Anesthesia in Stevens–Johnson Syndrome: Report of a Case

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The Stevens–Johnson syndrome is an uncommon acute eruptive disorder of skin and mucous membranes with systemic manifestations of variable severity.[1] The incidence is unknown and, although any age group may be affected, it occurs more often in children than in adults. The occurrence of this syndrome concurrently with indications for surgery necessitating general anesthesia is even more unusual, and presents several anesthetic hazards similar to those attendant upon hereditary epidermolysis bullosa and hereditary angioneurotic edema.[2–4] Although no current description of anesthetic care of patients with this syndrome is available in the literature, it is interesting that one of the original patients of Stevens and Johnson underwent surgery, probably with general anesthesia. The anesthetic management of our patient with this syndrome who needed emergency surgery and general anesthesia is described.

REPORT OF A CASE

A 14-month-old Caucasian girl was first seen because of otitis media, which was treated with benzathine penicillin and decongestants. A maculopapular erythematous rash then appeared, increasing in severity during a two-week period. The patient became febrile (104 F), with generalized desquamation of skin, lesions of oral mucosal surfaces, cracked and scaling lips, nasal crusts, conjunctivitis, and lid edema. Steroid and antihistamine therapy was begun. Food was refused, and intravenous feeding via surgical cannulation of peripheral veins was necessary. The child's cry became hoarse, and rales were present over both sides of the chest. A roentgenogram showed patchy areas of infiltrate in the central portions of both lung fields. Hemoglobin decreased to 6.2 g/100 ml, and transfusion was necessary. Bullae developed on the chest and shoulders, the four cutdown sites were grossly infected, and blood culture revealed *Staphylococcus aureus*. Generalized edema appeared, with low serum proteins. Gastrointestinal bleeding then developed, and steroid dosage was decreased gradually. A week later, the edema had decreased significantly, and the scalp in the occipital area became fluctuant, with serosanguineous drainage from a 4-by-3-cm ulcerated area exposing the cranium. Extensive undermining radiated 3 cm from the edges of the ulcer. It became apparent that immediate surgical debridement was necessary for the patient's survival. The patient's skin was "doughy" and thin, with some desquamation and residual evidence of oral lesions still present, and the four cutdown sites had not yet healed.

General anesthesia was necessary. Ketamine hydrochloride was selected as the sole anesthetic agent, with spontaneous ventilation with room air. An intramuscular induction dose of 8 mg/kg was given with the patient in the lateral position. An 18-gauge plastic catheter was then inserted percutaneously into a preauricular vein and secured, and a vinyl blood-pressure cuff was applied to an area free of desquamated epithelium. Spontaneous respiration was maintained without a mechanical airway or intraoral manipulation. The heart beat was monitored with a chest stethoscope. Anesthesia was maintained with ketamine, administered slowly intravenously as needed. During the hour of the operation, 125 ml of blood were replaced. Blood pressures ranged from 100 to 120 mm Hg, and pulse rates ranged from 60 to 140/min. The patient tolerated the procedure well, and emergence from anesthesia was uneventful.

Three further debridements were similarly managed with ketamine and spontaneous respiration of room air. Six weeks after the first procedure, a fifth ketamine anesthetic was given. Split-thickness skin was taken from the patient's back for subsequent grafting. The patient accepted the skin grafts, continued to gain weight, and was dismissed three months after admission, with complete resolution of the Stevens–Johnson syndrome.

DISCUSSION

The many manifestations of the Stevens–Johnson syndrome have been reviewed.[5,6] Those of particular anesthetic interest are briefly considered.

Temperature elevations to 104 F are common. Cutaneous lesions progress rapidly from

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erythematous areas to macules, papules, vesicles, and bullae, followed by scaling and sloughing of epithelium, with large areas being left denuded. These areas weep fluid and protein until they are re-epithelialized. Mucous membranes of the entire body may become similarly involved, particularly in the respiratory tract. Ulcers occur in the nose, oropharynx, larynx, trachea, and bronchi, and thick purulent secretions are produced.[7, 8] The visceral pleura contains large surface bullae appearing simultaneously with cutaneous bullae, and pleural effusions have been reported. A case in which the Stevens–Johnson syndrome progressed rapidly has been reported. In this case, the visceral pleural bullae ruptured, causing bronchiolar-alveolar-pleural fistulae and subsequent bilateral pneumothoraces with subcutaneous emphysema.[9] Pneumonitis commonly occurs, with roentgenographic and clinical features similar to those of primary atypical pneumonia.[10] Acute, transitory atrial fibrillation may occur, while acute interstitial myocarditis has been found at autopsy.[11] Ocular involvement ranges from mild conjunctivitis to panophthalmitis with corneal destruction.[12] An acute inflammatory renal lesion may occur, with albuminuria, casts, microscopic hematuria, and uremia.[13] Urethritis is common. Ulcerative lesions of the pharynx, esophagus, and colon have caused hematemesis, perforation, severe rectal bleeding, and death.[14] Painful changes in buccal mucosa, pharynx, and esophagus prevent the patient from taking adequate nutrition orally.

The syndrome is precipitated by single or multiple factors. Bacterial and viral infectious agents, drugs, foods, vaccinations, deep roentgen rays, and malignancies have been implicated.[5, 6] There is no specific therapy. Steroids and antihistamines have been advocated, but results are inconsistent. Antibiotic therapy is usually necessary because of the severe infection. The prognosis is guarded. The syndrome may recur after apparent resolution. Mortality rates vary from 3 to 12 per cent. Pulmonary complications and severe infections are the most common causes of death.[15, 16]

Basic information important to the safe conduct of anesthesia may be difficult to obtain. Applications of a blood-pressure cuff to denuded or desquamated skin may be painful and may cause further epithelial damage. To avoid aggravation of existing lesions, an esophageal stethoscope and indwelling thermistor probe should not be used. However, a weighted chest stethoscope may be used. The occurrence of atrial fibrillation, myocarditis, and pericarditis in the syndrome warrants the use of an oscilloscopic ECG tracing. Because febrile episodes are part of the syndrome, body temperature must be monitored continuously and a cooling blanket used. The axilla is a convenient and atraumatic monitoring site.

Long-term intravenous support is necessary when the disease is severe. Percutaneous venous puncture is desirable when possible, because cutdown sites become infected and heal poorly. Bladder catheterization to measure urinary output should be avoided because of the frequent occurrence of urethritis. Anemia secondary to infection and poor nutrition is common. Edema may occur secondary to hypoproteinemia. Consequently, intravascular volume, degree of extracellular hydration, and electrolyte balance may vary considerably. The severity of gastrointestinal involvement, fever, and protein and fluid loss from weeping epithelium must all influence individual adjustments of fluid therapy.

Because of lesions of the oral, laryngeal, and tracheal mucosa, manipulation of or trauma to these areas must be avoided. Simple application of an anesthetic mask can abrade and denude involved facial epithelium. Soft oral airways may cause bleeding and push tissue debris into the pharynx or larynx. Endotracheal intubation is particularly hazardous in these patients. Trauma to the laryngeal and tracheal mucosa by the tube may cause abrasions and bleeding in ulcerated areas or cause bulla formation, with severe compromise of the airway after extubation.

The occurrence of pleural blebs and pneumothorax suggests that positive-pressure ventilation should not be used. The frequent occurrence of pneumonitis with intrapulmonary shunting may necessitate high concentrations of oxygen to maintain adequate arterial oxygen tension. The frequent presence of infection by *Mycoplasma pneumoniae* and other severe pulmonary infections necessitates careful decontamination of any anesthetic apparatus.[17, 18]

With regard to the choice of anesthetic technique, local infiltration may be used with the usual caution applied in other seriously ill patients, if appropriate for the surgical procedure. Regional anesthesia also may be considered, depending on the condition of the skin overlying the area of entry and the presence of systemic infection. However, the meninges are epithelial surfaces, and death from Stevens–Johnson syndrome secondary to meningoencephalitis has been reported.[14] The administration of general anesthesia presents serious problems. The delivery of inhalation agents involves some instrumentation or manipulation of the airway, and even the use of a mask and an oral airway can lead to difficulty. Also, intravenous agents generally cause respiratory depression, loss of airway control, and cardiovascular depression. Use of ketamine, the only agent used in our case, can avoid these effects; fortunately, the scalp lesion with its somatic innervation permitted the use of this drug. In patients who need obtundation of visceral pain, a different technique may be advisable. In such instances, the anesthesiologist must compromise the adequacy and safety of anesthesia with the risks inherent in the Stevens–Johnson syndrome.

Summary

The Stevens–Johnson syndrome is an uncommon eruptive disorder of skin and mucous membranes. Involvement of the entire respiratory tract and pleura makes the administration of general anesthesia to patients with this syndrome particularly hazardous. A 14-month-old infant undergoing emergency surgery was managed with ketamine and spontaneous ventilation with room air without instrumentation of the airway.

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