

Mycoplasma pneumonia, Stevens-Johnson syndrome, and chronic obliterative bronchitis

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Stevens-Johnson syndrome is a variant of erythema multiforme characterised by a polymorphous vesicular and bullous eruption of the skin and mucous membranes, with systemic manifestations of variable severity.¹ Pulmonary complications, which may be fatal, generally occur during the acute phase of the illness.²⁻⁴ In this paper we present a patient who died of a chronic obliterative bronchitis 10 months after the rash had resolved.

Case report

An 8 year old girl was admitted on 4 January 1982 with a vesicular and papular rash involving the skin, mouth, and vulva. Six days before she had developed a respiratory tract infection, which had been treated with amoxycillin, and she still complained of cough and wheeze. She had previously been healthy but had an asymptomatic atrial septal defect. On examination apart from the rash there was a fever of 39°C and severe conjunctivitis. Rhonchi and crepitations were heard in both lung fields and a chest radiograph showed linear opacities in the left upper lobe. Subsequently the rash became bullous; there was serological evidence of a mycoplasma infection and cold agglutinins were present. A diagnosis of Stevens-Johnson syndrome secondary to mycoplasmal pneumonitis was made. She was treated with erythromycin and discharged after eight days. At this stage the rash was subsiding and the respiratory symptoms, although still present, had improved.

In February she presented again with progressive dyspnoea, wheezing, and exertional cyanosis. The response to steroids and bronchodilators was poor and recovery was complicated by mediastinal emphysema. Subsequently her respiratory symptoms persisted and she became emaciated and emotionally disturbed. She was admitted for the last time in October with dyspnoea, cyanosis, a collapsed right lung, and a hyperinflated left lung. At bronchoscopy the main and segmental airways were normal. An open lung biopsy showed non-specific patchy dilatation and collapse of the lung parenchyma, with a little mucoid material in the bronchioles. No bronchi were present in the specimen. Despite vigorous treatment she died 17 days later.

At necropsy about 100 ml of straw coloured fluid was present in both pleural cavities. The left lung was hyperinflated and the right lung collapsed. The trachea and the larger bronchi were inflamed but otherwise normal. Hilar

lymph nodes were slightly enlarged. There was a fibrinous pericarditis. The heart was of normal configuration but the foramen ovale was patent at its superior end.

Microscopically there was an obliterative bronchitis affecting many of the subsegmental airways of both lungs. Affected bronchi were scarred and flattened or narrowed to 1-5 mm in diameter. Cartilage plates were drawn inwards, the lamina propria was thickened and folded, and lumina were often partially or completely occluded by cellular fibrous tissue (fig 1). An abrupt transition from patency to complete stenosis could be traced in some sections. Glandular tissue was represented only by occasional cystic spaces, which were sometimes large enough to distort the bronchial lumen (fig 2). In some bronchi the muscle coat was well preserved but in others it was absent. The surviving lining epithelium consisted of columnar ciliated cells; there was no goblet cell hyperplasia. The pulmonary vasculature was normal, although pulmonary arteries accompanying stenosed bronchi appeared disproportionately large. Bronchial arteries, on the other hand, were dilated and enlarged (fig 1).

A few subsegmental bronchi were dilated rather than narrowed and in these cartilage, gland, and muscle were completely replaced by fibrous tissue. In bronchi of segmental size and larger there was hyperplasia of gland and muscle but no stenosis or scarring. The spleen contained a few necrotic granulomas of uncertain nature.

Discussion

Stevens-Johnson syndrome is now accepted as a secondary disorder, although the underlying cause may not always be apparent.⁴ Up to 30% of cases are preceded by an atypical pneumonitis and an association with *Mycoplasma pneumoniae* infection is well established.^{4,5} Other precipitating factors include connective tissue disorders, malignant tumours, various microbial diseases, and exposure to drugs, vaccines, and physical agents.⁴

Stevens-Johnson syndrome is a generalised disease and symptoms referable to the heart, central nervous system, gastrointestinal tract, and urinary tract may occur. Bronchopulmonary manifestations during the acute phase are well documented,¹⁻⁵ but it is not clear to what extent they are due to primary lesions in the respiratory tract; the roles of secondary infection and pre-existing pneumonia are still undefined.⁴ Few data are available on the pathology of such cases. Changes that have been reported are suggestive of a primary atypical pneumonia.² Pneumothorax due to rupture of bullous lesions in the

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Fig 1 Collapsed bronchus showing complete obliteration of the lumen by fibrous tissue. (Elastic-Van Gieson, $\times 27$.)

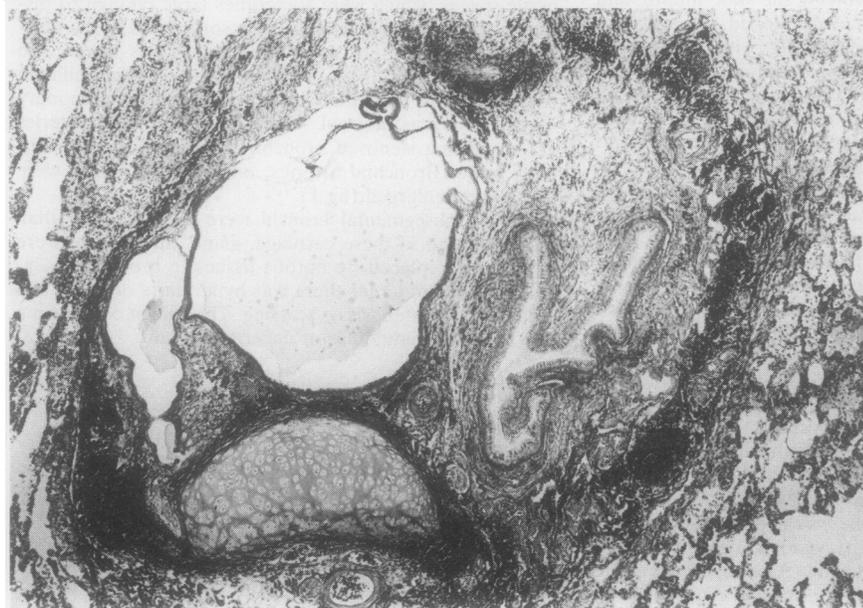


Fig 2 Bronchus showing cystic atrophy of glandular tissue, which has displaced the lumen to the right. (Elastic-Van Gieson, $\times 27$.)

pleura and mediastinal and subcutaneous emphysema have also been described.^{4,6}

Healing may be complicated by conjunctival adhesions, fibrosis of the lachrymal apparatus, oesophageal stricture, or vaginal stenosis,^{4,7} but so far as we are aware there are no previous reports of chronic obliterative bronchitis. There are two clinical descriptions in published reports of children who developed airways obstruction after Stevens-Johnson syndrome,^{8,9} but unfortunately no tissue was available for study in either of these cases.

The cause of the obliterative bronchitis in this case is uncertain. Although Macleod's or Swyer-James syndrome may complicate *M pneumoniae* infection⁹ the bronchioles rather than the bronchi are primarily affected. This child would appear to have had a tracheobronchitis due to a combination of Stevens-Johnson syndrome and secondary infection. Later the exudate was cleared from the larger airways and healing proceeded normally, but in the smaller airways it was retained and subsequently became organised.

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