Differentiating non-paraneoplastic from Glucagonoma associated necrolytic migratory erythema

Dear Sir,

There are 10 reported cases of non-paraneoplastic necrolytic migratory erythema (NME) for the past 20 years[1-6]. We have seen four cases of NME confirmed clinically and histopathologically for the past 10 years at Hospital Kuala Lumpur. Two patients had the classical Glucagonoma Syndrome with liver metastasis. The third patient had cirrhosis associated with hepatitis C infection. The fourth patient had past history of hepatitis B infection and rheumatoid arthritis. The CT scan and coeliac angiogram did not reveal any presence of pancreatic tumour in the third and fourth patients.

Retrospective study of these four cases indicates that the presence of a severely pruritic and extensive NME involving almost the entire trunk and limbs points more towards paraneoplastic NME especially if there is constitutional symptoms and sign of anaemia and hypoproteinaemia. Surgical removal of the pancreatic tumour led to clearance of NME in Glucagonoma patients. Despite the presence of liver metastasis, debulking of pancreatic tumour can induce remission of cutaneous lesions. The recurrence of NME in one of them alerted us to the presence of recurrent pancreatic tumour. One wonders whether triggering of NME requires the presence of pancreatic tumour. Octreotide in our experience does cause NME to clear but it does not improve the intractable diarrhoea nor prevent tumour growth.

The NME in the two patients with viral hepatitis associated liver disease tends to localise acrally and remains non-pruritic. Both has low serum zinc levels. We noted clearance of the NME with administration of zinc sulphate 200 mg three times a day initially followed by a maintenance of 200 mg daily. Stopping zinc supplement caused the reappearance of NME. This leads one to ask whether there is any relationship between this and Acrodermatitis enteropathica which responds to zinc supplement too. In non-paraneoplastic NME, the correction of the primary cause clearance of NME. This is seen with the administration of gluten free diet for coeliac disease associated NME, instituting total parenteral nutrition and replacing pancreatic enzymes for pancreatic insufficiency associated NME[3].

Diagnosis of NME can be difficult but an early recognition of NME clinically and histologically is very important to establish if it is associated with a Glucagonoma, early treatment of which will result in better outcome for the patient.

ACKNOWLEDGMENTS

We thank Dr M A Norizan, Prof S Samad, Prof K Khalid, Prof T T Tan and Dr Mehta for managing the patients.

Yours Sincerely,
R Rohna,
H H Suraiya

Department of Dermatology,
Hospital Kuala Lumpur,
Jalan Pahang,
50586 Kuala Lumpur,
Malaysia

REFERENCES

1. Marinkovich MP, Botella R, Datloff J, et al. Necrolytic migratory erythema without Glucagonoma in patients with liver disease. J Am Acad Dermatol.
2. Tanner M, Braschh J, Christopher E. Erythema necroticans migrams without glucagonoma, Hautarzt 1994; 45:480-3.
3. Thorisdottir K, Camisa C, Tomecki KJ, et al. Necrolytic migratory erythema: a report of three cases. J Am Acad Dermatol 1994; 30:324-9.
4. Kelly CP, Johnston CF, Nolan N, et al. Necrolytic migratory erythema with elevated enteroglucagon in celiac disease. Gastroenterology 1989; 96:1350-3.
5. Blackford S, Wright S, Robert DL. Necrolytic migratory erythema without glucagonoma: the role of dietary essential fatty acids. Br J Dermatol 1991; 125: 460-2.
6. Burton JL, Zinc and essential fatty acid therapy for necrolytic migratory erythema. Arch Dermatol 1993; 129:246.