

Type 1 cryoglobulinaemia presenting as digital ischaemia in chronic lymphocytic leukaemia

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DESCRIPTION

A 60-year-old Caucasian man with untreated chronic lymphocytic leukaemia presented with a 3-month history of recurrent arthralgia and joint swelling, predominantly affecting the knee, wrist and DIP joints. He subsequently developed painful, dusky fingers and was referred to the haematology department at the Royal Free Hospital for investigation (figure 1).

On admission, a full blood count revealed: Hb 92 g/L, white cell count $86.7 \times 10^9/L$ with a lymphocyte differential of $77.2 \times 10^9/L$ and platelets 332. Renal, liver and bone profiles within normal ranges. Cryoglobulins were detected, and subsequent immunofixation showed the presence of a type 1, monoclonal IgM lambda cryoglobulin.

The patient was managed with a continuous iloprost infusion, high dose corticosteroid therapy, plasmapheresis, rituximab and subsequently a dose of cyclophosphamide. On discharge, he was referred to University College Hospital for long-term management of CLL. He was referred to plastic surgery for amputation of affected digits (figure 2).

Type 1 cryoglobulinaemia is a rare disorder characterised by monoclonal production of immunoglobulins that precipitate at cold temperatures and can cause microvascular damage. The damage in this case led to end tissue damage and significant changes to the patient's quality of life.¹ It can manifest in a number of ways, and it is therefore



Figure 1 Before treatment.



Figure 2 After treatment, showing halting of necrosis. important to appreciate the accompanying underlying disorders that can drive it.

Learning points

- ▶ Type 1 cryoglobulinaemia is a rare disorder characterised by monoclonal production of immunoglobulins that precipitate at cold temperatures and can cause microvascular damage.
- ▶ It makes up 10%–15% of cryoglobulinaemias and is typically associated with lymphoproliferative disorders.
- ▶ Response to treatment is often monitored using cryocrit percentage or paraprotein quantification.

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