GLUCAGONOMA SYNDROME

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Glucagonoma is a rare syndrome with pancreatic tumour, necrolytic migratory erythema, weight loss and diabetes mellitus. Two such cases are reported here to highlight the variety of presentation of this syndrome and also to stress the importance of early diagnosis of this condition.

Key words : Necrolytic migratory erythema, Diabetes

Introduction

The skin can be a mirror of various internal diseases. Some of the skin lesions if recognised early, may enable both the dermatologists and the physicians alike in the early detection and treatment of the more serious internal diseases. Necrolytic migratory erythema is one such paraneoplastic skin lesion associated with glucagon secreting alpha cell tumour of the pancreas, diabetes mellitus, weight loss, anaemia and elevated serum glucagon levels. We are reporting two cases here to stress the importance of early diagnosis of this entity.

Case 1

A 42-year -old housewife presented with extensive crusted and erosive lesions since 6 years with exacerbations and remissions. She also complained of diarrhoea on and off, progressive loss of weight, weakness and occasional fluid filled lesions over the trunk. The medications received earlier were ineffective. On examination, she appeared pale and thin, weighing 30 kgs. There was diffuse thinning of hair over the scalp and generalised skin lesions in the form of annular plaques (Fig.1) with polycyclic margins with erosions, crusts and a few pustules at the periphery. The predominant areas involved were the trunk, groins, perianal area and legs. A mild glossitis and stomatitis were also noted. However systemic examination was within normal limits.

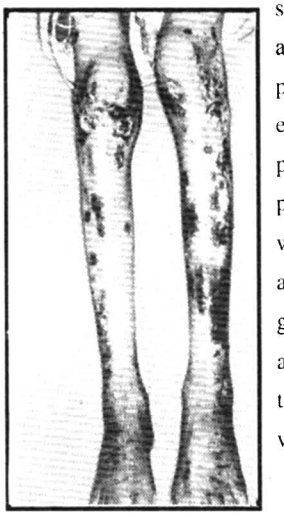

Fig 1. Annular and crusted plaques with polycyclic margins of necrolytic migratory erythema (Case-1).

Laboratory tests showed normocytic, normochromic anaemia, elevated blood sugar levels, raised ESR, glycosuria, proteinuria and reduced total protein and albumin levels. Her skin biopsy revealed hyperke- ratosis, acanthosis and

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perivascular lymphocytic infiltrate. CT scan of abdomen showed a tumour mass in the tail of pancreas.

Based on these findings, a diagnosis of glucagonoma syndrome was made and the patient was put on intravenous amino acid infusions apart from topical management. This led to partial improvement in skin lesions. Later distal pancreatectomy with splenectomy was done and the excised tissue on histopathological examination, confirmed the mass as malignant islet cell tumor.

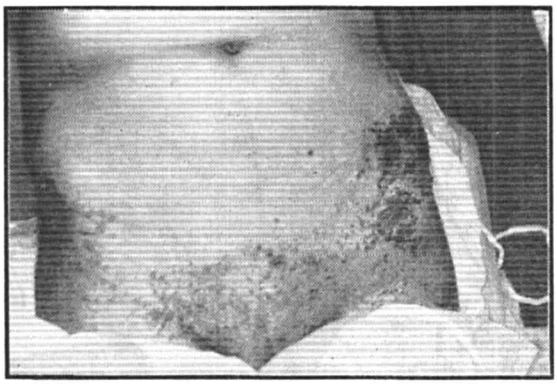

Fig 2. Erythematous and crusted plaques with polycyclic margins of necrolytic migratory erythema (Case-2).

Resection of the tumor resulted in dramatic improvement both in her skin lesions and her general condition. The patient had only post inflammatory hyperpigmentation when examined after 9 months.

Case 2

A 62-year -old woman presented with generalised crusted and erosive skin lesions along with weight loss, loss of appetite and occasional vomiting of 6 months duration. She was a known diabetic since 2 years. The treatments received earlier for her skin lesions were ineffective.

On examination she appeared ill and weighed 37 kgs. Her vital signs were normal. There was diffuse scaling of the scalp and mild scaling and erythema of the nasolabial folds. Skin lesions in the form of scaling. brownish crusting and focal erosions in the background of erythema were present over the axillae, abdomen, flanks, buttocks, perineal area and legs (Fig.2). Her systemic examination was within normal limits. Investigations revealed normocytic normochromic anaemia, raised ESR, elevated blood sugar levels, glycosuria, reduced total serum protein and albumin levels and reduced total serum amino acid levels. Her liver enzyme levels were also raised. CT scan of the abdomen showed a tumor mass in the body of pancreas with multiple secondaries in the liver. Based on these findings, she was diagnosed to have glucagonoma syndrome in advanced stage.

As the general condition of the patient was not good, she was put on conservative management with nutritional supplements, topical emollients and intravenous amino acid infusions. This led only to a partial improvement in her skin lesions. She was planned to undergo a debulking operation. But in the mean time, her general condition deteriorated and the patient expired due to pulmonary embolism.

Discussion

The diagnostic features of necrolytic migratory erythema have been reviewed by several authors.[1-3] They include a chronic migratory cutaneous eruption with advancing borders, that often contain vesicopustules. Patients also usually have glossitis, angular cheilitis, blepharitis, weight loss, and abnormal glucose tolerance.[2] The patients described here had eruptions and histopathologic findings identical with those of necrolytic migratory erythema and both were ultimately shown to have a pancreatic tumour. However we could not estimate the plasma glucagon levels because of the nonavailability of the test.

Though glucagonomas are slow growing tumours, nearly 50-80% are malignant.[4] Several theories have been proposed to explain the mechanism of the cutaneous eruption. Hyperglucagonaemia, zinc deficiency, fatty acid deficiency, hypoaminoacidaemia and hepatic impairment have all been considered as potential causes.[5,6] The treatment of choice is surgical resection. In case of metastatic tumours, however, palliative debulking could be supplemented with chemotherapeutic agents like streptozotocin and dacarbazine.[6]

Since glucagonoma is a slow growing tumour and good recovery is possible after surgical resection, an early diagnosis is mandatoy. Therefore, a high degree of clinical suspicion is essential to diagnose this otherwise fatal entity early in its course.

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