

Too Much?

As previously mentioned, I want to continue sharing some of the thought-provoking and relevant clinical scenarios that arise during my time on the inpatient lymphoma service.

One of my current patients is a 73-year-old man with chronic lymphocytic leukemia (CLL). Although he has experienced minimal progression of his CLL, he has demonstrated a collapse of his immune system. This patient was diagnosed with CLL when he presented with dyspnea that was found to be the result of *Pneumocystis pneumonia*. He subsequently developed autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia that responded to rituximab treatment, which was followed six months later by pancytopenia associated with parvovirus and cytomegalovirus (CMV) viremias. The CMV very quickly became difficult to control because he developed ganciclovir resistance and required foscarnet infusions. His CMV would be suppressed after an induction of foscarnet, only to relapse each time he was placed on maintenance treatment. The course of the parvovirus mimicked that of the CMV, recurring each time the intravenous immune globulin (IVIG) was held after achieving remission.

The degree of immune system dysfunction we saw with this patient, affecting the B- and T-cell lymphocyte compartments, is a classic clinical scenario for patients with CLL. Since his initial presentation, he has had two further relapses of AIHA. Unfortunately, his third episode has been extremely refractory to therapy and has required an excessive number of transfusions to maintain his hemoglobin above 7.0 g/dL. Because of his T-cell dysfunction, I have avoided using corticosteroids in his AIHA treatment. At this point, his condition has not responded to rituximab, high-dose IVIG, Bruton tyrosine kinase inhibition, or fostamatinib. Although he is well educated, having worked as an engineer in Russia before emigrating to the United States, he has fought me over every intervention.

Several weeks ago, I first recommended rituximab, cyclophosphamide, and dexamethasone (RCD) as treatment for the hemolytic anemia, but he refused this regimen based on general concerns about chemotherapy as well as a fear that his CMV and parvovirus would recur. He has received approximately 30 units of red blood cells so far and continues to receive an additional 2 units each week. He is easily encouraged each time he sees a slight, albeit temporary, improvement in his hemoglobin. Each time that I bring up the RCD regimen, he refuses, instead attributing his worsening condition to prostatitis, an upper

respiratory infection, or something else. With each unit of red blood cells, he increases the chance of being further alloimmunized and unresponsive to transfusions. Further increasing the risk from these transfusions is the fact that I have been unable to obtain insurance approval for him to receive oral iron chelation therapy. His reticence has sorely tested my patience, which I had always considered to be extremely good when it came to patient misbehavior.

In retrospect, I worry that I have harmed my patient by attempting to be understanding and allowing him to make misinformed and bad medical decisions. He is entitled to bodily autonomy, and I would never give chemotherapy to a patient who refuses it, but I can't help feeling that he might acquiesce if I were adamant and threatened to dismiss him from my practice. Ironically, the family worships me and is eternally grateful for everything I have done for them, all the while refusing my next recommendation. Could I argue for a court-appointed guardian given his inability to make sound medical decisions? This is not a patient opting to forgo medical care; he is more than happy to come to my office daily.

His case also raises issues related to resource utilization. The amount of IVIG he has received has been extremely costly. Regarding the number of red blood cell transfusions, I personally have donated 74 units of red blood cells throughout my life, and this one patient has used nearly half this amount. The supply of red blood cells in New York City has just a two- to three-day cushion. Should one individual be able to use so much of a limited resource when they are not utilizing available interventions?

My greatest fear is that the patient is going to have a bad outcome. I am worried that he will develop an inability to respond to additional transfusions, a complication of chronic hemolysis, or liver and heart damage from iron overload. I imagine that fear and lack of sophistication are driving his poor decision-making. Still, I continue to do my best for this patient and hope for a good outcome. I believe it is better to try and fail than to fail by not trying, but hope springs eternal. I can only hope I have not failed my patient.

Sincerely,



Richard R. Furman, MD

