Living with Sezary Syndrome

As shared by Mahanth J.

Chances are your network of close family and friends has been deeply affected by serious conditions such as cancer, heart disease, or diabetes regardless of who or where you are in the world. We are all in this together as members of the same species and a part of nature. Across borders, languages, climates, and net worths, our bodies are the vehicles that transport our consciousness through our physical reality for a finite number of years.

Like cars, trucks, ships, airplanes and spacecraft, human bodies are complex machines, except far more so. Robust health and quality of life require endless maintenance and major interventions because critical parts and systems break down over time. There is an entire universe of mysterious activity within us, constantly evolving, involving trillions of organic living cells. There's truly a Star Wars saga playing out at the microscopic layer with invisible visiting cells, both friendly and threatening, crossing the barriers daily without a passport to fight or cooperate. Even the most advanced researchers in any specialty of medicine have much to learn.

Like wealth, good health has to be earned constantly and requires preventive care and courageous struggle. Those of us in the smaller, more exclusive club of ultra-rare disease patients impacted by cutaneous lymphoma, including my own poorly understood subgenre called Sezary syndrome, might face the challenge of our lives.

Diagnosis

I was diagnosed on St. Patrick's Day 2023, though I probably suffered from the condition for at least 4 years before that day without knowing it for part or all of my 30s. Like many people afflicted worldwide by any of a variety of scarce skin diseases, I was incorrectly diagnosed and treated by a long parade of experienced medical staff as an eczema case. This is a pretty common (and nefarious) feature of cutaneous lymphomas (CLs) as eczema is a widespread autoimmune malady in the human population, which presents with most of the exact same visual and other symptoms, worst of all nonstop, unbearable itching that for me resulted in many sleepless nights for many years without complete relief from topical treatments. But eczema is seldom deadly, while Sezary Syndrome is extremely lethal, and the itching is even worse.

As the months and years went on the mutated Sezary cells spawned and spread rapidly to cover most of my skin, swim in increasing numbers through my bloodstream, and then lodge painfully into dramatically swollen lymph nodes in my neck, armpits, thighs, and groin by early 2023. The lymph nodal involvement triggered the diagnostic tests we needed to finally encounter a dreaded but accurate verdict. Skin biopsies, lymph node biopsies, and blood tests including one I'd never heard of before called Flow Cytometry were conducted, which taken together gave a snapshot of proliferating mutant cancerous T-cells whose nuclei were grotesquely enlarged and render these types of lymphocytes useless to the immune system. But they sure can reproduce fast and attack the healthy parts of the body.

It was a very shocking and frightening diagnosis for me and my family, made worse by my age at the time, 43, and my complete ignorance of Sezary syndrome. Sure, I'd heard of skin cancer, leukemia and lymphoma but I understood little about these cancers, and never could have imagined there was an ultra-rare subgenre within this world of scary ailments that combined all three of them into one single disease from Hell named after the dermatologist who discovered it in Paris, Dr. Albert Sezary, in 1934.

Until the answer came for me unexpectedly, out of the blue. In one day, everything changed. I didn't have eczema at all. I had something much worse, and it was already flowing freely throughout my body to cripple three different bodily systems. Only to be unearthed at Stage IV due to the extremely aggressive growth pattern of Sezary syndrome.

Accepting Uncertainty

At learning and digesting the bad news, I liked to think of myself as a brave and tough guy, but for a long time I didn't feel that way. For the first month I struggled with denial and thoughts of helplessness. I fought mightily, and failed, to keep up a persistent positive mindset for the next 6 months. I was addled with unhealthy doses of fear, anxiety, and self-pity over that period.

A battery of strong medications, chemotherapy, lifestyle changes, topical steroid ointments and creams, and brutal side effects followed starting in April 2023 with earnest hopes of beating down the lymphoma. Hundreds of hours were spent at uncountable hospital visits with numerous specialists and staff. Measurable progress was agonizingly slow and non-linear, but in November 2023, I had a good cry while sitting at an isolated beach in Cartagena, Colombia. I finally had a whole week off from treatments and work for my first post-diagnosis vacation to a place resembling paradise on Earth. After more tears were shed back at home, I finally accepted my unpleasant fate, acknowledged the trauma and dark uncertainty about the future, and the real possibility of my own death happening multiple decades before I had hoped for.

Throughout the ordeal I never completely gave up the fight through many unexpected ups and downs, although throwing up my hands in defeat was mighty tempting. In early 2024 my condition worsened and my body went the wrong way after the sustained progress I was celebrating towards the end of 2023.

If facing CL or any serious condition, you should expect the unexpected and be ready to take it on. That does not change if a disease is rare or poorly understood without established cause, or if things are going in the right direction for a long time. Do not get caught up in the factors out of your direct control. Focus on the choices you can make, medically and otherwise, to get better. That includes the effort to find doctors you trust with making major recommendations about life and death, while seeking out expert second opinions along the way. These efforts will keep your mind occupied and give your family confidence that you are doing the best you can.

As adult patients with our wits about us (which Sezary syndrome thankfully never took away from me), we have the agency to do independent research, ask tough questions, take calculated risks based on the existing knowledge and data, and follow the experts' high level advice before acting. Surrender to the choices you make and let the universe do the rest. This stuff is way, way too complicated for us to understand, even for top medical doctors and PhD scientists in the pharmaceutical industry, so make the plan, work the plan, and then reiterate when needed.

Throughout the last few difficult years, I also enjoyed family support, good company and reconnecting with old friends and relatives, some after many years, who were concerned about my health. I know in my heart that important social connections helped me recover. My version of CL did not take away the ability to enjoy a hot cup of coffee, a good movie or TV show, or a fine meal. I could still listen to my favorite music or curl up with a good book. In fact, while largely isolated with minimal in-person social life, I had plenty of spare time at home or in the hospital. I decided to make the most of it instead of wringing my hands.

Stem Cell Transplant

After long hours of consultation and independent research, and seemingly never-ending courses of other treatments including romidepsin, vorinostat, ECP and, mogamulizimab I decided to pursue a bone marrow stem cell transplant in November 2024, a risky and involved procedure which required great preparation and over 4 months entirely off from work for recovery.

During that buffer period many complications arose including Graft vs Host Disease (GVHD), yet another thing I'd never heard of. It was all made possible thanks to an anonymous 25-year-old female donor, a total stranger who registered with National Marrow Donor Program (NMDP) and donated her stem cells for completely altruistic reasons. I was very blessed to have found a suitable HLA typing match, which is not a given. Her selfless gesture not only rebuilt a new immune system and blood type in my body to attack the cancer remnants. Not only extended my life with complete remission that I still enjoy a year later. Her action restored my faith in humanity at a time when I really needed hope for myself and a wider society both undergoing dramatic turmoil.

But crisis always has been and always will be. It's how we deal with the challenges that matters. Fortunately, we are not alone, because we are all in this together.

Mahanth is open to connecting with others living with Sezary syndrome to talk about living your best life with the disease. Should you care to connect, please contact the Foundation at info@clfoundation.org.