A Case Report:

Living with Severe Aplastic Anaemia for 22 Years

A Patient Tells His Story

(Supplemented with Information from the Haematology Clinic Basel, Switzerland)

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Ernst Kienle: In 1977, feeling progressively tired, I was diagnosed with aplastic anaemia at the age of 22 years. My local doctor referred me to Prof. Speck in Basel, Switzerland, who took care of me until his death, in 1998.

I was given bone marrow (*Comment: E.K. was treated with a first course of antilymphocyte globulin, followed by an infusion of HLA-haplotype-matched bone marrow from his brother. At that time, and for the following years, antilymphocyte globulin was used to induce micro-engraftment of mismatched bone marrow*).

My blood values gradually normalised (*which, at the time, suggested that it was the combination of antilymphocyte globulin and mismatched marrow that induced autologous haematopoietic recovery*).

In 1984, as my blood values deteriorated, I was given a second course of antilymphocyte globulin, which was less successful than the first. Several treatments were then tried and my spleen was removed (*E.K. had become refractory to platelet transfusions, and splenectomy was performed at a time when platelet counts were very low*).

In 1985, I suffered painful haemolytic attacks and was diagnosed with paroxysmal nocturnal haemoglobinuria, which lowered my haemoglobin values still more. Erythropoietin (*which had been developed in the meantime*) transiently helped, so that I did not need any transfusions. Cyclosporin A was also tried (*but had to be stopped because of kidney damage and severe hypertension*). E.K.,

Fig. 1. Patient E.K., May 1999, 3 weeks after bone marrow transplantation from an unrelated donor.

with a remarkable will-power, learned to live with a haemoglobin value around 5 g%, doing regular physical exercise, refusing transfusions).

During that period, I felt really well. In 1998, I was diagnosed with myelodysplastic syndrome, and (*since the percentage of blasts increased rapidly*) I was given bone marrow in May 1999 (*from an unrelated donor, after con-*

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ditioning with VP16, cyclophosphamide and TBI. After very severe complications during the early post-transplantation phase, he rapidly recovered and was discharged 3 weeks after transplantation). Today, 4 months after transplantation, I feel completely healthy at the age of 44.

Without Prof. B. Speck, his team and my entire confidence, all this probably would not have happened. Prof. Speck took care of me and my disease for over 20 years, and I am greatly indebted to him.

Comments

E.K. was lucky

- to be diagnosed with severe aplastic anaemia at a time when antilymphocyte globulin had just become available as a treatment alternative for patients who did not have a matched sibling donor for bone marrow transplantation, and

- to survive a long period of pancytopenia until erythropoietin and cyclosporin became available, and matching techniques for selection of unrelated donors were improved.

He was wise to refuse transfusions over all the years, since, being previously heavily transfused, engraftment of the unrelated bone marrow could have been compromised, and iron overload or acquisition of hepatitis C could have caused irreversible organ damage.

The success of the unrelated bone marrow transplantation after a 22-year history of therapeutic trial and error was hardly predictable. The case of E.K. illustrates how being 'off protocol' can be a chance by provoking experimental treatment. For E.K., the development of new therapeutic tools and strategies has kept pace with the natural history of his disease. Though few patients may be as lucky, it is encouraging to know that in severe aplastic anaemia, even in an apparently desperate situation, the outlook is never hopeless.