THE AUSTRALASIAN COLLEGE
OF DERMATOLOGISTS

Necrolytic Migratory Erythema

Also known as … Glucagonoma syndrome, pseudo-glucagonoma syndrome

What is Necrolytic Migratory Erythema?
Necrolytic migratory erythema (NME) is a rare, painful, eroded skin eruption that is highly associated with an underlying glucagonoma (a type of pancreatic tumour). This is why sometimes it is also called "glucagonoma syndrome". NME usually occurs in mid-age patients. Men and women are affected equally.

What causes Necrolytic Migratory Erythema?

Most cases of NME are caused by an underlying glucagonoma, a very rare tumour. Only 400 cases of glucagonoma have been reported to date.

NME may also be associated with disorders that mimic a glucagonoma, leading to the term "pseudo-glucagonoma syndrome". These disorders may include pancreatic insufficiency, celiac disease, intestinal malabsorption syndromes, inflammatory bowel disease, cirrhosis, non-pancreatic malignancies and myelodysplastic syndrome.

What does Necrolytic Migratory Erythema look like?

The rash usually starts as a non-specific itchy or painful patch of redness. The body sites commonly affected include the groin, genitals, and buttocks. It then evolves to form an expanding, round, crusty and eroded raised eruption. It tends to wax and wane over weeks. It may look like other common skin diseases such as eczema or psoriasis. Some patients may also have inflamed corner of lips, and inflamed tongue.

What other problems can occur with Necrolytic Migratory Erythema?

Due to the underlying glucagonoma, patients often present with weight loss (most common), anaemia (also quite common), tiredness, sugar intolerance, diarrhoea, neuropsychiatric symptoms and deep vein thrombosis.

How is Pseudoxanthoma Elasticum diagnosed?

The diagnosis is usually made by a dermatologist, often in conjunction with a gastroenterologist. A skin biopsy is useful, NME shows characteristic findings under the microscope. Initial screening blood tests may include glucagon levels. Further imaging studies are often required such as a CT scan.

How is Necrolytic Migratory Erythema treated?

NME management should target its underlying cause. In cases of glucagonoma, surgery is the cornerstone of management. However, often, the disease has already metastasised by the time of diagnosis and the role of surgery may be limited in these cases. In metastatic disease, oncologists

may prescribe chemotherapy. Somatostatin-analogues reduces the secretion of glucagon and may provide good control of symptoms.

What is the likely outcome of Necrolytic Migratory Erythema?

About half of the patients with NME secondary to glucagonoma have metastatic disease at the time of diagnosis, hence the prognosis is likely poor. However, if the underlying cause of NME can be treated e.g. tumour removed, then the skin changes will likely resolve.

This information has been written by Dr Cathy Zhao and Dr Tanumay Raychaudhury

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