however, of a granulomatous nature with numerous giant-cells—a picture that has been associated with “malignant granuloma” and “Wegener’s granulomatosis.”

Godman and Churg (1954) are of the opinion that “Wegener’s granulomatosis” and “granuloma gangraenescens” (malignant granuloma) are closely related conditions. These authors were able to study one case of the latter disease of slow progressive course in which at necropsy recent arteritic lesions were found. They also note with interest that in the case of “Wegener’s granulomatosis” of longest duration—48 months—only minimal vascular lesions were present *post mortem*. One of their patients is receiving maintenance doses of cortisone and antibiotics nearly three years since he presented with destructive sinusitis and pulmonary granulomata and nephritis.

Summary

The case history of a woman of 41 years who presented with granulomatous rhinitis and gingivitis and died in uraemia after a seven-weeks illness is reported. Necropsy and histological examinations revealed necrotizing glomerulonephritis and lesions of “periarteritis” in many organs, but absent from the lungs.

The relationship between “periarteritis nodosa” and periarteritis with granulomatous lesions is briefly discussed. The general occurrence of pulmonary lesions in the latter syndrome is noted.

Attention is drawn to a recent survey of “Wegener’s granulomatosis” and the possible relationship of this condition to “malignant granuloma.”

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REFERENCES

- Godman, G. C., and Churg, J. (1954). *Arch. Path.*, **58**, 533.
 Howells, G. H., and Friedmann, I. (1950). *J. clin. Path.*, **3**, 220.
 Klinger, H. (1931). *Frankfurt. Z. Path.*, **42**, 455.
 Kussmaul, A., and Maier, R. (1866). *Dtsch. Arch. klin. Med.*, **1**, 484.
 Lindsay, S., Aggeler, P. M., and Lucia, S. P. (1944). *Amer. J. Path.*, **20**, 1057.
 Mönckeberg, J. C. (1905). *Beitr. path. Anat.*, **38**, 101.
 Ophüls, W. (1923). *Arch. intern. Med.*, **32**, 870.
 Stratton, H. J. M., Price, T. M. L., and Skelton, M. O. (1953). *British Medical Journal*, **1**, 127.
 Veszprémi, D., and Jancsó, M. (1963). *Beitr. path. Anat.*, **34**, 1.
 Wegener, F. (1936). *Verh. dtsch. path. Ges.*, **29**, 202.
 — (1939). *Beitr. path. Anat.*, **102**, 36.
 Zeek, P. M., Smith, C. M., and Weeter, J. C. (1948). *Amer. J. Path.*, **24**, 889.
 — (1952). *Amer. J. clin. Path.*, **22**, 777.

The fortieth annual report of St. Dunstan’s, *Forty Years On*, gives an account of the work of St. Dunstan’s for the war-blinded since its inception. Some 5,000 Servicemen and a few Home Guards and Civil Defence workers have come to St. Dunstan’s for training and rehabilitation. To-day a total of 2,600 St. Dunstaners are living—1,450 from the first world war and 1,150 from the second. 58 new cases, including 27 from the second world war, recent casualties from Malaya, Korea, and East Africa, and men from 1914–1918 who have gradually gone blind from mustard gas poisoning, were received last year. Experience has proved that those who come to St. Dunstan’s in the early days of their blindness readjust themselves quickest to their new condition, and that neither home nor hospital is the best place for such adjustment. As many blind people have administrative and executive posts and help in the teaching and welfare departments at St. Dunstan’s, newcomers are taught by example how to reconcile themselves to their deprivation. To-day, 120 war-blinded are chartered physiotherapists, 140 telephone operators, 100 and more shopkeepers, 550 industrial workers, and some are doctors, solicitors, company directors, and welfare officers.

MALIGNANT CHANGE IN ERYTHEMA AB IGNE

BY

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[WITH SPECIAL PLATE]

The reddish-brown marbling on the skin of the lower legs often found in women who sit long in front of a fire is so common in Britain that it is taken as a matter of course and little attention is paid to it in our textbooks of dermatology. In these it is dismissed in a few lines as erythema ab igne or ephelis ab igne, though the word *epheles* (freckle) is wrongly used. Hartzell (1912) pointed out that the disease is essentially an inflammatory change and not simply a discoloration of the skin with blood pigment.

Possibly because of the ubiquity of central heating, the dermatosis appears to be rare in the United States, for Hartzell (1912) remarked that cases he has shown at meetings had not been diagnosed by competent dermatologists. He stated that “English dermatologists are apparently the only ones who see it with any degree of frequency.” In the same paper he described four cases of this disease and observed that none of the patients were in good health—a significant point. It seems likely that the condition is still often unrecognized in the U.S.A., for Hyman (1954) in discussing a case shown at the New York Academy of Medicine as purpura annularis telangiectodes (Majocchi’s disease) remarked that “the lesions are neither annular nor telangiectatic. So the two important features of Majocchi’s disease are lacking. In England one often sees this peculiar kind of mottling of the skin, with nothing telangiectatic about it at all, known as erythema ab igne and due to sitting in front of the fire.”

In the standard French textbooks, such as the *Précis de Dermatologie*, no reference to the disease can be found, though Erasmus Wilson (1863) quoted Rayer, who described “epheles ignealis in women sitting over charcoal burners, the histological appearances being those of congestion followed by chronic inflammatory thickening, and even vesication and considerable pigmentation is produced.”

In Algiers, Montpellier (1921) described this same reticulate condition as occurring on the inner aspect of the thighs in Moslem women from the use of a little charcoal heater held between the thighs in winter.

But erythema ab igne is probably most common in the colder parts of Asia; for instance, in Kashmir, in cold weather the inhabitants carry beneath their clothing a warming-pan or *kangri*, the temperature of which may reach 150° F. (65.5° C.). Neve (1910) described *kangri* cancer which followed heat burns similar to erythema ab igne. Within a period of fifty years Neve (1929) saw more than 2,000 cases of such cancers. Similar to these is the reaction seen in some parts of China where the people sleep on a heated brick bed or *kang*, so that the skin over the trochanter is burned and *kang* cancer, as described by Laycock (1948), may be provoked. An editorial in the *British Medical Journal* (1954) on “Skin Cancer and Burns” refers to these cancers, and to others such as the well-known carcinoma of the lips in clay-pipe smokers.

It is therefore not altogether unexpected that the erythema ab igne seen in this country should occasionally be the site of squamous-cell carcinoma, and since 1951 I have had four patients with erythema ab igne in which a squamous-cell carcinoma has developed. Because of the frequent occurrence of this unsightly marking of the legs in elderly, middle-aged, and even youthful females in Britain, it was confidently expected that such malignant changes would have been previously described in some book or article, but searching has failed to reveal any such picture.

Description of Cases

Case 1.—A woman aged 71 was first seen in May, 1951, with a particularly severe erythema ab igne, which had been present for years, and a hypertrophic ulcerated area of the left leg, present since September, 1949. A biopsy from the tumour was reported on as follows: "The section consists of the edge of an ulcer bordered by acanthotic epidermis. The floor of the ulcer shows a dense chronic inflammatory infiltrate that almost obscures a low-grade squamous-cell carcinoma. The acanthotic epidermis shows no increase in pigment, the upper dermis is oedematous, the vessels widely dilated, and there is a mild chronic inflammatory reaction round some of the vessels." The Wassermann reaction was negative. The patient was a thin, tired woman with a noticeable reticular pigmentation and telangiectasis on the sides and V of the neck, and erythematous-squamous patches on the brow, nose, cheeks, and tips of the fingers not unlike lupus erythematosus, except that there was no atrophy. A diagnosis of poikiloderma atrophicum vasculare (Jacobi) with erythema ab igne and squamous-cell carcinoma was made. This case was among those shown at a meeting of the North British Dermatological Society on December 13, 1951. Early in January, 1952, the carcinoma was excised and the raw area grafted.

Case 2.—A spinster aged 80 developed an adenocarcinoma of the left breast which was removed surgically in 1942: this was followed by deep x-ray therapy, and there has been no sign of recurrence. Apart from this she has been healthy. For many years she had been in the habit of huddling over a big coal fire in the winter, but when she noticed marked "fire-burns" she placed a board and a rug between her legs and the fire. Gradually the pigmentation and redness faded, leaving warty excrescences. In July, 1952, she was referred to me by Dr. Anna Herdman for treatment of a tumour which had grown on one of the warty lesions on her right shin and had reached a diameter of 2 cm. with early ulceration. A biopsy showed a moderately differentiated squamous-cell carcinoma, which was removed by thorough curettage followed by electrocautery. When this area had healed it was treated with x rays (2,000 r). She remained under observation. In February, 1955, the irradiated area was still healthy, but another squamous-cell carcinoma had developed on the other leg, and this was treated in the same way.

Case 3.—A woman aged 57 was admitted to the Skin Department of the Royal Infirmary, Edinburgh, on September 7, 1953, with multiple hypertrophic ulcerating tumours of the inner aspects of the thighs and lower legs, situated on areas of skin affected by a chronic erythema ab igne. She confessed that for years she was accustomed to sitting close to a coal fire and realized that this was responsible for her condition. The tumours had gradually developed over the past three years. Her skin was pale, very fair and fine, with almost complete absence of axillary and pubic hair, so as to suggest an ectodermal defect. On the sides of her neck was a reticulate pigmentation with telangiectasia but no true atrophy, and suggested poikiloderma atrophicum vasculare of the Thomson type rather than of the Jacobi variety. Histological examination of tissue removed from several of the tumours showed squamous-cell carcinoma. The tumours were treated by deep x-ray therapy, and all vanished satisfactorily except

one on the right and one on the left leg, each measuring 3.5 cm. in diameter. The patient was therefore transferred to the care of Mr. A. B. Wallace, plastic surgeon, who excised the carcinomata and grafted the raw areas. (See Figs. A and B.)

FIG. A.—Case 3: Showing erythema ab igne with hyperkeratotic areas and squamous-cell carcinoma.

FIG. B.—Case 3: Close-up of squamous-cell carcinoma.

Case 4.—A woman of 56 was sent to see me by Dr. A. E. Turnbull on June 5, 1954. For many years she had noticed discoloration of the skin, chiefly of the left leg, and realized that this was due to her "sitting on top of" a coal fire. As she sat to the right of it the left leg was more scorched than the right. About three years ago she observed a warty area on the marbling of her left leg, and this gradually increased in size. Her previous medical history is of no interest except that in 1946 a malignant ovarian cyst was removed, and following this she received deep x-ray therapy. Since then there has been no evidence of recurrence, but her left leg has been slightly oedematous from time to time.

FIG. C.—Case 4: Erythema ab igne with squamous-cell carcinoma of left calf. FIG. D.—Case 4: Infra-red photograph showing retiform pigmentation over deep veins.

The clinical appearance of the condition is shown in Fig. C, and an infra-red photograph for comparison in Fig. D. A biopsy was interpreted by Dr. R. F. Ogilvie, senior lecturer in pathology, Edinburgh University, as follows: "The cutis is very oedematous, markedly infiltrated with polymorphs, round cells, and siderophages in varying proportion, and characterized by a formation of highly vascular granulation tissue. The overlying epidermis is slightly raised and covered by a distinct cornified layer, while masses of keratin plug the epidermal crypts. Bacterial masses are present among the cornified material, and the epidermis as a whole is slightly infiltrated with polymorphs. The Malpighian layer is sprinkled with mitotic figures and includes numerous feebly stained cells, but is not significantly thickened. In other words, no evidence of malignancy is present in the section. The features are those of an organizing subacute inflammatory condition accompanied by some hyperkeratosis." (See Special Plate, Fig. 1.)

The patient was admitted to hospital on July 15, 1954, and a small hyperkeratotic nodule excised from her right leg showed the following picture: "The epidermis is normal in thickness at one end of the section, but becomes progressively thicker in the nodular region. Both Malpighian and cornified layers share in the thickening. The rete pegs in particular are distinctly elongated and characterized by a swollen and actively dividing condition of their cells. Some parakeratosis is also apparent at the nodular end. The corium shows oedema, infiltration with round cells, polymorphs, and siderophages, mainly the first, and formation of vascular granulation tissue, but these features are more marked at the nodular end than elsewhere. The thickened cornified layer at the nodular end is also locally infiltrated with polymorphs. No malignancy, however, is present in this section. The condition is erythema ab igne of particularly marked degree at the nodular end and here accompanied by conspicuous reactive epidermal thickening." (Plate, Fig. 2.) Another biopsy was taken from the tumour on the left leg, of which Dr. Ogilvie reported: "The section shows: (i) the features of well-marked erythema ab igne with reactive epidermal thickening as described for the nodular area in the previous section; and (ii) invasion of the tissue at one end of the section by a moderately differentiated squamous carcinoma." (Plate, Figs. 3 and 4.)

The carcinoma was excised at the plastic unit of Mr. A. B. Wallace and thick razor grafts applied.

Another case of erythema ab igne exemplifies the same reaction (precancerous changes) as that seen in the section taken from the small hyperkeratotic nodule of the right leg in Case 4.

Case 5.—A woman of 77 was referred to me by Dr. A. Y. Cochrane on November 6, 1953. For many years she had had an erythema ab igne affecting the left leg only, and 12 years ago relinquished her habit of sitting over a coal fire. In August, 1953, she observed that an exudative crusting was developing on the "fired" area, and this soon became a heaped-up ulcerated growth. The presence of a squamous-cell carcinoma was suspected, and the tumour was removed by curettage and the base thoroughly destroyed by electrocautery. Section of the lesion showed no evidence of carcinoma, the histological report being: "A keratosis. Much of the epidermis is acanthotic, but the normal cellular architecture is retained. There is a diffuse chronic inflammatory reaction in the dermis. The epidermis is obviously unstable, but no definite malignancy was seen." The skin healed satisfactorily in a little over four weeks, and there has been no recurrence nor have fresh tumours arisen. Apart from a fissure-in-ano her general health has always been good.

Discussion

In taking the history of a patient with squamous-cell carcinoma one is frequently informed that the growth appeared subsequent to a minor injury to the affected part—a point which sometimes has to be considered in industrial compensation. More often, however, these carcinomata tend to appear after repeated minor insults to the skin, so

that the appearance of an epithelioma on the site of chronic irritation is a well-known phenomenon, as witness the high incidence of skin cancers in sunny countries such as Australia and South Africa, or the fungating tumours that used to be only too frequent in chronic lupus vulgaris.

The heat-burns due to the *kangri* pot and the *kang* bed have also produced their quota of carcinomata in Asia, and the precarcinomatous lesions in these cases are indistinguishable from erythema ab igne, which has also been reported as due to prolonged application of a hot-water bottle to the abdomen or back. Surprising features of the cases here described are: (1) there appears to have been no previous description of similar cases in any medical book or journal, despite the high incidence of erythema ab igne in this country; and (2) in this small number two of the four patients with carcinoma had been cured of cancer of other organs some years previously, while the other two had skin changes compatible with a diagnosis of poikiloderma atrophicans vasculare, a disease which may be the first sign of an internal carcinoma, leukaemia, or mycosis fungoides, but may also be found in subjects with hereditary ectodermal defects, such as existed in Case 3. Exposure to sunlight and to cold seems to have played a part in some cases of this type of poikiloderma, and Downing *et al.* (1947) have suggested that physical factors might well be precipitating factors.

In the cases now presented the disease began as an acute erythema, sometimes with vesiculation, following exposure to the heat of a coal fire. This erythema died down and recurred repeatedly after numerous similar experiences. Gradually a retiform pigmentation appeared on the skin over the deep veins, as shown by infra-red photography (see Fig. D). After years of such incidents, hyperkeratoses appeared on the dusky-red pigmented areas, as can be seen around the tumour in Figs. A and B. These successive processes ranging from a chronic inflammatory reaction to squamous-cell carcinoma are shown in the histological reports on Case 4 and the photomicrographs (Plate, Figs. 1-4).

It may be impossible clinically, and even histologically, to distinguish when a tumour such as those described has ceased to be a pseudo-epitheliomatous hyperplasia and has become a true squamous-cell carcinoma. In Case 5 the growth appeared to be a hyperplasia only, as serial sections showed no real invasion and comparatively few mitoses, but no doubt it was a precancerous lesion. The carcinomata which developed in these cases were of low malignancy with no evidence of secondary spread, so that the prognosis appeared to be good.

Though erythema ab igne is extremely common in women in Britain, it does not seem to be of frequent occurrence in other parts of Europe, and is decidedly rare in America. This may be accounted for by the universality of central heating, though there are still plenty of coal fires in use in the United States. Any form of heat, if the exposure is long enough, would probably produce the disease, and there is some evidence that erythema ab igne has been provoked by gas and electric fires. It would almost seem as if the inhabitants of North America were not so prone to develop this type of reaction, for Danto and Maddin (1954) described the case of a woman in Vancouver who had developed what they termed "thermal telangiectasia" caused by an electric fire, an electric heating pad, and a car heater. This patient developed numerous discrete, pinhead-sized telangiectases on the posterior aspect of her legs, with no sign of hyperpigmentation, freckling, or mottling. One cannot help thinking that in Britain this person would have shown a typical erythema ab igne.

During a long experience with out-patients, I have observed many women with erythema ab igne, some in the early twenties, but mostly over the age of 40. I have not yet seen a single case of the disease in a male, even though many have confessed to the habit of sitting huddled over a fire for long spells. It may be that endocrine factors play some unknown part in the production of the lesions.

Summary

The chronic inflammatory changes of the skin known as erythema ab igne, due to prolonged sitting over a fire, are common in this country but rare in the United States.

Similar cutaneous changes due to the heat from the *kangri* bowl in Kashmir and the *kang* bed in China have been shown to produce cancers.

Four cases are described in which the patient has developed a squamous-cell carcinoma on an area of erythema ab igne, and one case in which there were premalignant changes. Two of the four patients had been treated for, and cured of, cancer of other organs some years previously; the other two showed the cutaneous changes known as poikiloderma atrophicum vasculare, sometimes found in association with malignant disease.

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REFERENCES

- British Medical Journal*, 1954, 1, 204.
 Danto, J. L., and Maddin, W. S. (1954). *Arch. Derm. Syph. (Chicago)*, 70, 667.
 Downing, J. G., Edelstein, J. M., and Fitzpatrick, T. B. (1947). *Ibid.*, 56, 740.
 Hartzell, M. B. (1912). *J. cutan. Dis.*, 30, 461.
 Hyman, A. B. (1954). *Arch. Derm. Syph. (Chicago)*, 70, 839.
 Laycock, H. T. (1948). *British Medical Journal*, 1, 982.
 Montpellier, J. (1921). Quoted by C. du Bois, *Ann. Derm. Syph. (Paris)*, 2, 83.
 Neve, E. F. (1910). *British Medical Journal*, 2, 589.
 — (1929). *Practitioner*, 122, 355.
 Wilson, W. J. E. (1863). *Diseases of the Skin*, p. 599, Churchill, London.

PNEUMATOSIS CYSTOIDES INTESTINALIS

REPORT OF A CASE

BY

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[WITH SPECIAL PLATE]

Cases of gastro-intestinal pneumatosis have been reported in the literature from time to time. The condition was known to John Hunter, who described its occurrence in swine. There is a specimen in the Hunterian Collection of the Royal College of Surgeons: this was sent to John Hunter by Jenner and the gas from the cysts was analysed by Cavendish; a description of the specimen is to be found in Hunter's *Works* (Nitch, 1923). The first reference to the condition in man is in the catalogue of the Anatomical Museum of Amsterdam in 1737 (quoted by Lamont, 1929), where it describes "a portion of jejunum of a man showing a tumour which arose from wind." Bang described the condition in 1876 and Mayer in 1825, the latter calling it emphysema bullosa. Since then there have been many descriptions and much discussion in the literature as to the possible aetiology of these gaseous cysts in the abdomen. Jackson (1940) collected 171 cases from the literature and described one personal case. There have been over 17 further cases reported (Somerville, 1951).

The case now described is of interest because of the confusion caused by air cysts around the terminal ileum, which had become displaced and lay in a suprahepatic position under the right diaphragm.

Aetiology

The association of pneumatosis cystoides with pyloric stenosis is well known. Berglund (1939) states that in 60% of the cases described in the literature pyloric stenosis has been present. Lamont (1929) claims that 50% of cases are definitely associated with duodenal or gastric ulcer.

The aetiology was first fully reviewed by Nitch in his presidential address to the Section of Surgery of the Royal Society of Medicine (1923). He collected 85 cases from the literature, analysing the aetiological factors, and described two cases of his own.

Many theories have been put forward. Neoplastic, bacterial, and mechanical factors have been considered. Eveleth and Biester (1938) discovered that in hogs fed on polished rice cysts developed on the serosa of the intestine. This led many observers to suspect malnutrition as being the main causative factor in this disease. Schorr *et al.* (1951) drew attention to the fact that patients with this condition were usually undernourished, owing mainly to the long history of associated peptic ulceration and subsequent pyloric stenosis. If the pyloric stenosis is relieved the cysts tend to regress immediately, and have been observed to do so by Berglund (1939) both radiographically and by laparotomy. The disease affects infants, and Moore (1929) reported four cases of pneumatosis in infants who died of enteritis with ulceration. Botsford and Krakower (1938) also reported six cases occurring in children in association with diarrhoea or vomiting; no mucosal defects were found on necropsy in these cases. Morison (1952), in discussing the pathology of this condition in infancy, postulated that some of the gas might have been produced by micro-organisms invading the tissues before or after death.

Various authors have suggested that gas is driven into the lacteals through mucosal ulcers, and that the raised intra-abdominal pressure from repeated vomiting might lead to gaseous distension of lacteals.

Case Report

A man aged 51 was brought to St. Bartholomew's Hospital after having collapsed on the platform of a railway station. He complained of severe upper abdominal pain, worse on the right side. He was shocked, dehydrated, and vomiting blood. He gave a 20-year history of dyspepsia, having had a course of medical treatment in hospital 19 years ago for peptic ulcer. Four years ago he was operated on at another hospital for perforated gastric ulcer; the records show that no other abnormality was found in the abdominal cavity. Since this time he had developed two distinct types of pain, one coming on 1½ hours after meals and situated in the epigastrium, and the other in the right hypochondrium which radiated to the back and right shoulder and was not associated with meals. There was a weight loss of 4 stones (25.4 kg.) and he had been unemployed for four years (since his operation for perforation) on account of his disability. Recently repeated vomiting had developed and he had noticed melaena stools on several occasions. Since his operation he had suffered from very severe bronchitis, which had become worse in recent months.

Three days before admission he experienced sudden epigastric pain which persisted; 48 hours later the pain became worse and colicky. He vomited altered blood on four

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FIG. 1.—Case 4: Photomicrograph (×68) from nodular lesion on left leg.

FIG. 2.—Case 4: Photomicrograph (×68) from small nodule on right leg.

FIG. 3.—Case 4: Photomicrograph (×15) of section from large nodule on left leg, showing squamous-cell carcinoma.

FIG. 4.—Case 4: Photomicrograph (×68) of squamous-cell carcinoma.

J. D. GRIFFITHS AND D. F. ELLISON NASH: PNEUMATOSIS CYSTOIDES INTESTINALIS

FIG. 1.—Antero-posterior radiograph of abdomen. (a) Loculated gas beneath right dome of diaphragm. (b) Complete obscuring of central abdominal area by stomach grossly distended with blood clot.

FIG. 2.—Photograph of specimen of resected small intestine showing on left relatively collapsed segment with submucosal cysts, and on right (top) the opened proximal portion containing large intramural cysts. The intervening mass consisted of strictured bowel and contracted mesentery packed with large and small cysts.