A CASE OF STEVENS-JOHNSON SYNDROME WITH EXTENSIVE SURGICAL EMPHYSEMA

BY

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The condition known as Stevens-Johnson syndrome, a skin eruption resembling erythema multiforme, with lesions of the mouth, conjunctivae, genitalia, and anus, was described by Stevens and Johnson in 1922. A similar condition had been recorded in the last century, but does not appear to have been thought worthy of special notice. The cause of the disease is unknown. It is suggested that the eruption may be due to hypersensitivity or allergy, because some cases give a history of recent upper respiratory infection or of the administration of sulphonamides. A virus has been suggested, but at present there is no proof of this. The condition usually runs a benign course, with full recovery in ten days, but respiratory complications may occur (Liddle, 1951).

The following case of Stevens-Johnson syndrome is reported because of its severity and because of the unusual complication of surgical emphysema.

Case History

A boy aged 9 years 11 months was admitted to hospital on May 1, 1954, with the following history. Five days previously he had complained of headache, and was listless. His throat and conjunctivae were inflamed, and his temperature was 102° F. (38.9° C.). He was started on a course of "tresamide" (each 0.5-g. tablet containing sulphamerazine 0.1 g., sulphadiazine 0.2 g., sulphacetamide, 0.2 g.), which was replaced by chloramphenicol on the fourth day of the illness because of the appearance of a morbilliform rash on the chest. On the next day the rash spread all over the face, chest, and limbs, and many of the lesions became vesicular. He had severe toxæmia, headache, and nausea, and his temperature rose to 105° F. (40.6° C.).

He was admitted to an isolation hospital as a suspected case of smallpox, and thereafter transferred to the Grove Hospital on the sixth day of the illness.

On admission he was seen to be a well-built boy, with no history of previous illness. He was semicomatose and profoundly toxæmic. His temperature was 99° F. (37.2° C.), pulse 108, and respirations 20. There was the dusky-red rash of erythema multiforme on the face, neck, trunk, and limbs. In places the rash was confluent and there were many large vesicles containing yellow fluid. Some vesicles had burst, particularly round the eyes, ears, and neck, and the underlying dermis was fiery red. The lips were purplish, cracked, and bleeding. The buccal mucosa was overlaid with collapsed vesicles, and gingivitis and ulceration of the tongue were present. The conjunctivae were deeply injected and oedematous. He had photophobia, but no corneal lesions were seen. There was some ulceration of the glans penis and dysuria, probably due to lesions within the meatus. There was no urethral discharge. Gross surgical emphysema was present on the chest, stretching from the neck to the lower abdomen. Respiration was a little laboured and there was a suggestion of cyanosis. Anteriorly, breath and heart sounds were difficult to hear, but auscultation of the back of the chest revealed moist crepitations in all areas. The spleen and liver could not be felt. A diagnosis of Stevens-Johnson syndrome was made.

The patient was placed in an oxygen tent and parenteral penicillin (250,000 units six-hourly) and streptomycin (0.5 g. six-hourly) were given. On the day after admission the general condition was unchanged and the surgical emphysema had spread downwards to the groins and into the upper parts

of the thighs. In the region of the lower ribs large pockets of air were palpable under the skin.

On the fifth day after admission exudation into the vesicles was maximal, with dehydration and circulatory collapse. The temperature rose to 104° F. (40° C.), and the pulse was frequent and weak. Intravenous plasma was given (3 pints—1.7 litres) as it was thought that protein loss in the exudate was high. The plasma was followed by glucose-saline. Intravenous therapy was continued for two days, after which the patient's condition began to improve. On the sixth day, when the mouth lesions were clearing, oral chlortetracycline was given (250 mg.) six-hourly, in place of the penicillin and streptomycin. By the tenth day the rash had begun to fade and the raw areas were drying. There was heavy desquamation of the hands and feet. By this time the conjunctivae were nearly normal, and there were no corneal lesions.

The chest was slow in clearing. Three weeks after admission the patient was still raising much thick sputum, although by then the pyrexia had subsided. From the third week onwards recovery was uninterrupted. The patient was discharged after five weeks, the only sequelae being some staining from the rash and loosening of all finger- and toenails.

Chest X-ray Examination.—Films were taken on the third, seventh, and twelfth days after admission. In the first film pronounced surgical emphysema was seen in the soft tissues of the neck and thoracic wall, but the lungs were clear. On each side of the superior mediastinum thin vertical lines of opacity were visible. These lines appeared to represent lateral displacement of the mediastinal pleura, suggesting the presence of mediastinal emphysema. The second film showed patchy shadowing of the lung fields, and the mediastinal appearances were the same. The final film showed some left basal collapse, the surgical emphysema was less marked, and the mediastinal pleura was no longer displaced.

Comment

This case represents a severe form of the Stevens-Johnson syndrome, in which skin, mucous membranes, and respiratory systems were all heavily affected. In addition there was severe toxæmia. The chief point of interest was the surgical emphysema which occurred early in the disease and which persisted for nearly three weeks. The likely explanation for the surgical emphysema seems to be as follows. In respiratory infections, such as pertussis, influenza, or measles, and in bronchial asthma, valvular obstruction of terminal bronchioles may be produced by mucous plugs or by inflammatory oedema of the lining mucosa. Alveolar air trapped behind the obstruction may be forced into the interstitial spaces of the lung by expiratory pressure. The air may then pass along the vascular sheaths (Aisner and Franco, 1949), to reach the hilum and thence the mediastinum. Air may then escape from the mediastinum into the tissues of the neck.

Similar cases of mediastinal emphysema have been described in association with the muscular straining of parturition (MacRae, 1949) and the lifting of heavy weights. Here the expiratory muscles are forcibly contracted with the glottis closed, causing rupture of alveoli.

In the present case it seems probable that the obstruction was caused by thick mucus or by the swelling of the mucosal lining of the small air passages.

The area of collapse seen in the third x-ray film may be significant. Possibly it marks the site at which a number of bronchioles became obstructed, causing the air in the alveoli distal to the obstruction to be forced into the vascular sheaths of the lung.

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