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. Alopeia 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/dl, 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. A computed tomography scan showed hilar, splenic, paracolic and paracaval lymphadenopathy suggestive of lymphoma. No lymphomatous infiltration was seen on bone marrow examination, but there was noticeable dyserythropoiesis and abnormal megalocytes as well as 39%, myeloblasts and 21%, lymphocytes. Ringed sideroblasts were not found. The blasts were myelomonocytic. Sudan black and naphthol acetate esterase positivity confirmed a diagnosis of acute myelomonocytic leukaemia (FAB M4). Chromosome analysis of the bone marrow showed 66%, normal mitoses with hyperdiploidy, 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 monoclonal layer 52%, of cells were positive for lymphoid markers—that is, CD19 (B cell) 26%, CD2 (T cell) 26%—of which 22%, were CD4 and 8%, CD8 positive. Immunoperoxide studies on the skin biopsy specimens showed CD4 positivity at the dermo-epidermal 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, arbinoamide and thioguanine). He had a succession of infective episodes and died subsequent to completing chemotherapy; a post mortem examination showed fungal pneumonia.

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 inclusions 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.

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Fecal leukocytes in Escherichia coli 0157:H7 enteritis

The cases and histopathology described by Hunt et al continue to broaden the disease spectrum which we believe is associated with Escherichia coli 0157:H7.

Our prospective studies, however, contrast with their suggestion that absence of neutrophils in the stool may help distinguish the haemorrhagic colitis associated with E coli 0157:H7 from bacterial dysentery or inflammatory bowel disease.

We examined the stools of 33 children that yielded E coli 0157:H7 on culture when associated with a diarrhoea illness. Twelve of eight patients had diarrhoea alone and five had already progressed to haemolytic uraemic syndrome. Stool smears were fixed in methanol and stained with methylene blue. Polymorphonuclear leucocyte counts greater than or equal to 5 per high power field (1000 x) in at least four fields were considered positive.

Sixteen of 35 (45%) stools specimens obtained over one to 10 days after onset of illness were positive. All 16 positive stools contained blood and mucous macroscopically. Stools which were non-bloody or semiformed did not contain clinically important numbers of fecal leukocytes. Within one to two days, three to four days, or more than five days of onset of symptoms, six of 14, nine of 14, and one of seven specimens were positive respectively.

Although the incidence of positive examinations for fecal leukocytes should vary according to microscopic criteria and timing of stool specimens, our findings proposes an incidence approaching that seen in other bacterial diarrhoea secondary to Campylobacter, Salmonella, and Shigella. While the predictive values for E coli 0157:H7 enteritis of a stool which is positive or negative for fecal leukocytes would not be very high, the presence of fecal leukocytes certainly does not favour other bacterial diarrhoeas or inflammatory bowel disease. Rather, E coli 0157:H7 enteritis in its advanced form should be regarded as a subset of the bacterial dysenteries.

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*J Clin Pathol* 1990 43: 347
doi: 10.1136/jcp.43.4.347-a

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