Successful treatment of systemic AA amyloidosis associated with underlying Hodgkin lymphoma

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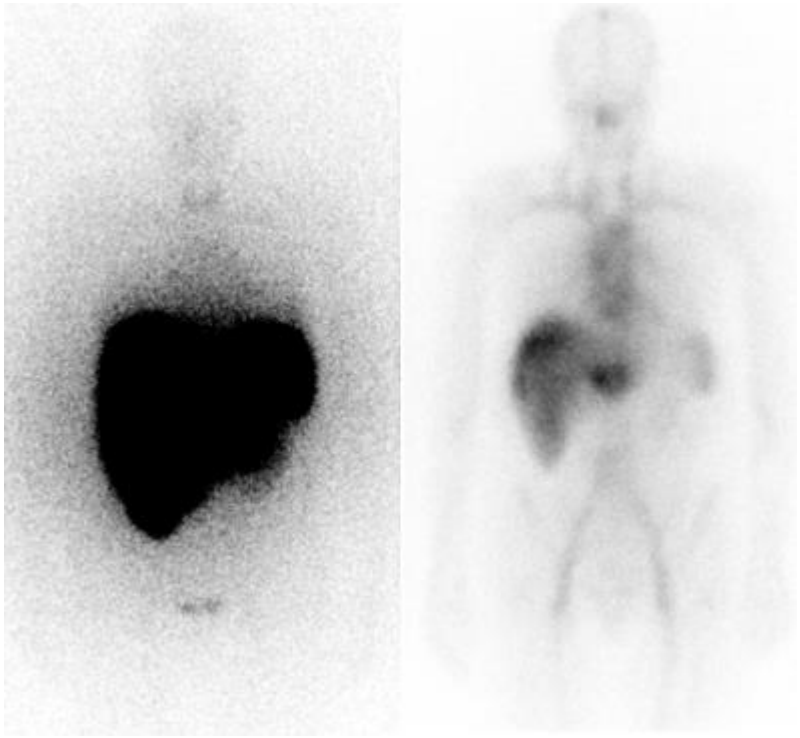
A 47-year-old female with a history of breast carcinoma in remission was found to have iron deficiency anaemia, deranged liver function tests and nephrotic syndrome on routine testing. Further investigations revealed a mediastinal mass, biopsy consistent with classic Hodgkin lymphoma (nodular sclerosis subtype). Positron emission tomography-computed tomography (PET-CT) demonstrated fluorodeoxyglucose (FDG)-avid Stage 2 nodal disease and non-FDG avid massive hepatomegaly. Gastric and colonic biopsies revealed AA amyloid deposition. ^{123}I-labelled serum amyloid P component (SAP) scintigraphy (a sensitive, specific imaging modality for visceral amyloid deposits) demonstrated a large amyloid load in the liver and spleen, obscuring the kidneys (left). There was no evidence of cardiac amyloidosis by echocardiography, and she had normal serum NT-proBNP and cardiac troponin T levels. Her inflammatory markers were markedly abnormal: C-reactive protein (CRP) 121 mg/l (normal range 0–5), serum amyloid A (SAA) 296 mg/l (normal range < 10), with no evidence of infection. The patient was treated with six cycles of doxorubicin, bleomycin, vinblastine and dacarbazine to a PET Deauville 2 response. She received reduced dose (50%) vinblastine due to risk of paralytic ileus in gastrointestinal amyloidotic involvement. She developed bleomycin-induced lung toxicity, which has resolved.

After one chemotherapy cycle, CRP and SAA levels normalised and have remained within satisfactory limits. Her proteinuria and serum albumin have improved over time. Now, four years since diagnosis, her liver function tests, urinary protein and serum albumin levels are normal, and her estimated glomerular filtration rate is 56 ml/min. She has no evidence of recurrence of Hodgkin lymphoma or breast carcinoma. SAP scintigraphy shows substantial regression of amyloid deposits (most recent imaging, right).

AA amyloid fibrils arise from misfolding of the acute-phase reactant, SAA, in patients with SAA overproduction. Curiously, systemic AA amyloidosis occurs in only a small proportion of patients with chronic inflammatory disorders, the most frequent of which is inflammatory arthritis. Rare causes include vasculitis and malignancy, although systemic AA amyloidosis associated with Hodgkin lymphoma is exceptionally rare. This case demonstrates regression of AA amyloid deposits on SAP scintigraphy and improved function of amyloidotic organs following successful treatment of underlying Hodgkin lymphoma, in a patient with previous breast carcinoma.

Figure 1

a) b)



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