

Images in paediatrics

Severe refractory dermatitis and cystic fibrosis

An 80-day-old infant was admitted for severe dermatitis, resembling acrodermatitis enteropathica, characterised by erythematous scaly plaques with crusts and ulcerations, predominantly in the diaper area, perioral area and extremities (figure 1). She had generalised severe dermatitis from the first weeks of life unresponsive to conventional treatment. At admission, the patient was below the 3rd percentile for weight and at the 25th percentile for length, both of which were at the 50th percentile at birth. She was exclusively breast fed and any allergic sensitisation was excluded. Laboratory findings included low levels of albumin (16.2 g/dl) and haemoglobin (5.5 g/dl). Albumin and blood transfusions were required. Being deficient in zinc levels (43.2 µg/dl), she was treated with oral zinc supplementation (40 mg/day). The neonatal screening for cystic fibrosis (CF) was negative, but steatocrit was 28% and chymotrypsin was 1 U/g in the stools. The sweat chloride test was attempted but insufficient amount of sweat was obtained because of severe dermatitis. Based on a suspicion of CF, pancreatic enzyme supplementation was started. The dermatitis showed gradual resolution, preceded by extensive desquamation. This was associated with clinical and laboratory improvements. Finally, mutation analysis for CF showed the homozygous mutation $\Delta F508$.

Skin manifestations of CF are underestimated.¹ Acrodermatitis enteropathica-like dermatitis may concur and allows a correct and early diagnosis in atypical presentations of CF, particularly in association with failure to thrive or pathological laboratory findings (anaemia and hypoalbuminaemia).^{2–4} Even in cases of negative neonatal screening, the sweat test and/or genetic analysis are required if there are symptoms suggestive of CF.¹

Diego G Peroni,¹ Claudia Banzato,¹ Sara Guerresi,¹ Donatella Schena,² Marco Deganello,¹ Attilio L Boner,¹ Giorgio L Piacentini¹

¹Department of Paediatrics, University of Verona, Verona, Italy

²Section of Dermatology and Venereology, Department of Medicine, University of Verona, Verona, Italy



Figure 1 Erythematous scaly plaques with crusts and ulcerations in the diaper area and legs.

Correspondence to Professor Diego Peroni, Department of Paediatrics, University of Verona, Policlinico GB Rossi, Piazzale Scuro, 37134 Verona, Italy; diego.peroni@univr.it

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Accepted 29 September 2011

REFERENCES

1. **Bernstein ML**, McCusker MM, Grant-Kels JM. Cutaneous manifestations of cystic fibrosis. *Pediatr Dermatol* 2008;**25**:150–7.
2. **Perafan-Riveros C**, Franca LF, Alves AC, *et al*. Acrodermatitis enteropathica: case report and review of the literature. *Pediatr Dermatol* 2002;**19**:426–31.
3. **Muñiz AE**, Bartle S, Foster R. Edema, anemia, hypoproteinemia, and acrodermatitis enteropathica: an uncommon initial presentation of cystic fibrosis. *Pediatr Emerg Care* 2004;**20**:112–14.
4. **Patrizi A**, Bianchi F, Neri I, *et al*. Acrodermatitis enteropathica-like eruption: a sign of malabsorption in cystic fibrosis. *Pediatr Dermatol* 2003;**20**:187–8.



Severe refractory dermatitis and cystic fibrosis

Diego G Peroni, Claudia Banzato, Sara Guerresi, et al.

Arch Dis Child published online October 28, 2011

doi: 10.1136/archdischild-2011-300731

Updated information and services can be found at:

<http://adc.bmj.com/content/early/2011/10/28/archdischild-2011-300731.full.html>

These include:

References

This article cites 4 articles

<http://adc.bmj.com/content/early/2011/10/28/archdischild-2011-300731.full.html#ref-list-1>

P<P

Published online October 28, 2011 in advance of the print journal.

Email alerting service

Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

Advance online articles have been peer reviewed, accepted for publication, edited and typeset, but have not yet appeared in the paper journal. Advance online articles are citable and establish publication priority; they are indexed by PubMed from initial publication. Citations to Advance online articles must include the digital object identifier (DOIs) and date of initial publication.

To request permissions go to:

<http://group.bmj.com/group/rights-licensing/permissions>

To order reprints go to:

<http://journals.bmj.com/cgi/reprintform>

To subscribe to BMJ go to:

<http://group.bmj.com/subscribe/>