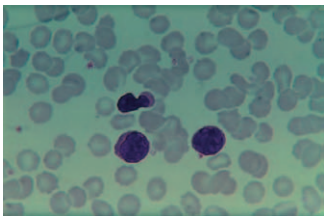
Case 6.8 Sézary syndrome

A 66-year-old man presented with generalized itching and redness of his skin for 2 months, with severe keratosis on the soles of his feet and the palms of his hands. On examination, he had characteristic exfoliative dermatitis with bilateral axillary lymphadenopathy but no hepatosplenomegaly. Investigation showed that his haemoglobin was normal (139 g/l) but he had a raised white-cell count (12.8×10^9/l). A blood film showed an increase of small cleaved lymphocytes, 90% of which were T lymphocytes (see Table C6.8). Electron microscopy of buffy coat cells confirmed that the nuclei of these cells had multiple clefts, an appearance characteristic of cells known as Sézary cells. The patient had early-stage *Sézary syndrome* (see Case Figure 6.8) and was treated with psoralens and ultraviolet A therapy (PUVA) to the skin.

Table C6.8 Immunophenotyping in 6.8 and 6.9

	Lymphocyte marker*								
	Surface membrane immunoglobulin (SIg)								
Case	**kappa**	**lambda**	**mu**	**gamma**	**alpha**	**CD3**	**CD19**	**CD19 and CD5**	**Diagnosis**
Normal/reactive lymphocytosis	7	4	6	3	2	75	12	2	Normal/reactive lymphocytosis
Case 6.8	1	1	2	0	0	92	2	0	Sézary syndrome
Case 6.9	60	2	60	1	0	10	60	0	Hairy cell leukaemia

*Results expressed as percentage of peripheral blood lymphocytes positive for marker.



Case Figure 6.8 Sézary cells. The lymphocytes are cleaved and have an overlapping nuclear conformation.

Essentials of Clinical Immunology, Sixth Edition. Helen Chapel, Mansel Haeney, Siraj Misbah, and Neil Snowden.

Case 6.9 Hairy cell leukaemia

A 48-year-old asymptomatic man was found to have gross splenomegaly on a routine life-insurance examination. Investigation showed a normal haemoglobin but a mildly raised white-cell count (14.2×10^9/l). On the blood film these cells were mainly small mononuclear cells resembling lymphocytes, but had a spiky or 'hairy' appearance, had B-cell markers on their surface and were positive for kappa but not lambda light chains (see Case Table C6.8). They stained positively for tartrate-resistant acid phosphatase, an enzyme characteristically found in the cells of *hairy cell leukaemia* (see Case Figure 6.8). Bone marrow aspirate was 'dry' and trephine showed a characteristic infiltrate.

Splenectomy is now reserved for those with pancytopenia or splenic infarction. This patient was treated with pentostatin, which is the treatment of choice for hairy cell leukaemia. He has remained well for 5 years. Interferon or cladribine are also used in some countries, with good effect in some cases; stem cell transplantation is under consideration.

Table C6.8 Immunophenotyping in 6.8 and 6.9

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