**Follicular mucinosis, mycosis fungoides, and acute myeloid leukaemia**

Follicular mucinosis is a skin condition characterised by papulo-follicular lesions and histologically by mucinous degeneration of the hair follicles. Alopecia and lymphoma may occur, and often mycosis fungoides may develop subsequently. In published series, totalling 153 patients, progression to lymphoma occurred in 20%.

### Case report

A 60 year old man was admitted with an itchy papulo-nodular rash, multiple follicular abscesses, and alopecia totalis. Small lymph nodes were palpated in the neck only and there was no hepatosplenomegaly. Previous medical history included alcohol abuse and pulmonary tuberculosis. Investigations showed haemoglobin at 9.8 g/l, white cell count at 4.2 x 10^9/l with normal white cell proportions, and a platelet count of 202 x 10^9/l. Red cells were dimorphic, with normocytic and hypochromic forms. Skin biopsy specimens showed mucinous degeneration around hair follicles characteristic of follicular mucinosis. Atypical lymphoid cells were also seen in the dermis and infiltrating the epidermis (figure), with a lymphoid infiltrate also seen at the epidermal-dermal junction, as found in mycosis fungoides.

### Comments

This patient had follicular mucinosis, mycosis fungoides, and acute myeloid leukaemia. The association between the two skin disorders is well recognised, but there is only one report linking dysmyelopoiesis with the Sézary syndrome. More generally, an association between lymphoproliferative and myeloproliferative disorders is now recognised.

In this patient the presence of H inclusion in the red cells indicated leukaemic transformation of an early progenitor or stem cell. It seems unlikely that the skin and marrow disorders were unconnected. Primitive myeloid cells were present in the dermis; in the bone marrow the normal predominance of CD8 compared with CD4 lymphocytes was replaced by a CD4:8 ratio of 11:1. This suggests that an abnormal lymphoid proliferation was present in the bone marrow in addition to acute myeloid leukaemia.

H W HABBOUTH, N P LUCIE, Department of Haematology, R M MACKIE, J ASHWORTH, M TURBTT, Department of Dermatology, Western Infirmary, Glasgow G11 6NT


A computed tomography scan showed hiliar, splenic, paraaortic and para caval lymphadenopathy suggestive of lymphoma. No lymphomatous infiltration was seen on bone marrow examination, but there was noticeable dyserythropoiesis and abnormal megakaryocytes as well as 39%, myeloblasts and 21%, lymphocytes. Ringer sideroblasts were not found. The blasts were myelomonocytic; Sudan black and naphthol acetate esterase positivity confirmed a myelomonocytic leukaemia (FAB; M4). Chromosome analysis of the bone marrow showed 66%, abnormal mitoses with monosomy p11-p12, 47-48 or more chromosomes, breakages and rearrangements. Although haemoglobin electrophoresis was normal, haemoglobin H inclusions were detected after incubation with Brilliant cresyl blue.

Immunofluorescence studies of the bone marrow showed that the blast cells were positive for the myeloid markers CD13, 14, and 33. In the mononuclear layer 52% of cells were positive for lymphoid markers—that is, CD19 (B cell) 26%, CD2 (T cell) 26%, of which 29%, were CD4 and 31%, CD8 positive. Immunoperoxidase studies on the skin biopsy specimens showed CD4 positivity at the epidermal-dermal junction, characteristic of mycosis fungoides. CD13 and 14 positive cells were also present in the dermis. These probably represented blast cells as morphologically recognisable neutrophils and monocytes were scarce.

One week after diagnosis myeloblasts were seen in the blood in rapidly increasing numbers. Remission induction was attempted with a standard seven day regimen (daunorubicin, cycosine, arabinoside and thioguanine). He had a succession of infective episodes and died subsequently after completing chemotherapy; a post mortem examination showed fungal pneumonia.

H W HABBOUTH, N P LUCIE, Department of Haematology, R M MACKIE, J ASHWORTH, M TURBTT, Department of Dermatology, Western Infirmary, Glasgow G11 6NT