

CLINICAL ALLERGY IMAGES

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This is one of a series about clinical images relevant to the practice of allergy and immunology. Please submit interesting images and discussion for publication to the Section Editor, Dr George du Toit (details below).

ECZEMATOUS SYNDROMES IN CHILDHOOD

Eczema is a common dermatological syndrome characterised by inflammation of the skin, redness, itching, and the outbreak of lesions that may discharge serous matter and become encrusted and scaly (Fig. 1). The intense itching and disfigurement associated with



Fig. 1. Severe eczema.

Table 1. Selection of eczema-associated syndromes and presenting features

Primary immunodeficiencies

Wiskott-Aldrich syndrome: a primary immunodeficiency disorder characterised by recurrent infection, eczema, and micro-thrombocytopenia (Fig. 2). Inherited in an X-linked recessive pattern.

Hyper-IgE syndrome: a primary immunodeficiency characterised by recurrent skin abscesses, recurrent pneumonia with pneumatocele formation (Fig. 3), eczema, eosinophilia, and elevated levels of serum IgE.

Ataxia telangiectasia: an autosomal recessive primary immunodeficiency, characterised by cerebellar ataxia. It usually presents by the age of 2 years, with ataxia (upper and lower limbs), speech and eye movement abnormalities. Other important features are translocations, immunodeficiency, a high serum alpha-fetoprotein concentration, growth retardation, telangiectasia – most noticeably on the bulbar conjunctiva (Fig. 4) – and a very high risk of developing a lymphoid tumour. Patients also show an increased sensitivity to ionising radiation. Eczematous skin rashes are commonly associated.

X-linked agammaglobulinaemia (also known as XLA or Bruton's sex-linked agammaglobulinaemia): those affected suffer recurrent and severe bacterial infections. This usually begins in early childhood although the condition is sometimes diagnosed in young adults who have a milder form of the disease. Children may also have enlarged joints and lack of the usual lymphoid tissue such as the tonsillar beds.

Immunological disorders

Dermatitis herpetiformis: a chronic, extremely itchy rash consisting of bumps and blisters. Dermatitis herpetiformis is linked to sensitivity of the intestine to gluten in the diet (coeliac sprue). Onset is usually in young adulthood.

'Eczematous-like' skin diseases

There are many other eczema-like variants, including: **seborrhoeic dermatitis, allergic contact dermatitis, nummular dermatitis, asteatotic eczema, lichen simplex chronicus and palmoplantar psoriasis**. The baboon syndrome is a form of 'systemic contact dermatitis'.

Infectious skin diseases

Scabies and dermatomycosis

Genetic syndromes

Netherton syndrome: Netherton syndrome is a rare disorder inherited in an autosomal recessive pattern consisting of ichthyosiform dermatosis, hair shaft abnormalities (trichorrhexis invaginata), and an atopic diathesis.

Dubowitz syndrome: features include intra-uterine growth retardation (with primordial shortness of stature), microcephaly, variable degrees of eczema and mental retardation, and a characteristic facial appearance. The syndrome is presumed to be recessively inherited.

Nutritional deficiencies

Acrodermatitis enteropathica (Fig.5): an inborn error of metabolism resulting in zinc malabsorption and severe zinc deficiency. Dermatitis, diarrhoea, mood changes, anorexia, and neurological disturbance are described. Growth retardation, alopecia, weight loss and recurrent infections may be present in toddlers and schoolchildren.

There are many other **nutritional deficiencies** which may present with eczema-like rashes. These include: **copper deficiency, marasmus, kwashiorkor, vitamin C deficiency**, among others.

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eczema result in significant reduction in the quality of life of both patients and their caregivers.

In young children, eczema is frequently the harbinger of the 'allergic march'. The skin barrier may serve as a portal for both aeroallergen and food-allergen sensitisation. Indeed, the overwhelming majority of food allergic patients either have current eczema or report a history thereof. Eczema is also a strong predictor for the development of subsequent asthma. It is therefore important that the attending physician consider co-morbid conditions such as asthma, rhinitis and food allergy, when faced with a young patient who has 'typical' eczema.

However, not all that is dry and itchy – a diagnosis of eczema makes! It is therefore the prerogative of the physician to consider differential diagnoses when faced with atypical 'eczematous' rashes or patients who present with eczema in association with systemic symptoms and/or signs. There are many syndromes which include eczema as one of the cardinal signs. Although these syndromes are rare, they are nonetheless important to consider when dealing with 'atypical' eczema presentation. Table I details a selection of syndromes and presenting features.

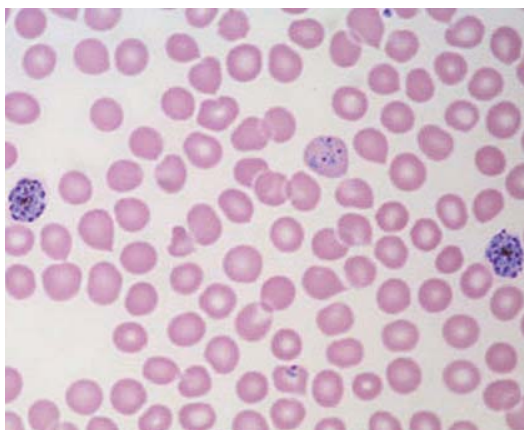


Fig. 2. Thrombocytopenia.

Declaration of conflict of interest

The author has no conflict of interest.



Fig. 3. Pneumatocele.



Fig. 4. Telangiectasia on the bulbar conjunctiva.



Fig. 5. Zinc deficiency.

ERRATA

In the article 'Metals and allergy' in our last issue (June 2006; 19: 62) some information was incorrect.

The authors' details are as follows:

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