81 Case 81

A 55-year-old man with a history of recently treated stage IVB diffuse large B-cell lymphoma presented some 8 months after completion of R-CHOP chemotherapy. His initial presentation had included bone marrow involvement and extranodal disease, both known risk factors for CNS relapse. The best approach to prevention of CNS relapse of occult CNS disease at presentation remains a topic of intense debate. His new complaint was of progressive weakness in his lower limbs with difficulty walking. There was no significant bladder or bowel disturbance.

On examination he appeared well and was without palpable lymphadenopathy or organomegaly. The most notable feature was of a lower limb paresis and sensory disturbance without a clear sensory level. His leg muscle groups were wasted and lower limb power was impaired. As he had been treated with vinca alkaloid therapy he was generally hyporeflexic rendering this modality of assessment less reliable. He reported no significant lower limb pain.

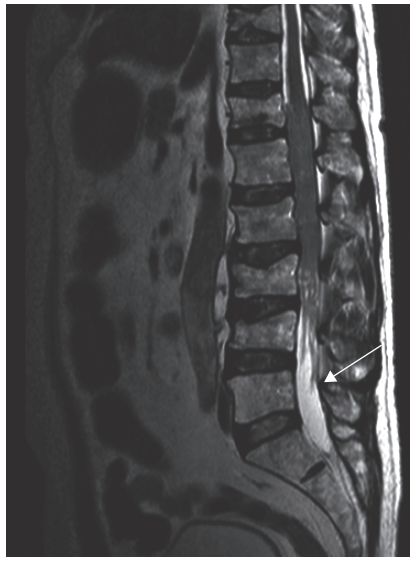

Figure 81.1 MRI.

Laboratory investigations

The FBC and blood film were unremarkable.

U&Es, LFTs, bone profile, LDH and CRP were all normal.

A bone marrow aspirate (being a site of disease at presentation) showed no abnormal lymphoid population, or other significant abnormality.

Imaging

Lower spinal cord pathology was suspected. MRI imaging showed a lesion affecting the lower cord and conus medullaris (arrow, Figure 81.1); this would explain the clinical presentation, although the sparing of bladder and bowel function was somewhat unusual. CT imaging of the chest, abdomen and pelvis showed no significant abnormality. PET/CT imaging highlighted an area of increased isotope uptake in the terminal spinal cord correlating with the abnormal area seen using MRI (arrow, Figure 81.2). No other areas of abnormal uptake were evident.

Flow cytometry

In view of the localised area of possible disease relapse and after considering the implications of neurological

Practical Flow Cytometry in Haematology: 100 Worked Examples, First Edition. Mike Leach, Mark Drummond, Allyson Doig, Pam McKay, Bob Jackson and Barbara J. Bain.

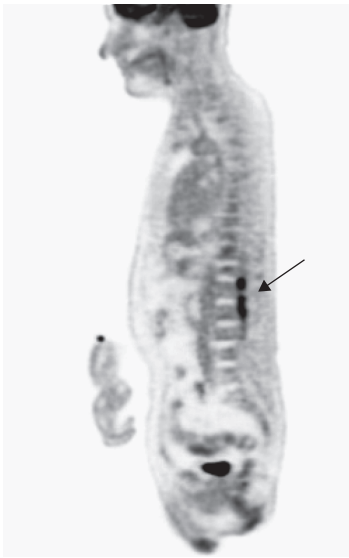

Figure 81.2 PET/CT.

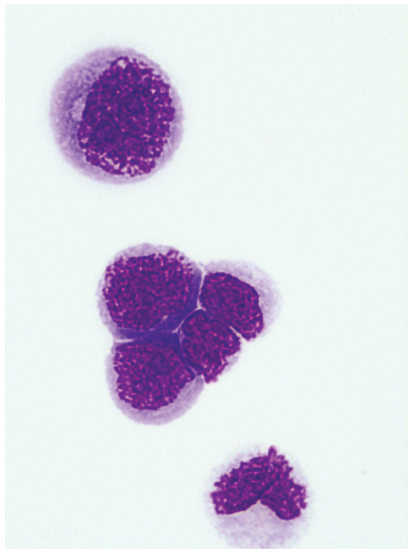

Figure 81.4 MGG, ×1000.

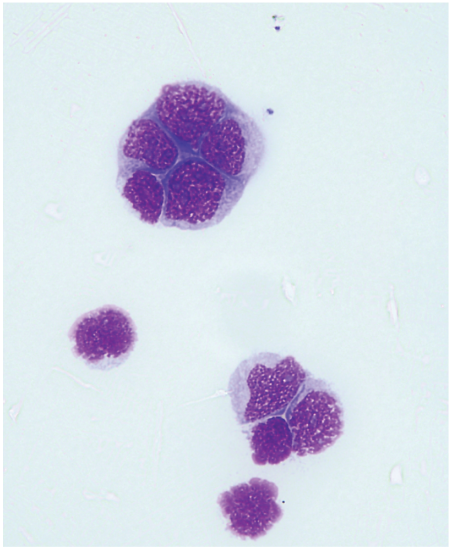

Figure 81.3 MGG, ×1000.

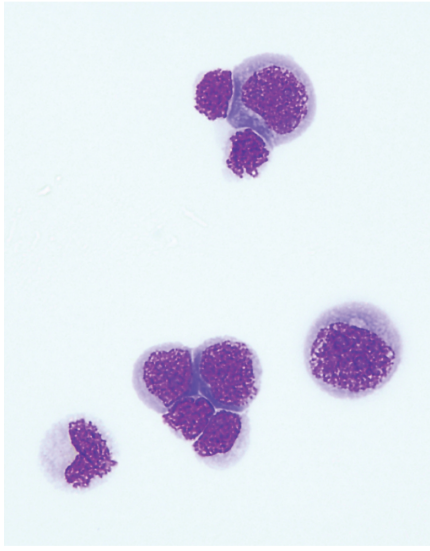

Figure 81.5 MGG, ×1000.

decompensation we proceeded to a radiologically guided L5 approach lumbar puncture to acquire a CSF specimen. No other tissue was available for a definitive diagnosis. A cytospin preparation showed a population of pleomorphic large cells with a minor reactive lymphocytic response. Flow analysis using a CD2/CD19 gated approach, focussing on the large cells, identified a significant excess of B cells. The total cell count was 0.16×10^9/L.

The B-cell population was abnormal, showing kappa surface light chain restriction; the phenotype was mature with expression of CD19, CD20, FMC7, CD22 and CD79b. The cell morphology was in keeping with a lymphomatous population (Figures 81.3–81.6).

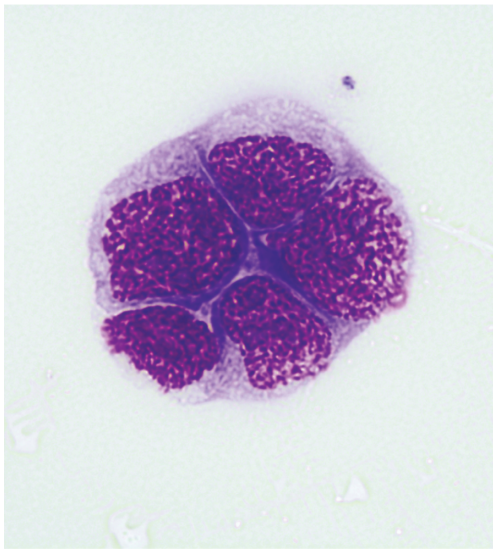

Figure 81.6 MGG, ×1000.

Discussion

This case illustrates the rare and unfortunate CNS relapse of diffuse large B-cell lymphoma. The case is even more unusual in that relapse occurred within the terminal part of the spinal cord. Despite the highly unusual features, a relapse was suspected in view of the prior history, the new neurological presentation and the findings on imaging. Morphological and flow cytometry analysis of CSF allowed a definitive confirmation which informed the subsequent treatment plan.

Final diagnosis

Isolated lower spinal cord and intraconal CNS relapse of diffuse large B-cell lymphoma.