CLINICAL IMAGES

Mycoplasma pneumatoceles

Savvas Andronikou, Afshin Eimany, Philip J Robinson, Andrew Kemp

Hyperimmunoglobulin E syndrome (HIE) or Job's syndrome is a rare complex immune disorder characterised by chronic eczema and recurrent cutaneous, subcutaneous, paranasal sinus and pulmonary infections primarily due to *Staphylococcus aureus* and fungi. Pneumatoceles characteristically develop as a result of staphylococcal infection. We present an unusual case of Job's syndrome where the patient lacked the characteristic cutaneous manifestations and presented with a mycoplasma pneumonia which was followed by the formation of multiple pneumatoceles. The causal relationship between mycoplasma and pneumatocele formation is suggested by the patient's response to appropriate antibiotic treatment with the formation of pneumatoceles.

The patient developed a facial rash at 3 weeks of age, diagnosed as infantile acne, occipital boils at 6 and 6.5 months and boils on the buttock at 22 months. An immunoglobulin E (IgE) level at 22 months was markedly elevated at 8 320 kU/l normal (0 - 25) (Fig. 1). At 23 months he developed a moist infected area on his scalp, which grew *S. aureus*. He did not have generalised atopic dermatitis. At 29 months he presented with a right upper lobe pneumonia and a pleural effusion, which was drained. No organisms were identified and mycoplasma-specific antibodies were not detected by particle agglutination test. The pneumonia resolved with the formation of right upper lobe pneumatoceles (Figs 2a and 2b). IgE levels

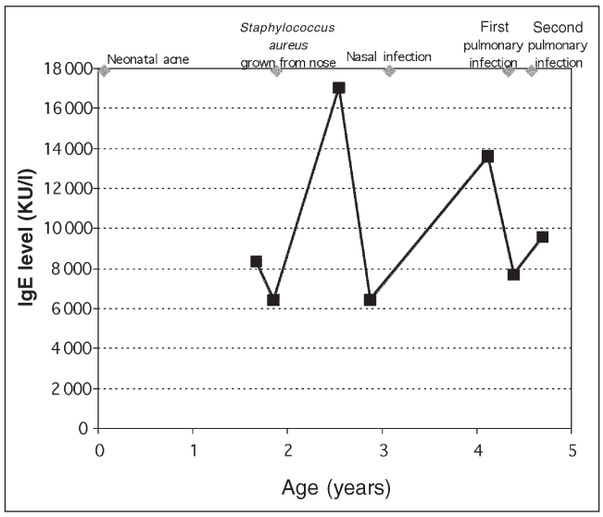
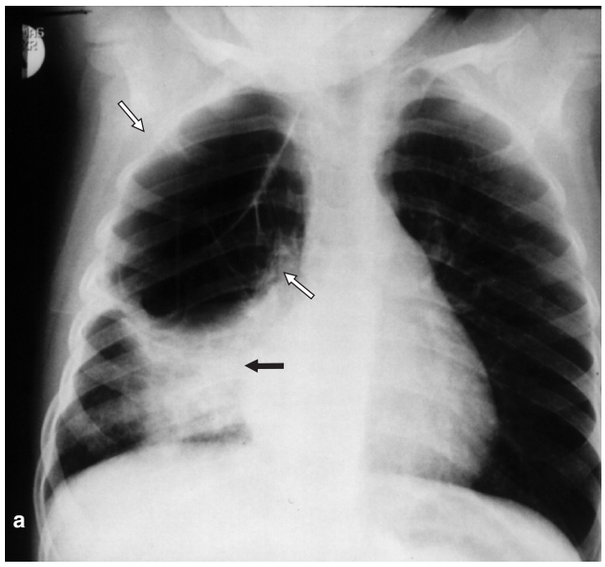
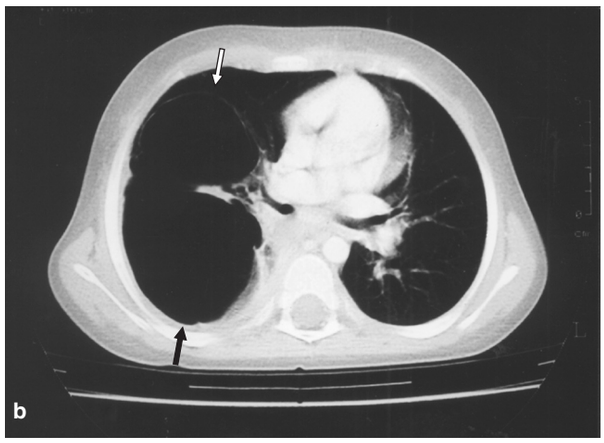


Fig. 1. Patient's IgE levels with indicators of clinically significant events.





Figs 2a and b. Initial imaging at second presentation. (a) Radiograph demonstrates a large pneumatocele of the right upper lobe (white arrows) and acute infective change in the lower lobe (black arrow). (b) CT scan confirms the pneumatocele in the right upper lobe (arrows) without intracavitory body or fluid level.

Department of Paediatric Radiology, Red Cross War Memorial Children's Hospital and School of Child and Adolescent Health, University of Cape Town

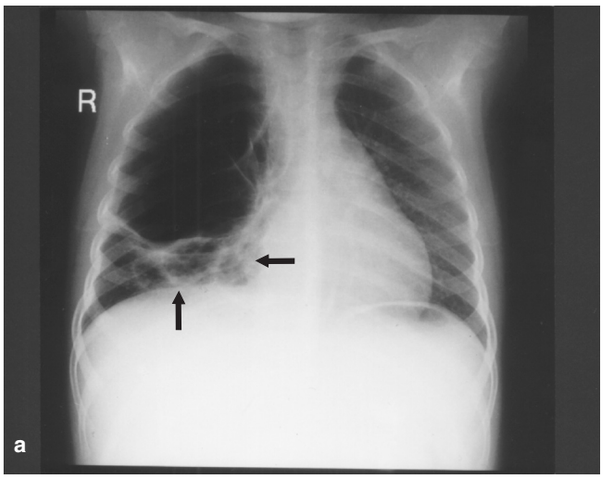
Savvas Andronikou

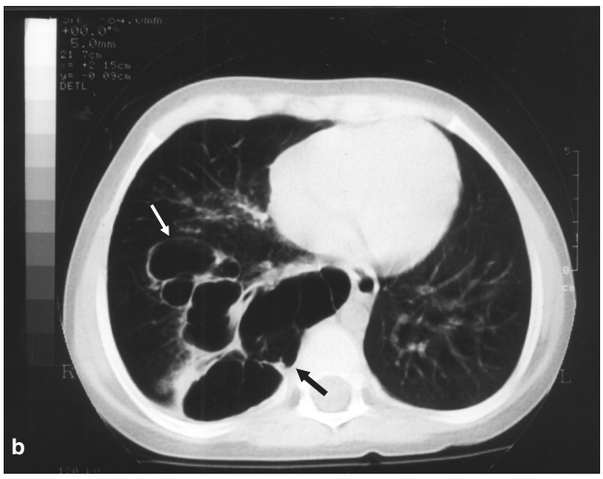
Departments of Paediatric Radiology, Paediatric Immunology and Paediatric Respiratory Medicine, Royal Children's Hospital, Melbourne, Australia

Afshin Eimany
Philip J Robinson
Andrew Kemp

(7 670 KU/l) remained elevated (Fig. 1).

He presented again at 31 months with right lower lobe pneumonia, which was treated with flucloxacillin and cefotaxime. Radiographs revealed a large right upper lobe pneumatocele and consolidation in the right lower lobe. A swinging fever continued until day 7 of treatment. Serology on day 6 of treatment was positive for mycoplasma (particle agglutination positive, mycoplasma-specific IgM enzyme immunoassay positive). A macrolide was commenced and his temperature settled within 24 hours and remained normal until discharge from hospital 15 days later. This was associated with resolution of the right lower lobe consolidation and the formation of pneumatoceles in the right lower lobe (Figs 3a and 3b). The patient was discharged after a decision not to remove the pneumatoceles surgically.





Figs 3a and b. Imaging after clinical response to erythromycin therapy. (a) Pneumatoceles in the right lower lobe (arrows), the site of the current infection, in addition to those shown in Fig. 2. (b) CT scan confirms pneumatoceles in the right lower lobe (arrows) with only a small amount of residual air-space disease.

Discussion

HIE is a rare syndrome that includes a deficit of immunity with high levels of IgE specific for *S. aureus* and *Candida albicans*.[1-6] The syndrome appears to be an autosomal dominant disorder with variable expression and incomplete penetrance.[5] It usually presents before the age of 2 years[3] as a multisystem disorder affecting cutaneous tissue, dentition, the skeleton and the lower respiratory tract.[5] The major characteristics of the disease are recurrent staphylococcal skin abscesses, pneumonia with pneumatocele formation and extreme elevations of IgE. These features in varying combinations are considered diagnostic.[5]

Eczematoid rashes are considered universal in early life and 'cold' abscesses (so called because of the lack of surrounding inflammation), which yield *S. aureus*, are characteristic of the syndrome.[5,6] However, the triad of cold abscesses, pneumonia and high IgE occurs in only 77% of patients.[5] Although our subject did not have an eczematous dermatitis, which is almost universally described, the recurrent cutaneous infections, extreme elevation of IgE and development of pneumatoceles on two occasions following lower respiratory tract infections are indicative of the HIE syndrome.

Pulmonary involvement is common. One large study[5] demonstrated that 26 of 30 patients had pneumonia, and of these 23 developed pneumatoceles, which is considered a characteristic finding in this syndrome, the most common pathogens identified being either *S. aureus* or *Haemophilus influenzae*. Superinfection of pneumatoceles is associated with *Pseudomonas aeruginosa* and *Aspergillus fumigatus*.[5,6] The natural history of staphylococcal pneumatoceles is resolution with fibrosis, but in HIE they frequently fail to resolve spontaneously.[4] In the HIE syndrome the pathogenesis of pneumatoceles is thought to be the same as that resulting in subcutaneous 'cold abscesses', so called because of the lack of surrounding inflammatory response.[5,6] In our case the serological findings, failure of response to antistaphylococcal therapy and rapid response to a macrolide are consistent with mycoplasma infection. It is possible that the pneumatoceles resulted directly from the mycoplasma infection or alternatively that there was undetected bacterial infection in addition to the mycoplasma which contributed to pneumatocele formation. The appearance of pneumatoceles after infection with mycoplasma in either normal subjects or subjects with the HIE syndrome has not previously been reported in the literature.

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