Pemphigoid and Carcinoma of the Pancreas

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The association of internal malignant neoplasms with diseases of the skin other than secondary deposits has been recognized for over a century. There are, however, only a handful of cases in the literature where pemphigoid is recorded as the skin lesion accompanying a malignant neoplasm, and it is undecided whether such a combination is fortuitous or not. Such an association is difficult to analyse statistically, because there are so few cases, and it is therefore helpful to record examples when they occur.

CASE HISTORY

A woman aged 76 was blind owing to glaucoma. There had been recurrent urinary infections for 20 years. She was first seen in April 1963 with two months' history of loss of weight, anorexia, regurgitation, vomiting, and lower abdominal pain. On examination she had a furred tongue and belched frequently. B.P. was 160/100. In the abdomen there was a vague mass in the left hypochondrium; the only other finding was of mild Parkinsonism.

Investigations showed a haemoglobin of 15.2 g.%, E.S.R. 29 mm./hour, and a white-cell count of 14,000/c.mm., 84% polymorphs. The leucocytosis was explained by an *Escherichia coli* urinary infection, which was treated first with nitrofurantoin ("furadantin") and later with sulphamethoxypyridazine. Barium meal and follow-through showed scattered diverticula in the duodenum, jejunum, and colon. Tests for occult blood were negative.

The patient continued to eat little, had several small vomits daily, and the lower abdominal pain persisted. Three weeks after admission she suddenly collapsed with raised jugular venous pressure, tachycardia, and hypotension. An electrocardiogram showed right branch block and T-wave inversion in V6. A diagnosis of massive pulmonary embolism was made and treatment was started with metaraminol, heparin, and phenindione. She slowly recovered and the right bundle branch block disappeared. The phenindione was continued. The only new sign was oedema of both legs, which was treated with mersalyl.

Four weeks after the pulmonary embolism a bullous eruption developed; it involved the four limbs, the back, and the buccal mucosa. There were petechiae on the lower legs, and the oedematous feet became purple. Nikolsky's sign was negative. A biopsy specimen of the skin showed a subepidermal blister containing fibrin and eosinophil polymorphs, which were also numerous around the small blood-vessels of the upper cutis. The appearances were consistent with the diagnosis of dermatitis herpetiformis or pemphigoid and were distinct from those of true pemphigus and erythema multiforme.

The mersalyl was stopped at the first sign of the rash, and the phenindione tailed off over a period of four days. Prednisone, 30 mg. daily, did not control the eruption, and the dose was increased to 45 mg. daily. Over the next week no new bullae developed, and there was considerable clearing of the existing lesions.

At this point the patient again collapsed, and the right bundle branch block reappeared, but this time there was no response to treatment and death followed half an hour later.

At necropsy a carcinoma of the pancreas was found involving body and tail, without necrosis. There were deposits in the peri-pancreatic lymph glands, liver, and right lung. Thrombotic emboli were found in both pulmonary arteries, and ante-mortem thrombus filled the right internal iliac vein. The heart weighed 320 g., with slight hypertrophy of the left ventricle, and showed brown discolora-

tion. Both kidneys were slightly shrunken, with a few cortical scars; the left one contained a cyst 3 cm. in diameter, and some minute cysts in the medullary pyramids. There was a submucous fibroid in the uterus.

Examined histologically, the pancreatic tumour was a mucus-secreting adenocarcinoma. Additional findings were focal myocarditis, with eosinophils and histiocytes, and, in the kidneys, linear scars associated with calculous material in the collecting tubules.

COMMENT

Forman (1960) presented a patient to the Royal Society of Medicine who developed pemphigoid six months before rectal bleeding; an adenocarcinoma of the rectum was later removed surgically. At that time Wilson (1960) recalled two cases, with carcinoma of breast and stomach, and Hellier (1960) mentioned pemphigoid with carcinoma of breast and uterus in his own experience. He thought the association to be not uncommon, and suggested that in all patients with pemphigoid a search should be made for a neoplasm.

A year later Marks (1961) demonstrated a further case of pemphigoid of skin and buccal mucosa, which had been preceded for seven months by a malignant melanoma that later metastasized. Gold (1961) then stated that he had seen two patients with pemphigoid and carcinoma of the lung where buccal involvement had been prominent. Wilson recalled that the patient he had seen with breast carcinoma had had mouth lesions (this is presumably the case mentioned a year earlier).

Chadfield and Kanagasundaram (1962) described a case of carcinoma of the bronchus and a bullous eruption, with marked ocular and buccal manifestations, which could be pemphigoid. Sneddon (1963) recorded carcinoma of the stomach with a bullous rash and ocular lesions.

There are thus nine previous cases in the literature, three with a primary tumour in the bronchus, two in the breast, one each in the uterus, rectum, and stomach, and one malignant melanoma. The association of pemphigoid with carcinoma of the pancreas has not previously been recorded.

Of the seven cases where details are known, five had mouth lesions. This incidence is far in excess of that found usually in pemphigoid; indeed, Rook and Waddington (1953) give a figure of 5% of cases as having buccal bullae. It could well be that mouth involvement is a peculiarity of patients with neoplasms. Further study is needed to ascertain whether the relationship is causal or not.

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