Section of Dermatology

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[*January* 15, 1953]

Sclerema Neonatorum.—R. T. BRAIN, M.D.

The patient, H. S., was a female infant aged 5 weeks when brought to the Hospital for Sick Children, Great Ormond Street. She had been a full-term baby delivered after a normal labour lasting about seven hours and the birth-weight was 8 lb. The mother's pregnancy had been uncomplicated and it was unlikely that she had had any dietary deficiencies. About the third or fourth day of the infant's life an extensive dusky bluish area was noticed on the back, extending from the lower rib margins to the base of the neck and in the lower part of this area two large swellings were observed (Fig. 1). The infant was breast fed and her general condition and nutrition were good.

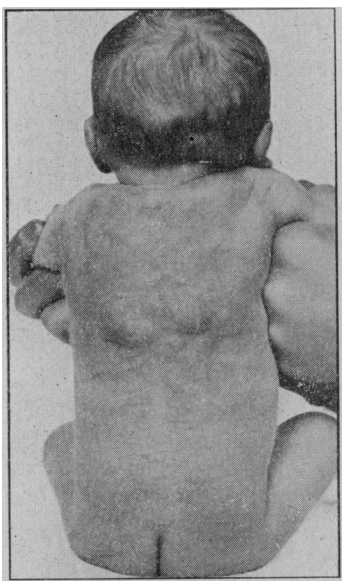

FIG. 1.—H. S., aged 6 weeks, showing two cystic swellings.

Examination.—The infant was sturdy and healthy and the only abnormal feature was that the skin across the back and shoulders was of a brownish-red colour with an irregular nodular surface, here and there causing coarse creases near the edges. In the lower part on each side of the mid-line were two soft fluctuating communicating cysts 3–4 cm. in diameter. When the centres of these soft swellings were depressed with the fingers, a firm collar of indurated subcutaneous tissue was felt to surround the base of each. This induration was continuous over the affected area and resembled a waxen plaque approaching 1 cm. in thickness. It was possible to insert the finger-tips beneath the well-defined edges, especially near the neck, and it seemed that the induration involved the hypoderm. The skin was firmly attached to it but was otherwise relatively normal in texture.

On aspiration of the cyst a thick creamy sterile fluid was obtained containing no cells but only fat droplets and doubly refractile crystals.

X-ray of soft tissues showed no calcification.

Biopsy.—The fat excised from the affected area was hard, opaque, white and avascular. Sections (Dr. M. Bodian): "The cutis shows only minimal changes, slight œdema and very little cellular infiltration. There is some variation in the size of fat cells which have, on the whole, preserved their outline. Needle-like crystals lie in the fat (and some outside) mostly in radiating tufts and some singly. Whenever they lie outside the cells they are colourless but within the fat cells they appear to take up the stain for neutral fat; this, however, is probably spurious. A granulomatous infiltrate in the fibrous septa around the fat lobules spreads also into the lobules. This consists of histiocytes, lymphocytes and polymorphonuclear leucocytes, but foreign body giant cells are also present in apposition to some of the fat cells."

Comment.—I reported a very similar case in 1947 but no cystic lesions were present. Resolution in that instance was almost complete in two months.

The special interest of this case is in the two fluctuant swellings over the mass. Such swellings are distinctly rare although Sir Archibald Gray's (1926) first two patients showed them. They may be mistaken for abscesses or cavernous hæmangiomata.

No notable contribution has been made since the work of Channon and Harrison (1926) on the chemistry of the fat. The conclusion was that there was a relative deficiency of olein so that the preponderance of stearin and palmitin gave a high melting point to the body fat and that, possibly, cooling of the skin led to the deposition of needle-like crystals which provoked a foreign-body granulomatous reaction. Although the investigations of Harrison led to the inference that the crystals were natural fat, neither the evidence nor the explanation was entirely satisfactory since in view of the fact that an infant's fat contains less olein than that of an adult, it is surprising that sclerema neonatorum occurs so rarely and is not much more common in cold climates.

The differential diagnosis involves the separation of this specific entity from œdema neonatorum, scleroderma, pre-agonic or cadaveric induration of the fat of the newly born and possibly from what is described as subcutaneous fat necrosis of the newly born. One would expect subcutaneous fat

necrosis to be localized and usually traumatic but, whether traumatic or idiopathic, saponification of the necrotic fat tissue would result in the early formation of free fatty acids and deposition of calcium salts. Harrison was unable to find free fatty acids in his investigations of material from sclerema neonatorum. Pre-agonic or cadaveric induration is so obviously an ante-mortem solidification of the fat in a marasmic, moribund infant that it could scarcely be confused with sclerema neonatorum, except in the rare case when the latter disease involves the trunk and limbs with, in consequence, a serious prognosis.

I am indebted to Dr. C. H. Whittle and to Dr. O. P. Bewers, the attending practitioner, for this interesting patient, and to Mr. D. Martin for the excellent photograph.

After showing a photograph of his case, Dr. Brain showed a second photograph and said that it illustrated the patient he saw with Dr. E. Davis in 1945. The clinical condition was very similar to that of the patient shown to the meeting but no cysts were present (*see* Davis and Brain, 1947).

POSTSCRIPT (June, 1953).—Progress: The condition had clinically resolved in two months. R. T. B.

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GRAY, A. M. H. (1926) *Arch. Derm. Syph., Chicago*, **14**, 635.
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HARRISON, G. A. (1926) *Arch. Dis. Childh.*, **1**, 63 and 123.

Urticaria Pigmentosa with Bone Lesions. Two Cases.—C. D. CALNAN, M.B.

I.—K. P., female, aged 24.

History.—Three years ago: Onset of eruption on legs and arms, and some lesions scattered on trunk. They become red and wheal with friction or heat.

Examination.—The arms, legs and chest show urticaria pigmentosa of the adult type.

Investigations.—Biopsy: Numerous mast cells in the dermis, especially around blood vessels. X-ray of skull: There is an annular clear-cut deficiency about 1 cm. diameter of both tables in the L. parieto-frontal region in the line of a suture. The margins are sclerosed (Fig. 1).

II.—E. F., female, aged 52.

History.—Five years: Rapid onset of the present eruption which itches on occasions especially when the skin is exposed to a fire or hot water. It is present on the arms, legs and more sparsely on the trunk.

Examination.—Lesions of urticaria pigmentosa of adult type on the forearms, arms and lower legs, and a few on the trunk.

Investigations.—X-ray skull: There is a small ill-defined defect in the right frontal bone about 5 mm. diameter (Fig. 2).

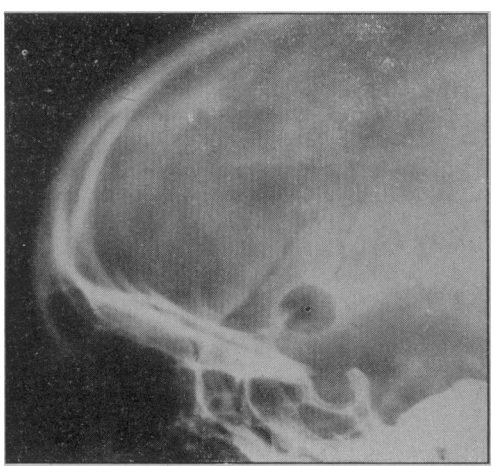

FIG. 1.

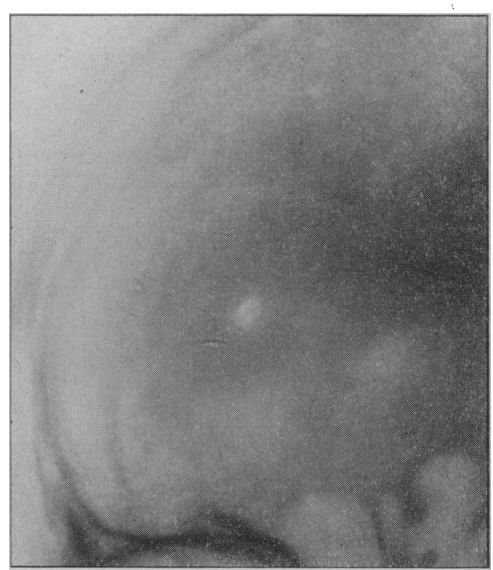

FIG. 2.

Comment.—These 2 cases are presented in order to discuss the significance of bone lesions in urticaria pigmentosa.

I became interested after reading of Felix Sagher's patient who had cystic osteoporosis of the ribs and vertebræ (Sagher *et al.*, 1952). (Clyman and Rein, 1952, also have examined about 6 patients.) There is no specific diagnostic feature about the X-rays of my patients, and the abnormalities could well be due to other incidental pathological processes. There is, however, some evidence for regarding this condition as a systemic disorder, and French authors not infrequently classify their cases as mast-cell reticulosis (Hissard *et al.*, 1951).

REFERENCES

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Congenital Ichthyosiform Erythrodermia (Brocq) with Bullæ.—R. G. HOWELL, M.R.C.P. (for H. J. WALLACE, F.R.C.P.).

R. W., male, aged 23.

This patient shows the characteristic lesions of congenital ichthyosiform erythrodermia, with large plaques of horny skin on the trunk and limbs. The face and extremities are less markedly involved. The lesions have remained unchanged since early childhood. On the trunk and legs he develops bullæ, 2·5–5 cm. in diameter. They are flaccid, subepidermal, and readily become secondarily infected. There is no seasonal incidence.

Treatment.—Liquid paraffin is the most suitable local application. Vitamin A, 150,000 units daily for nine months, made no change to the lesions. Sulphapyridine, 0·5 gramme t.d.s., appears to reduce the frequency of blister formation.

Comment.—The occurrence of bullæ in this disease has been attributed to sepsis, to sweat retention and to trauma. Sulphapyridine may have some effect if sepsis is causative.

Partial Congenital Ichthyosis.—R. G. HOWELL, M.R.C.P. (for G. B. DOWLING, M.D., F.R.C.P.).

J. K., male, aged 38.

This patient has had thickening and roughness of the skin of the arms and legs since infancy. The lesions are symmetrical and involve the dorsum of the hands and feet, the arms to above the elbows and the legs to the mid-thigh. There is a sharp line of demarcation between affected and normal skin and there are bilaterally symmetrical islands of normal skin in the hyperkeratotic areas. The palms and soles are not affected. The lesions are symptomless and do not vary.

Comment.—This case resembles that described by Vilvandré as "an unusual case of symmetrical keratodermia". In both cases the lesions are symmetrical, sharply demarcated from normal skin, and remain unchanged from early childhood. In Vilvandré's case the hyperkeratosis was more florid, and the palms and soles were involved.

REFERENCE

VILVANDRÉ, G. (1918) *Brit. J. Derm.*, **30**, 202.

Dr. H. J. Wallace: There is one detail I would like to add. The cause of the blistering in the patient is uncertain but we have another patient with a less severe form of this disorder in whom the blistering invariably appears to be traumatic.

Dr. H. R. Vickers: I would suggest that the second case is of the same order as the first. I do not like the diagnosis of ordinary ichthyosis; there is no involvement of the palms.

Dr. R. T. Brain: I agree. One can see the exaggerated pattern of hyperkeratosis in both cases. I think this may have started as a congenital ichthyosiform erythrodermia. The erythrodermia disappears and leaves the ichthyotic elements which become more conspicuous.

Dr. F. Ray Bettley: I think the second case would be best called widespread hyperkeratotic nævus because it does not really have the characteristics which are present in the first case which is a true congenital ichthyosis, with extremely thick "crocodile" skin. The second case does not progress to that degree.

Sarcoid Following Injury.—R. G. HOWELL, M.R.C.P. (for G. B. DOWLING, M.D., F.R.C.P.).

M. F., male, aged 32.

This man was blown up by a "booby-trap" in Palestine in 1945. His face and shoulders were peppered with soil particles. When seen in 1951 there were many small brown-red papules scattered among the black pigmented marks of his injuries. He could not say when the nodules first appeared, and they have remained unchanged since 1951. The spleen was not palpable and there were no enlarged lymph glands. The general health was good.

Investigations.—X-ray of chest and hands revealed no abnormality. Mantoux 1/1,000 negative.

Biopsy of a nodule from the shoulder (Dr. Ian Whimster).—"The dermis contains numerous discrete and confluent tubercles. Doubly refractile, colourless, crystalline foreign bodies are present in the tubercles, sometimes inside giant cells, but are also present in the dermis where they do not appear to be exciting any inflammatory reaction."

Treatment.—Foreign bodies have been removed from both eyes and a right corneal graft done. No treatment has been given for the skin lesions.

Comment.—Silica particles in the skin may excite a particular "tuberculoid" local inflammatory response, or if the patient develops "sarcoidosis" subsequent to the injury, the silica particles may preselect the site of the lesions.

In R. D. Sweet's case, sarcoid tissue was found in lymph glands quite independent of silica deposits. Silica particles in the skin are only rarely associated with "sarcoid" tissue. It appears then that they may preselect the sites of development of lesions in a generalized disease.

REFERENCE

SWEET, R. D. (1950) *Proc. R. Soc. Med.*, **43**, 173.

The following cases were also shown:

Scleroderma.—Dr. C. H. WHITTLE and Dr. J. MOFFATT.

Granuloma of Face: For Diagnosis.—Dr. G. A. BECK and Dr. C. H. WHITTLE.

Case for Diagnosis: ? Granulomatous Mycosis. ? Squamous Carcinoma.—Dr. BRIAN RUSSELL.

Epidermolysis Bullosa Dystrophica.—Dr. P. J. FEENY.

Hypertrophic Lichen Planus with Epitheliomata.—Dr. HAROLD WILSON.

(These cases may be published later in the *British Journal of Dermatology.*)

[*February* 19, 1953]

Mycosis Fungoides.—I. B. SNEDDON, M.B., Ch.B., M.R.C.P.

A. W., male, aged 66. Monumental Mason.

History.—In October 1952 he developed an itching red eruption on the head, face and neck and his face became swollen. Apart from this he felt well; he had taken no drugs. The rash had spread slowly down his trunk and the proximal parts of the limbs.

Past history and family history.—Not relevant.

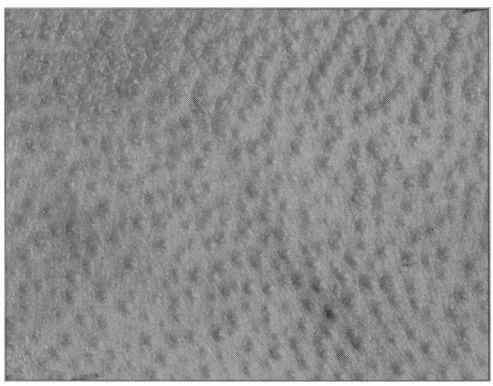

FIG. 1.—The skin of the upper trunk showing the follicular papular eruption.

On examination.—A confluent bright erythema was present over the scalp and face; the skin of the face was indurated and deep fissures were present in the natural skin folds around the mouth. The neck, upper trunk and arms showed a discrete red follicular papular eruption which was becoming confluent around the neck (Fig. 1). The hands and feet were unaffected. No enlargement of lymph glands, liver or spleen could be found.

Investigations.—Skiagram of chest showed no significant abnormality. W.R. negative. Blood count: Hæmoglobin 128%; red cells 6,400,000; leucocytes 9,000; differential count normal; marrow puncture within normal limits.

Histological examination of the follicular rash on the trunk (Dr. L. C. D. Hermitte): The epidermis shows a slight degree of acanthosis. Its surface is wavy but there is no hyperkeratosis or parakeratosis. The mouths of the follicles are, however, filled with lamellated keratin. There is a dense perifollicular infiltrate with lymphocytes and histiocytes and the centre of the follicle appears to contain keratin. The sebaceous glands show some degree of atrophy. Some of the capillaries in the corium are dilated and surrounded by a few chronic inflammatory cells. The appearances suggest a mild degree of pityriasis rubra pilaris.