All cases earlier reported developed the symptoms within 2-8 weeks of starting the offending drug. Our patient also presented with the triad of fever, rash, lymphadenopathy and hepatosplenomegaly, abnormal liver functions and leucocytosis about one-and-half-months after starting the drug.

Due to fever, lymphadenopathy and rash, this condition has to be differentiated from viral infections especially infectious mononucleosis and also bacterial infections. Awareness of this entity helps in an early diagnosis. Treatment consists of omitting the offending drug and systemic steroids, if necessary. Our patient was managed symptomatically.

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Chronic Lung Disease Following Stevens-Johnson Syndrome

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Stevens-Johnson syndrome (SJS) is an acute self-limited eruption of the skin and mucous membranes which represents a hypersensitivity reaction to various etiologic agents(l). Organ involvement, including acute pulmonary lesions, have been described previously(2,3). However, chronic lung disease is an extremely rare complication of SJS. We present here a case of chronic lung disease following SJS in a previously well child.

Case Report

An 8-year-old boy who was well till 2 months prior to admission, received oral

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medication for an upper respiratory tract infection. Sixteen hours later, he developed redness in both eyes followed by a generalized erythematous, non-itchy, maculopapular skin rash involving the face, palms and soles. He also had high grade intermittent fever. The rash progressed to become vesiculobullous in nature with peeling of skin. He also developed oral ulcers with foul smelling purulent oral discharge, eye discharge with symblepharon, wheezing, cough and breathlessness. These symptoms lasted for 2 weeks, following which the skin lesions healed but the eye condition worsened with resultant visual impairment. He was treated with prednisolone during this acute phase with which his wheezing improved minimally. Two weeks prior to admission to our hospital his respiratory distress worsened and he became oxygen dependent, resulting in his referral.

Physical examination revealed a tachypneic child with marked chest indrawing, with no cyanosis or clubbing. Oxygen saturation by pulse oximetry in room air was 80% and with supplementary oxygen (2 lit/min) by nasal cannula it was 95%. Palmar and plantar erythema were observed along with hyperpigmented and hypopigmented macules all over the body. He had cheilosis and ulcers over the oral mucosa and tongue. Both eyes showed circumciliary congestion, peripheral corneal neovascularisation and symblepharon; vision was poor, with finger counting not possible beyond 1 meter distance. Bilateral extensive expiratory rhonchi and fine rales were auscultated over the chest.

Investigations revealed a total leucocyte count of 17,300/cu mm, with a differential count of 5% band forms, 75% neutrophils, 19% lymphocytes and 1% monocytes. The erythrocyte sedimentation rate was 6 mm in 1 hour, serum total bilirubin 0.4 mg/dl (direct 0.2 mg/dl), serum total protein 6.7 g/dl, albumin 4 g/dl, alkaline phosphates 94 U/l, SGOT 32 U/l, SGPT 10 U/l, blood urea 20 mg/dl, serum creatinine 0.4 mg/dl, and serum complement 80%. The cold agglutinin titer in the blood was nil. Bacterial and fungus cultures of the sputum were negative and induced sputum was negative for *Pneumocystis carinii* antigen. Fasting gastric juice was negative on smear for acid fast bacilli. The chest roentgenogram revealed hyperinflated lung fields with pneumatocele formation and a computed tomogram of the thorax revealed extensive bilateral parenchymal lung disease with areas of patchy opacities, pneumatocele formation and patchy areas of emphysema *(Fig. 1)*. Arterial blood gas analysis in room air showed: pH-7.441; pCO_2-34.9 mm Hg; pO_2-33.5 mm Hg; HCO_3-23.4 mmol/lit; tCO_2-24.4 mmol/lit; ABE-0.3 mmol/lit; and O_2 saturation-67.6%

The patient was hospitalized for 4 weeks and treated with oral erythromycin (40 mg/kg/day), prednisolone (1 mg/kg/day), aerosolized salbutamol and ipratropium bromide. As a result of this therapy, his oxygen requirement decreased and he was able to maintain normal oxygen saturation in room air. However, clinical and radiological abnormalities in the lungs persisted. He was discharged home on tapering doses of prednisolone.

Fifteen months later his parents reported that his respiratory distress was present with Grade IV effort intolerance although he was no longer oxygen dependent. His current medications include oral theophylline and some ayurvedic (herbal) products.

Discussion

Acute pulmonary complications of SJS such as pneumonia and bronchiolitis are well documented(3) but reports of chronic

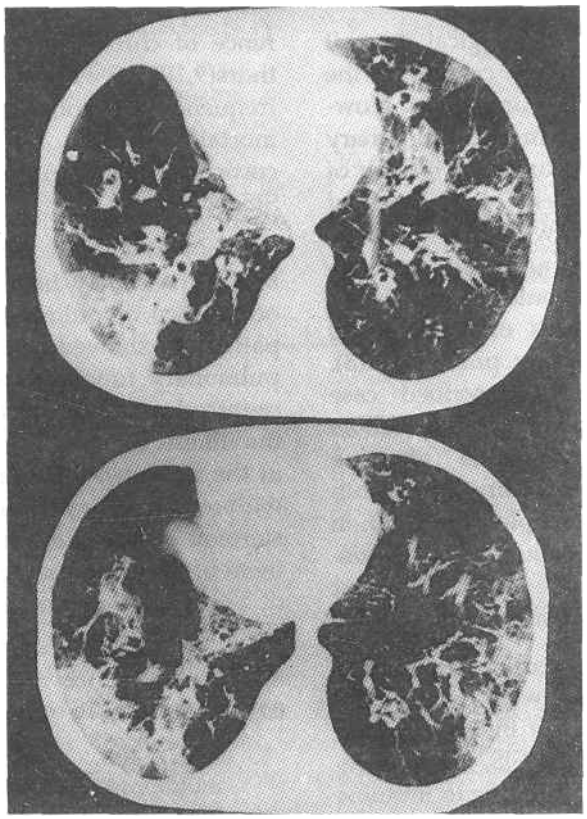

Fig. 1. *Computed tomogram of the thorax shows extensive involvement of both lungs with patchy opacities, pneumatocele and emphysema.*

pulmonary complications are rare. We could find only 3 such reports in the English language literature(4-6).

Our patient presented to us late in the course of his illness after his skin lesions had healed. Yet, his history was characteristic of SJS and he had typical oral and ocular lesions at the time of presentation to our hospital. The severity of the ophthalmic complications is thought to be related to the severity of disease in SJS(7). The severe nature of the ophthalmic lesions, namely, bilateral corneal opacities, synechiae and blindness seen in our patient suggests that he had severe systemic disease during the acute phase of his illness.

There were many similarities between our patient and the three reported cases in the literature. All the children had cough and wheezing in the acute phase, followed by a period with minimal respiratory symptoms with subsequent worsening of these symptoms(4-6). The appearance of chronic respiratory signs and symptoms occurred 2-4 weeks after the onset of SJS in all cases. Radiologic findings such as, areas of atelectasis with local areas of emphysema were common features in all whereas pneumatocele formation as seen in our patient was described in only one of the other reported cases(6). The long term outcome in the reported patients was poor with one patient having died(5) and the rest, including our patient, having persistent pulmonary signs(4,6).

In the absence of histological data, it was difficult to accurately categorize the chronic lung disease in our patient. However, the presence of chronic pulmonary manifestations with waxing and waning of symptoms and the presence of areas of atelectasis interspersed with areas of organizing pneumonia and areas of emphysema on radiography would be consistent with a clinical diagnosis of bronchiolitis obliterans with organizing pneumonia(8). In the only previously described case where histology was available, the authors found obliterative bronchitis affecting many of the subsegmental airways of both lungs with partial or complete obliteration of the lumina with cellular fibrous tissue(7). Thus, SJS could be added to the list of etiologies for bronchiolitis obliterans.

The pathogenesis of chronic pulmonary sequelae in SJS is not well understood. Whether this is a result of the primary infection which resulted in SJS, or a consequence of mucosal damage due to immune complex deposition secondary to SJS or a combination of both is unclear. *Mycoplasma pneumoniae* which is a common predisposing condition for SJS(l) has been associated with bronchiolitis obliterans and organizing pneumonia.

Our patient received corticosteroids in the acute phase after developing respiratory symptoms. This was also true for the case described by Edell *et al.{6)* who postulated that steroid treatment during the acute stages of SJS in their patient may have contributed to the long term detrimental sequelae. Both the patients described by them as well as our patient had pneumatocele formation which could have possibly resulted from weakening of collagen due to steroid administration, with secondary cystic development. Thus, steroid treatment in the acute phase of SJS nay also have contributed to the occurrence of chronic lung disease. Other authors(9,10) have also reported an increased frequency of complications and higher mortality rates among patients treated with corticosteroids -during the acute phase of SJS.

Though steroid treatment during the acute phase may have contributed to the occurrence of pulmonary sequelae in our patient, he showed clinical improvement in pulmonary functions when steroids were re-introduced later in the chronic phase of the illness. Steroids are also recommended in the treatment of bronchiolitis obliterans w4th organizing pneumonia(8). Thus, while steroids may be detrimental in the treatment of the acute pulmonary complications of SJS, their role in the treatment of the chronic pulmonary sequelae is yet to be fully defined.

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