SHORT REPORT

Relapse of chronic inflammatory demyelinating polyneuropathy 5 years after autologous stem cell transplantation

M Vermeulen, M H J van Oers

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We describe relapse of chronic inflammatory demyelinating polyneuropathy in a patient who had been in remission for 5 years after treatment with autologous stem cell transplantation. Before the transplant, he needed higher doses of immunosuppressive treatment than are now necessary to maintain his improved condition. He now receives intravenous immunoglobulins at monthly intervals.

In 2002, we reported on a patient with chronic inflammatory demyelinating polyneuropathy (CIDP) who had never had spontaneous remissions over a 10 year period, but who was in remission for 2 years after autologous stem cell transplantation (ASCT)[1]

Here we report relapse of CIDP 5 years after ASCT in this patient.

CASE REPORT

This patient was 38 years old in 1988 when he developed signs and symptoms which were later considered to be consistent with CIDP. He needed high doses of corticosteroids and intravenous immunoglobulin treatment to maintain his improved condition. Because of the side effects of this treatment, ASCT was considered. He was treated with BEAM myeloablative chemotherapy (1,3-bis(2-chloroethyl)-1-nitrosourea (BCNU), etoposide, cytosine arabinoside and melphalan) followed by stem cell reinfusion (CD 34+ cells).

Recovery of peripheral blood cells occurred within 16 days. Prednisone was tapered off but doses lower than 5 mg were accompanied by complaints of fatigue, probably due to adrenal insufficiency. He remained on this prednisone dosage and had no other immunosuppressive treatment. The only neurological sign that remained was mild numbness of his fingertips. He worked fulltime without limitations in his daily life activities.

Remission after ASCT lasted 5 years. His relapse started with tingling in his feet and hands. The sensory symptoms progressed to his forearms and upper legs, followed by diffuse weakness of the arm and leg muscles. He was no longer able to walk without assistance.

Electrophysiological studies were repeated and showed deterioration (table 1). Standard blood tests were repeated and were no different from earlier test results.

The signs and symptoms had progressed over 12 weeks when intravenous immunoglobulins were started. Initially he received 150 g in 3 days followed by 50 g at monthly intervals. With this treatment, the signs and symptoms improved. Within 10 days he was at the level he had been before his relapse. The main difference compared with the period before ASCT is that he now needs lower doses of immunosuppressive treatment which are better tolerated.

Table Electrophysiological studies before and after relapse

Right median nerve	Before	After
Distal CMAP (mV)	7.3	3.3
Distal latency (ms)	12.3	14.8
Nerve conduction velocity (m/s)	24	13

CMAP, compound muscle action potential.

DISCUSSION

We are not aware of other reports on ASCT treatment in CIDP. Improvement in the signs and symptoms of polyneuropathy after ASCT has been reported in five patients with POEMS syndrome.[2] This is a multisystem disorder with polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy and skin changes. This syndrome is associated with plasma cell dyscrasia. The pathogenesis of this syndrome is unknown but the symptoms are probably secondary to plasma cell proliferation.

There are several reports of ASCT treatment in patients with multiple sclerosis (MS).[3 4] Saiz *et al* reported 14 patients with severe MS who had been treated with ASCT. Three year probability of progression free survival was 86% and that of disease activity free survival was 46%. On MRI, the mean change in T2 lesion volume from baseline to the third year was 20%.[3]

In a similar study in 16 patients, 3 year progression free survival was 92% but 3 year disease activity free survival was less than 20%.[4]

The conclusion from the experience in patients with MS is that ASCT is not a definitive cure of MS but may change the aggressive course of the disease.

Similarly, ASCT is probably not a definitive cure for CIDP. This treatment may change the course of CIDP, but ASCT is a toxic and demanding treatment. The question is whether similar effects, as observed in our patient, can be obtained with less demanding treatments.

Authors' affiliations
M Vermeulen, Department of Neurology (H2-227), Academic Medical Centre, University of Amsterdam, Amsterdam, the Netherlands
M H J van Oers, Department of Haematology, Academic Medical Centre, University of Amsterdam, Amsterdam, the Netherlands

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Correspondence to: Dr M Vermeulen, Department of Neurology (H2-227), Academic Medical Centre, University of Amsterdam, PO Box 22700, 1100 DE Amsterdam, the Netherlands; m.vermeulen@amc.uva.nl

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Abbreviations: ASCT, autologous stem cell transplantation; CIDP, chronic inflammatory demyelinating polyneuropathy; MS, multiple sclerosis