A young woman with massive weight loss, neuropathy and cardiopathy

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In December 2000 a 45-year-old woman presented with a history of fatigue, light-headedness, nausea, vomiting, poor appetite, early satiety, diarrhoea and substantial weight loss (18 kg in the last 6 months). She had been complaining of progressive dyasiaesthesia and paraesthesia of the lower extremities for the previous eighteen months. On admission the physical examination revealed facial purpura particularly around the eyes (fig. 1), oral submucosal bleeding spots and macroglossia (fig. 2). Blood pressure was 90/50 mm Hg and a symptomatic postural hypotension was detectable. Vibration sensation was reduced in the lower extremities and the Achilles tendon reflex was symmetrically impaired.

Laboratory tests showed hypalbuminaemia of 21 g/l (30–52 g/l), total proteins 59.8 g/L (60–82 g/L) low iron of 6 mmol/l (6–30 mmol/l), transferrin saturation of 11% (15–50%) and an erythrocyte sedimentation rate of 10 mm/h. The following investigations were normal, negative or non-diagnostic: full blood count, liver function tests, lactate dehydrogenase (LDH), sodium, potassium, calcium, phosphate, creatinine, thyroid stimulating hormone (TSH), clotting factor X, prothrombin time and anticardiolipin antibodies. Immunoelectrophoresis of the serum showed a monoclonal gammapathy of IgG lambda type (14.5 g/L). Proteinuria was 1.18 g/24 h (<0.15 g/l) consisting predominantly of monoclonal IgG lambda and free lambda light chains (0.05 g/24 h). Bone marrow aspirate and biopsy revealed 10–15% (in certain areas up to 30%) plasma cell infiltration.

An abdominal ultrasonography detected no abnormality. The gastro-oesophageal endoscopy revealed clearly reduced gastric motility with food retention and a moderate gastritis, positive for \textit{H. pylori}. Radiographic studies of the upper gastrointestinal tract to demonstrate gut paresis were not performed. Biopsy of both stomach and rectum showed evidence of massive amyloidosis with predominant vessel-associated deposition (fig. 3) of immunohistological type IgG lambda (fig. 4).

An echocardiogram revealed cardiac involvement with concentrically thickened ventricular walls and restrictive physiology, as typically observed in light chains (AL)-amyloidosis. Left ventricular ejection fraction was 57%.

Figure 1
Spontaneous periorbital purpura, giving the typical “raccoon-eyes” sign.

Figure 2
Macroglossia in amyloidosis. The tongue is frequently rimmed by the indentation of teeth.
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Figure 3
Congo red staining showing massive perivascular amyloid deposition.

Figure 4
Immunohistochemical staining for lambda light chains.

References
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