of the bone marrow and not a single lineage cytopenia, as its name implies.

Anemia (from the ancient Greek αναιμια, anaimia, meaning 'lack of blood') is defined by a decrease in the total amount of hemoglobin or the number of red blood cells.[2] Values vary depending on the age group.[3] Aplasia indicates defective development, absence, or cessation of the production of a particular tissue. "Aplastic Anemia" is not really only an "anemia," but instead it is a pancytopenia in which leukopenia, anemia, and thrombocytopenia are almost always found. This disease comprises a total bone marrow failure where stem cells are unable to generate all mature elements. Supportive care is not only limited to transfusion of red blood cells, but it also entails control of bleeding and management of infection. Moreover, ultimate treatment is much more complex including immune suppression or challenging bone marrow transplant.

The incidence of Aplastic anemia is reported to be biphasic, with peaks at 10 to 25 years, and the majority of patients presenting beyond 55 to 60 years of age.[1] Heredity, infection, immune diseases, exposure to chemicals and radiation have been attributed to the development of aplastic anemia. The diagnostic criteria are based on the percentage of gross cellularity of the bone marrow and two or more affected cell lines.[4] Despite the precision of the diagnostic criteria, aplastic anemia has always been a diagnosis of exclusion.[5]

In any case, none of this purports to change what we do know: that aplastic anemia is pancytopenia rather than just simply anemia.

Other conditions that are more worthy of the name "aplastic anemia" due primarily to involvement of the red cell lineage are pure red cell aplasia, either congenital (like Diamond Blackfan anemia) or acquired; and "Transient erythroblastopenia of childhood," also known as "transient acquired pure red cell aplasia," which is an anemia following a temporary cessation of red blood cell production and maturity in the bone marrow. In patients with sickle cell disease, Parvovirus B19 infection destroys red blood cell precursors and halts the production, causing a pure RBC "aplastic crisis," in which anemia is severe. The truth is that these entities describe single lineage cytopenias, where there is low hemoglobin and a cessation of red blood cell production.

Anemia is not a synonym for leukopenia. A patient with thrombocytopenia is not the same as a patient with pancytopenia. Nomenclature and classification were created to depict the disorders. The term "Aplastic Anemia" does not describe the entity as it should. A bone marrow failure of this nature should be called "Aplastic Pancytopenia."

Even though there has not been a groundswell of discussion of this nomenclature issue among pediatric and adult hematologists, we are certain that many agree with this objective premise. A correction of this medical term is indicated. Unfortunately, we are all erroneously accustomed to the misnomer and doubt that a modification in the terminology can be accomplished. Perhaps the first step towards achieving this ambitious goal is a publication in an important journal such as this one.

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