Summary

Thirteen cases of dystrophia myotonica are reported in brief. All presented some radiological abnormalities of the skull. The most constant changes were a small to very small pituitary fossa, thickening of the calvarium, and hyperostosis interna. There was enlargement of the sinuses in some and in two the mandible was elongated. We believe the high incidence of these changes cannot be coincidental, and it is held they should now come to be accepted as some of the variable features of dystrophia myotonica.

It is a pleasure to thank various physicians who have given me access to their cases, and Dr. Alexander and Dr. Rolleston for reviewing the radiological reports.

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BULLOUS LESIONS IN LEUKAEMIA

BY

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The skin manifestations of leukaemia are many and varied: Arzt and Fuhs (1929) have mentioned eczematous, erythema-multiforme-like, purpuric, zosteriform, urticarial, bullous, and rupial lesions, in addition to exfoliative erythrodermia and the specific leukaemic deposits occurring in the form of small papules, nodules, or large tumours.

Wiener (1947) states that bullous eruptions may precede generalized exfoliative erythrodermia in lymphatic leukaemia, and that they are often encountered in the early stages of monocytic leukaemia. It would seem that remarkably few cases of leukaemia with bullous lesions have, in fact, been recorded. In an analysis of a series of 445 cases of the reticulosos (including 160 cases of leukaemia) at the Los Angeles County Hospital, Epstein and MacEachern (1937) reported cutaneous involvement in 194 (43.5%). Bullous or vesicular lesions were seen in six cases, of which three were Hodgkin's disease, two myeloid leukaemia, and one lymphosarcoma. No bullous eruption was seen in any cases of lymphatic, monocytic, or acute leukaemia.

Margarot, Rimbaud, and Roche (1935) reported the case of a boy of 12 years, who was first seen with a purulent bullous eruption on the lower limbs, associated with pyrexial attacks. It was only when, five months later, he developed a generalized erythrodermia associated with palpable lymph nodes that a blood count revealed a leucocytosis of 34,000 per c.mm., of which 91% were primitive cells of the lymphatic series. The diagnosis of acute leukaemia was confirmed by the subsequent course of the disease, with the development of ulcerative stomatitis and a severe bleeding tendency, and death within four months of establishment of the diagnosis.

Remenovsky (1935) described the case of a 51-year-old patient with an acute leukaemia (leucocytes 83,600 per c.mm., 91.7% immature lymphocytes). After systemic arsenic the blood picture changed dramatically, the total leucocyte count falling to 6,000 per c.mm., 40% being lymphocytes. At the same time bullous eruptions developed on the wrists and forearms.

It would seem that the few recorded cases of bullous dermatoses have been associated with acute leukaemia. The following case of chronic lymphatic leukaemia is therefore of particular interest because of several unusual features: the manner of presentation as a bullous eruption on the palms and soles; the remarkable bluish infiltrated erythematous eruption on the face; and the presence of four other skin manifestations—namely, purpura, erythema-multiforme-like lesions, cellulitis, and the specific papular infiltrate.

Case Report

The patient, a married woman aged 65, first reported to the skin clinic at the Norfolk and Norwich Hospital on July 26, 1950. She gave a history of a bullous eruption which started one month previously on the right sole, later involving the left sole and the left palm. She had had a sore throat at the onset. There was no relevant previous history.

On examination bullae up to 1 cm. or more in diameter were scattered over the soles of both feet. The dorsa of the feet and webs of the toes were quite normal. There were a few confluent bullae on the left thenar eminence. All lesions contained clear fluid at this stage. General examination revealed no abnormalities. Scrapings taken from the feet showed no evidence of fungus infection.

In the absence of a history of possible contact irritants, it was thought that the eruption was in the nature of a bullous pompholyx, and treatment was given in the form of baths of potassium permanganate, and phenobarbitone, $\frac{1}{2}$ gr. (32 mg.) thrice daily. The local treatment was changed after a week to Castellani's paint, and later to 0.5% gentian violet. No progress was made, and after a month both palms were severely involved.

In view of the discomfort and the lack of response to treatment, the patient was admitted on October 4. By this time bullae had begun to appear on the dorsa of both feet, and some haemorrhagic elements were present (Figs. 1 and 2). Glands were palpable in both axillae and groins, but the spleen was not felt.

A blood examination on October 11 showed a total leucocyte count of 79,800 per c.mm., of which 91% were lymphocytes. On November 3 her leucocytes had gone up to 123,200 per c.mm., of which 90% were lymphocytes, 4% monocytes, and 6% polymorphs. (Her haemoglobin at this stage was 98%.) No primitive cells were seen. Large numbers of smear cells were present in stained films. The platelets were not accurately estimated at the time, but the impression was that they were reduced. A sternal marrow examination on November 15 confirmed the picture of chronic lymphatic leukaemia.

A course of superficial x rays (to a total of 450 r) produced a dramatic effect on the bullae. After four weeks the hands were completely normal again and there were only a few small lesions on the dorsa of the feet.

During the period in hospital she developed cellulitis of the legs; this responded rapidly to penicillin.

One week before discharge on November 24 a few scattered pinhead-sized erythematous papules had developed on the anterior surfaces of the knees and thighs. When she was seen in the out-patient department on December 7 the hands were still normal but new purulent bullae had appeared on the soles. Both feet showed a diffuse erythema

FIG. 1.—The dark pigment is not due to haemorrhage but is gentian violet.

FIG. 2.—Showing the bullous lesions that developed later on the dorsa of the feet in addition to the soles.

and both legs were oedematous and very tender as far up as the knees. Large glands were palpable in both groins. It was considered that the oedema was partly inflammatory and partly due to pressure by glands on the deep veins. A few bluish infiltrated nodules were present around the right nipple, and some pinhead-sized erythematous papules over the upper part of the sternum; there was also a diffuse erythema of the forehead. The spleen was palpable 3 in. (7.5 cm.) and the liver 1 in. (2.5 cm.) below the costal margin. A radiograph of the chest revealed the presence of mediastinal glands.

A blood count on December 7 showed: haemoglobin, 82%; leucocytes, 169,000 per c.mm. (lymphocytes 95%, monocytes 1%, polymorphs 4%).

It was decided to irradiate the glands in the axillae, groins, and mediastinum.

Progress

December 20.—The oedema of the legs had increased, and many punctate purpuric lesions were seen, especially over the shins. The most striking feature was the indurated erythema of the face, which had now spread on to the cheeks; x-ray treatment was prescribed for this. The leucocytes numbered 160,000 per c.mm.

January 16, 1951.—Bullae reappeared on the hands and feet. Purpuric lesions were seen on the inner aspects of the right thigh and over the shins. Subconjunctival haemorrhages occurred. The general condition had clearly deteriorated, and there was evidence of much loss of weight.

February 20.—The patient was readmitted to hospital. The facial appearance was most remarkable: a livid purplish erythema involved the whole face except for the orbit. Ringed macules, not unlike the target lesions of erythema multiforme, were seen on the trunk. Many infiltrated bluish papules were scattered over the front of the chest and abdomen. These were almost certainly specific leukaemic infiltrations, but in view of the patient's general condition a biopsy was not carried out. All superficial lymph nodes were enlarged. The spleen extended to below the umbilicus. A blood count showed: haemoglobin, 60%; total leucocytes, 265,000 (lymphocytes 97%, polymorphs 3%). The bleeding-time was 5 minutes 45 seconds; the coagulation time 3 minutes 32 seconds. The platelet count was 60,000 per c.mm.

February 23.—Pharyngitis, with systemic upset, and copious vomiting were present. The mouth was painful, with multiple ulcers. A swab from the throat showed a few haemolytic streptococci.

March 12.—She now had a gastro-intestinal upset, with haematemesis and diarrhoea. Pathogenic organisms were not isolated from the faeces.

March 20.—Her general condition was deteriorating rapidly. The skin lesions, however, were fading noticeably. The bullae on the palms and soles had practically disappeared.

March 26.—The patient collapsed with abdominal pain and died a few hours later. This was thought to be probably due to a severe internal haemorrhage. A post-mortem examination was not made.

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Last year the New York State Legislature adopted an amendment excusing every pupil from "such study of health and hygiene as conflicts with the religion of his parents" (*New York Times*, January 6). This amendment, sponsored by the Christian Science Church, did not attract much attention until recently, when the New York Academy of Medicine and various teaching groups in the State demanded its repeal. They maintain that it would set back the cause of medicine and health; on the other hand the Christian Scientists state that the exemption of such children from health courses should not affect the examination questions which other children are required to answer. Their officials say that their only concern was that children of their faith whose parents request it should be permitted not to study disease symptomatology and the germ theory of disease causation. Every phase of "positive" health and hygiene is being studied by the children of Christian Scientists.