Once-Twice

By Herbert C. Flessa, M.D.

Her illness began in the spring of 1983 when she began having episodes of blurred vision in her right visual field as well as occasional numbness of her right upper lip and tongue. A complete physical examination revealed no abnormalities. A blood count, however, showed a slightly elevated white count and a significant elevation of one of the white cells called the eosinophils. These latter cells, as you may know, are often associated with allergy. She became pregnant and had a normal delivery in mid 1984. The symptoms of numbness and so forth, however, continued. She was first seen at the University Hospital Medical Center in October of 1986. Here again the physical examination was normal. The white count, however, was a good deal higher. On examination of a well-stained peripheral blood film, the eosinophils, as noted previously, were bizarre appearing.

Further studies of the bone marrow, including a search for an abnormal chromosome sometimes seen in leukemia patients, were normal. It seemed likely that the neurologic symptoms were related to intermittent plugging of small blood vessels in the brain by the sticky eosinophils. Therapy with aspirin was instituted as well as a chemotherapy agent called hydroxy urea in an effort to reduce the stickiness and also reduce the numbers of eosinophils. Throughout 1987 and 1988 the patient's clinical condition remained relatively stable, but her physical examination changed a great deal. By November of 1988 her spleen, an organ for filtering blood cells was enlarging at a rather rapid pace. She also developed a heart murmur and another abnormal heart sound called a gallop indicating that there was involvement now of the heart with this eosinophilic illness. We added corticosteroids to her treatment program but these did very little in controlling her disease. Throughout the summer of 1989 we discussed with the patient and her husband, and sometimes other members of her family, the possibility of bone marrow transplantation. The patient, her husband and all members of her family agreed to push ahead with exploring transplantation as a therapeutic option. We asked family members to go to the Hoxworth Blood Center where blood samples would be taken from each eligible member looking for someone to match the patient's blood antigens. We

specifically told them that mothers, fathers and other more distant relatives would not be satisfactory because matches in that situation would not be possible. I received a phone call a day or two later to tell me that blood had been drawn on all the patient's brothers and sisters and that her father, not a candidate for bone marrow donation, insisted that his blood be drawn because he wanted to be involved. That problem was easily solved, I said, "Draw his blood and let's see what it looks like". Shortly after the blood drawing we learned that she had a perfect match of her blood with her one of her brothers. We continued to move ahead.

I asked the patient to visit with a friend of mine, a hematologist/oncologist at Ohio State University, Dr. Earl Metz, to see if he and his staff agreed with the possibility of bone marrow transplantation. I should add that the medical literature revealed no reported case of successful bone marrow transplantation with someone with this form of illness, basically an eosinophilic leukemia. In a day or two the patient went to Columbus and on the day of her visit I received a call from Earl, which said simply, "If you guys don't want to do the transplant we will". We obtained the brother's bone marrow samples, and after other detailed discussions decided to proceed with the transplant. In September of 1989 using a brother's perfectly matched bone marrow, transplantation was achieved. Three weeks after the transplant her blood count was returning to normal without eosinophils. Further testing indicated that the cells present in her blood were her brother's karyotype. Her post-operative course went relatively smoothly. She developed minor graft versus host disease with skin itching, some liver abnormalities and episodes of diarrhea, which cleared in about six months. In June of 1990 she developed disseminated herpes zoster or shingles. Fortunately this responded promptly and dramatically to the use of an intravenous antiviral called Acyclovir. She is now in her 15th year post transplant taking no medication. She has full time job. Her daughters are raised and life is good.

Addendum: To my knowledge this may be the only patient with eosinophilic leukemia who has been successfully transplanted.

In 1973, when not quite 50 years old, he developed a Guillain-Barre Syndrome. This strange, rare auto-immune illness presents not unlike polio myelitis with muscle weakness and sometimes numbness in the extremities (autoimmune: the body develops

antibodies against some of its own tissue). This disease slowly cleared over a period of weeks. In 1988 the patient sought advice regarding decreasing vision in his right eye. Shortly thereafter the vision in the left eye began to decrease. An ophthalmologist, Dr. Joel Sachs, then Chairman of the Department of Ophthalmology at the College of Medicine here in Cincinnati, diagnosed retinal leukemia, an uncommon complication of chronic lymphocytic leukemia. I was invited in on the case. Fortunately the patient responded to high doses of corticosteroids as well as huge doses of a chemotherapy agent given intravenously at infrequent intervals. His vision returned to normal. From 1989-1996 the patient did reasonably well despite slowly falling hemoglobin numbers, and slowly rising white counts. His spleen also became involved and despite our best therapeutic efforts, the disease was slowly getting out of control. In February of 1996 he was admitted to the hospital with severe right upper lobe pneumonia caused by the pneumococcus. The infection had spread to his blood stream. While in the hospital and on appropriate antibiotic therapy his clinical condition deteriorated. I felt obligated to tell the patient and the patient's wife that although recovery was possible it was becoming increasingly unlikely. The white count rose to 180,000 with practically no infection fighting cells (neutrophils). Much to my pleasant surprise after two weeks of extreme therapy his condition stabilized and began to slowly improve. As he improved, oxygen therapy was interrupted. He became afebrile. Slowly the infiltrate in his lungs began to clear. Some time in late March or perhaps early April as his convalescence continued, I noted that his white count was falling and by mid-May the white count approached normal, and by mid July the total number of white cells was normal as well as the so-called infection fighting cells or neutrophils. The lymphocytes were no longer the predominant cell in the peripheral blood. His spleen, which had reached the size of a large football, had receded. By early autumn his strength had improved enough to return to work as an engineer at Armco Steel in Middletown. During these past eight years he has had repeated physical examination, as well as repeated blood counts. His exams continue to be normal. His health is excellent. His blood counts show no evidence of leukemia. It has simply disappeared. He has had an intervening illness, namely cancer of the prostate, treated successfully with radiation therapy. He asked from time to time whether we should do more bone marrow tests looking for any residual evidence of leukemia. My reply is always the same, "Absolutely not."

An occasional patient with advanced cancer of any variety will have the cancer disappear for reasons that are not measurable. This patient is one of the lucky few.