Dermatomyositis was originally and independently described and named in 1887, by three persons: Hepp—pseudotrichinosis; Wagner—acute polymyositis; and Unverricht—acute progressive polymyositis. Unverricht later attached the name of Dermatomyositis to the condition, and though in the absence of cutaneous symptoms the term polymyositis is proper (Wedgewood, Cook and Cohen, 1935), the full term of dermatomyositis is preferable as serving to distinguish this variety of polymyositis from all others. The acute form is still known as the Wagner-Unverricht type (Degos).

First reports of the co-existence of dermatomyositis and visceral cancer came from Stertz (1916), Gottron (1931), and Bezecny (1935). That this association was no mere chance was shown in the collected series of patients that followed: Shuerman (1951)—12.9% of patients with dermatomyositis showing malignancy; Curtis, Blaylock, and Harrel (1952)—17.7%; Christianson, Brunsting and Perry (1956)—6.7%; Williams (1959)—15.3%; Arundell, Wilkinson and Haseler (1960)—34.3% (52.2% if aged over 40). Curtis, Heckman, and Wheeler (1961) concluded that with the elimination of those cases occurring in the younger age group the incidence in adults becomes approximately 50%; and that dermatomyositis, when acquired by an adult, should always be considered as occurring with a malignancy until proved otherwise.

In the original report by Bezecny (1935) on two patients, one with carcinoma of the ovaries and the other of the breast, it was noted that there was a remarkable improvement in the dermatomyositis of that patient who had an extirpation of the carcinoma of the ovaries. Improvement following treatment of underlying malignancy, whether by surgery or radiotherapy, has since been reported frequently. By contrast, this is believed to be the first case report of radiotherapy of cancer leading to dermatomyositis.

Case Report

A.W., a 45-year-old widow, presented in Outpatients on 21.1.63 complaining of a lump in her right breast which had been present about six weeks.

On examination there was a small nodule in the right upper quadrant of her right breast, which was not attached to skin or underlying tissue. There was no evidence of lymph node involvement or secondary deposits.

Operation. On 28.1.63 a right radical mastectomy and bilateral oophorectomy was performed. Histology showed a scirrhous carcinoma and deposits within lymphatics. Her post-operative recovery was uneventful, and from 12.2.63 to 6.3.63 she received radiotherapy (Dr. W. G. Evans): DXR 250 KV to chest wall, T.D. 3,500r; and telecobalt to lymph node areas, T.D. 4,900r. On 27.3.63 she was noted to have a small patch of erythema and scaling over the right scapula and right anterior chest wall.

Progress. Three weeks later the rash had spread to her back, upper chest, arms, neck and face, and she had developed swelling and oedema of these areas, with complete closure of her left eye. She also complained of pruritus and weakness of her upper limbs; and later of some difficulty in swallowing.

On examination at this time she presented the typical appearance of a patient with dermatomyositis. The skin over the face, chest, and arms and back was poppy-red with a slight violaceous tinge, and felt hot to the touch. In addition she had oedema, brawny in places, at these sites. The skin over her knees and elbows was red and tender, and over her knuckles and extensor tendons showed faint red patches and streaks. Many dilated capillary loops were visible in the nail folds, which were sore, and splinter haemorrhages showed beneath the nail plates. There was marked muscle weakness of the sternomastoid and shoulder girdle muscles, and slight weakness of the pelvic girdle muscles. There were no neurological abnormalities; all reflexes were present and equal. The operation scar was healthy and some telangiectases were present over the sites of X-ray therapy; there was no evidence of any secondary deposits.

Investigations done at that time were as follows:
Hb. 11.8g. WBC 4,000/mm. Film normal. ESR 35 mm./hr. SGOT 70 units, SGPT 50 units. Blood urea: 35 mg./100 ml. Serum Protein: 6.4g./100 ml. Electrophoresis: a decrease in albumin and increases in alpha-2 and gamma globulin. C-Reactive protein: positive. Examination for LE Cells: negative. Urinary creatinine: 0.65g./24 hours; creatine 0.5g./24 hours. X-ray chest, pelvis, and cervical, thoracic and lumbar spine showed no evidence of secondary deposits. Barium swallow: normal.

Treatment. She was treated with 80 mg. prednisolone daily and testosterone propionate 200 mg. twice weekly, replaced later by Nilevar tabs. 1 daily. On this regime her skin lesions regressed and her muscle strength improved, so that by May 1963 she was able to return home on a maintenance dose of prednisolone 30 mg. daily, which was reduced to 15 mg. daily.

In January 1964 there was a recurrence of oedema and erythema of face, chest and arms, which required an increased dose of steroids for a short while.
Otherwise her progress to date has been uneventful. Present state 3.9.63 shows a remarkably fit woman, who has remarried and leads a reasonably normal life. She is at present taking prednisolone 20 mg. daily, calciferol and Nilevar. The only changes now present are those from prolonged steroid therapy. Apart from a mild weakness, there are no signs of, or changes left over from, her dermatomyositis.

Comment

Dermatomyositis is an inflammatory and degenerative parenchymatous myositis. The skin changes when present are non-specific and may be followed by atrophy. As such it falls into the broad group of connective tissue disorders or mesenchymoses. Though classified with the connective tissue disorders, it must be kept separate from systemic lupus erythematosus, systemic sclerosis, and the vascular collagenoses, which, though they may mimic it closely, show a general systemic involvement absent in dermatomyositis, and in which any association with malignancy is no more than fortuitous. It must also be kept distinct from the carcinomatous neuromyopathies. Henson, Russell and Wilkinson (1954) discussing carcinomatous neuropathy and myopathy stated that the constant absence of skin lesions and supporting pathological evidence excludes dermatomyositis. Brain (1963), in his Thom lecture on the neurological complications of neoplasms, remarked that, while dermatomyositis might occur in association with a carcinoma, it was doubtful whether the muscular changes in the myopathic-myasthenic group should be regarded in the same disorder.

Dermatomyositis occurs in two forms—Adult, in which association with malignancy may be as high as 50% (Curtis, Heckman and Wheeler, 1961; Arundell, Wilkinson and Harerick, 1960), and Juvenile, in which association with malignancy must be exceptional, but cannot be excluded. Sunde (1949) reported a girl aged 11 with dermatomyositis who subsequently developed a chromophobe adenoma. This state of affairs is analogous to that found in the malignant and juvenile varieties of acanthosis nigricans. Separation into types was foreseen by Williams (1959) who questioned whether the symptom complex which presented itself as dermatomyositis was truly one disease, particularly in view of associated malignancy in cases reported.

Dermatomyositis is popularly thought of as being a disease of auto-immunity, which would account for its rare haphazard occurrence in patients suffering from a wide variety of cancer and bring it into line with the other diseases of supposed auto-immunity. Curtis (1952) suggested that tumour catabolic products may serve as allergen initiating the disease, and with Heckman and Wheeler (1961) demonstrated skin-sensitising antibody to tumour extract in a patient with dermatomyositis and metastatic carcinoma of the lung from the sigmoid colon. Similar results had previously been obtained by Grace and Dao (1959) in a patient with carcinoma of breast, and they too postulated that sensitivity played a role in the genesis of dermatomyositis.

But dermatomyositis could also represent a failure of immunity.

Burnet (1961) considered that the first step in evolution of the immune process lay in self-recognition and the ability to delete somatic mutant (malignant) cells; and he (Burnet, 1964) has put forward the view that the immunological inadequacy of early childhood and advancing age may have some bearing on the high incidence of cancer at these times. Some allergic response to tumours should, therefore, be the rule rather than the exception, and the immunological inadequacy that may be the forerunner of cancer may at times also result in the failure to produce neutralising antibody against foreign toxic tumour products (carcinotoxins) which may then injure nervous tissue (neurotoxins), muscle tissue (myotoxins) and so on.

The sequence of dermatomyositis following radiotherapy points to products of tumour destruction as the primary cause, but does not help decide whether they act as allergens or carcinotoxins.

Summary

A case, believed to be the first, of dermatomyositis following treatment of a carcinoma with radiotherapy is reported. Dermatomyositis, an entity distinct from other connective tissue disorders and the carcinomatous neuromyopathies, occurs in two forms: Adult, often associated with malignancy; and Juvenile, not associated. The aetiology may lie in some form of auto-immunity, but in adult cases with malignancy, the converse, a failure of immunity, could be the cause. No conclusion can be drawn from the case reported.

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Gottron (1931): quoted by Degos.


Dermatomyositis following Treatment of Cancer

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