MAHA sounds more like a type of fish at a sushi bar than a serious oncological emergency. Early in my oncology career, a physician excitedly told me my patient "was having MAHA." I thought it meant he was eating dinner, so I couldn't quite figure out what the fuss was. I went to see the patient and to inquire about this MAHA. It must be tasty! As oncology nurses it is important for us to understand what MAHA is and why it can't be found on the menu at a local restaurant.

Several populations of oncology patients might develop MAHA (also known as CA-MHA), so it is important to recognize how this relates to other diseases the oncology nurse might know more about such as disseminated intravascular coagulation (DIC) or thrombotic thrombocytopenic purpura (TTP). DIC and TTP are types of microangiopathic diseases that really describe coagulation disorders where vessel endothelial tissue is injured by microscopic thrombosess forming throughout the body. These clog smaller blood vessels and, thus, create a dangerous mine field for future red blood cells to navigate.

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MAHA stands for *microangiopathic haemolytic anemia*. Breaking this down, the “micro” signifies small and, of course, “angi” refers to blood vessels. In MAHA, the endothelial layer of small blood vessels becomes littered with fibrin mesh and fragments. Think of fibrin as splintered trees washed downstream after a flood, piling up against anything that constricts the flow. What is created is a chaotic web that, in this case, literally slices red blood cells wanting to travel through the vessel. This action results in two things. First, the haemolysis (destruction) of the red blood cells creates schistocytes. Greek for the word *schistos*, which means “divided”, these are simply pieces of the red blood cells. Predictably, the schistocytes, also known as helmet cells, are irregular, jagged, and asymmetrical. Second, haemolysis of the red blood cells causes anemia, defined as a decrease in red blood cells or a lack of competent hemoglobin. For this reason, MAHA is considered an acquired haemolytic anemia.

MAHA is particularly associated with advanced cancer patients, hematologic patients with acute promyelocytic leukemia (APL), and patients receiving hematopoietic stem cell transplants (HSCT), particularly allogeneic transplants (Elliott et al., 2010). Solid tumour oncology patients receiving antineoplastic agents such as mitomycin C, cisplatin, gemcitabine, and vascular endothelial growth factor (VEGF) inhibitors will have a higher risk (Levi, 2009). As well, the carcinoma itself can release factors that interrupt coagulation, thus setting in motion a pathological process leading to MAHA (Fontana et al., 2001). Side effects of oncology treatments, including immunosuppression, can result in gram-negative bacteria sepsis (like E. coli), a notorious cause for coagulation chaos and higher risk for DIC.

Signs and symptoms of MAHA include many familiar to the oncology nurse. MAHA is the result of coagulation disorders like DIC and TTP and can be indistinguishable from them with initial assessment (Francis et al., 2007; Oberic et al., 2009). Therefore, closely following the coagulation status of patients at risk for these disorders provides the first level of nursing (and medical) inter-
vention. Oncology patients may have signs and symptoms such as fatigue, headaches, confusion, lethargy, pallor, hematuria, and abdominal pain. Respiratory symptoms, such as tachypnea (quick breathing), dyspnea (trouble breathing), hypoxia (low blood oxygen saturation), and shortness of breath (SOB) are the most common signs of MAHA (Elliott et al., 2010). Visible signs of MAHA include petechiae (little red spots on the skin) and spontaneous ecchymosis (bruising), particularly on the legs and torso. These indicate rampant microbleeds associated with platelet destruction and subsequent thrombocytopenia (low platelet count). Diagnostics will show low platelet counts and low RBC counts, as these cells are either destroyed (MAHA) or used up in the process of unfettered coagulation (DIC). Patients who developed MAHA will also show elevated fibrin degradation products (FDPs), elevated D-dimers, a high Prothrombin Time/International Normalized Ratio (PT/INR) and decreased fibrinogen (due to most being converted to fibrin). Of course, the physician will be able to see the schistocytes on blood smears.

Treatment of MAHA, as with many oncology interventions, includes being aware of what could happen. I like to call this pre-planning fortune telling, because, as the nurse, you are looking at what has been laid before the patient with the intention of divination. As front-line staff, we (as nurses) are also acutely aware of our responsibility to drive early and appropriate medical therapy. Safety of the MAHA patient is vital, and special care should be taken when helping the patient with ambulation and activities to avoid tissue injury via cuts and scrapes. In most cases, patients with MAHA will be admitted to hospital (if they are not there already), and transfer to the ICU should always be considered if they are unstable on acute care wards.

Anticoagulants are generally not an option, as quickly in the process leading to MAHA the coagulation factors, and platelets, are already exhausted. Platelet and RBC transfusions and fresh frozen plasma (which contain coagulation factors) may be ordered. In advanced cases, plasma exchange (PE) or renal dialysis may be considered. Intravenous fluid replacement, along with oxygen therapy, can provide immediate interventions meant to provide comfort and support to the patient. Stopping, or attempting to stop, free-flowing bleeding is important, and I have seen instances of patients with MAHA needing to have cocaine inhalation or intensive nasal packing in order to stop (or slow) epistaxis. Frequent lab work will be ordered, and patients should be monitored closely for new onset of symptoms. A new therapy called Xigris (drotrecogin alfa), a form of human-activated protein C that has profibrinolytic properties, could provide hope for DIC and MAHA patients.

Education of the patient and their support system as to what is happening and how the medical team is trying to address it is something nurses can embrace. Fear, anxiety, and frustration can exacerbate symptoms of MAHA. Unfortunately, prognosis for advanced oncology patients who develop MAHA is poor. The literature demonstrates a wide range of mortality rates, but general consensus seems to be about 70% for this patient population (Elliott et al., 2010, p. 49). Quick interventions, along with fortune telling, seem to be the best way to prevent and potentially resolve a run-in with MAHA.

References


