Sclerema Neonatorum
Complicating Surgical Procedures

In the past year, 3 infants from a single hospital service developed a condition following the description of sclerema neonatorum. Sclerema appeared to be a major factor in the deaths of two of the infants. Because so little attention has been paid to this process in surgical literature, it seems pertinent to report the experience with these 3 patients and to review the more recent literature.

Report of Cases

Case 1—A white male infant was brought to the Hospital at 3 days of age. Details of birth and past history were not available. Physical examination on admission revealed a 4 lb 6½ oz. (2 kg.) infant with a rectal temperature of 95 F. The baby was cyanotic and moderately dehydrated and appeared acutely ill. There was obvious respiratory distress, with coarse rales and rhonchi audible throughout the chest. Subcutaneous tissue over the buttocks and lower extremities was cold, firm, and fixed. The lower extremities were immobile, and there was no pitting. Laboratory data were within normal limits, except for a 4+ protenuria. Roentgenograms after injection of a 40% iodine addition product of poppyseed oil (Lipiodol) into the esophagus revealed atresia of the esophagus at T1. The gastrointestinal tract was distended with air. The child was first given hydrocortisone sodium succinate, intravenous fluids, and oxygen, and attempts were made to control body temperature. Despite these measures, hardening of the subcutaneous tissue spread in a cephalad direction and respiratory distress increased. The child died 19 hours after admission and before operation was attempted. Permission for autopsy was not granted.

Case 2—A 3-day-old white female infant was admitted from another hospital. Shortly after birth frequent suctioning was required to remove orals, and respiratory distress developed. The patient had received fluids by clysis before admission. Physical examination revealed a 6 lb 11½ oz. (3.05 kg.) infant whose rectal temperature was 97.2 F. The child was well developed and well nourished. There was obvious respiratory distress. No other abnormalities were noted at the time of admission. Laboratory data were within normal limits. Roentgenograms after injection of Lipiodol into the esophagus revealed atresia at the level of C7, with an increased amount of air in the gastrointestinal tract. The patient was initially given intravenous fluids and antibiotics. Seven hours after the patient's admission, esophageal atresia with tracheoesophageal fistula (gloss, Type C) was repaired through a right thoracotomy. Twelve hours after operation, intermittent periods of cyanosis were noted, and respiratory distress became progressive. Temperature remained below 98 F, reaching 96.4 at the lowest point. Seventy hours after operation, diffuse firmness of the subcutaneous tissue over the buttocks, thighs, back, and chest were noted, and respiratory distress increased. There was marked immobility of the chest wall, and despite the institution of hydrocortisone sodium succinate therapy, the patient died 78 hours after operation. At autopsy the anastomosis was intact; there were multiple abnormalities of the heart, and routine microscopic examination of the skin and subcutaneous tissue revealed no recognized pathologic changes.

Case 3—A white female child was delivered at the Medical Center after an uncomplicated gestation. Birth weight was 5 lb 7 oz. (2.47 kg.). Her respiratory distress was noted shortly after her arrival in the Newborn Nursery. Lipiodol injection

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into the esophagus was carried out when the patient was 36 hours of age, revealing esophageal atresia at T5, with increased air in the gastrointestinal tract. Fifty-two hours after birth, closure of the tracheoesophageal fistula with esophageos- sophagostomy was accomplished through a right thoracotomy. Postoperatively, the patient was given antibiotics and put in an incubator. Seven hours after operation, firmness of the subcutaneous tissue about the buttocks, thighs, and legs was noted. Shortly after this discovery, hydrocortisone sodium succinate, 25 mg. intramuscularly every 6 hours, was started, and increased efforts to maintain body temperature were instituted. Forty-eight hours after the onset of treatment there was noted softening of the subcutaneous tissue, and hydrocortisone sodium succinate was reduced to 15 mg. intramuscularly every eight hours. By the seventh postoperative day, all evidence of sclerema had disappeared and steroids were slowly withdrawn. The child was discharged 19 days after operation.

Comment

Sclerema neonatorum can be defined as a process occurring in the newborn infant, characterized by nonpitting induration of the subcutaneous tissue, usually beginning in the region of the thighs and buttocks and spreading in an ascending manner. The skin over the involved areas seems bound to the underlying structures and assumes a cool, smooth, purple appearance. Progression of the process to involve the skin of the chest with progressive respiratory distress is the usual terminal event.

Hughes and Hammond reviewed the literature pertinent to sclerema neonatorum up to 1948. Adding 3 cases of their own, Hughes and Hammond were able to collect 28 cases from the literature prior to 1948, and summarized them as follows:

1. The average age of onset was 4 days with extremes from birth to seventy days.
2. Twenty-five per cent of the mothers were ill at the time of delivery.
3. All but 2 deliveries were spontaneous.
4. Average birth weight was 2,800 gm., with variations from 2,150 to 4,100 gm.
5. The majority of the infants exhibited abnormal behavior and symptoms at birth; weakness and cyanosis were the most common.
6. Almost all had difficulty with body temperature control and evidences of other complications besides sclerema.
7. Seventy-five per cent died, the average age at death being 10 days.

Histopathologic examination of tissue involved in sclerema obtained both at biopsy and at autopsy shows no characteristic change when studied by the usual methods, though some observers have reported an increase in collagenous tissue. The process is to be differentiated from subcutaneous fat necrosis which has a characteristic microscopic picture.

Though the etiology of sclerema neonatorum is unknown, a number of possible factors have been enumerated. Some investigators have reported a difference in the composition of subcutaneous fat between infants and adults, and others have found that this difference is more marked in infants with sclerema. Because this difference involves changes in solidification with temperature changes, this theory has been popular, but at the present time it is considered to be of questionable importance. Hughes and Hammond suggested that sclerema neonatorum may be a manifestation of severe shock in infancy and that the gross changes observed may be due to insufficient peripheral circulation.

A number of therapeutic agents have been used in the treatment of sclerema. Among these are included thyroid extract, x-irradiation, hot baths, and blood transfusions. No effective treatment was available until the advent of corticosteroids. In 1951, Kendig and Toone reported a patient unsuccessfully treated with cortisone. In the following years, several subsequent reports of successful treatment with corticotropin and cortisone have been published, though the mortality rate remains high. The value of general supportive therapy, antibiotics, attention to fluid and electrolyte balance, and maintenance of body temperature have been stressed by most authors. In addition, it is felt that the intravenous route of administration is preferred because peripheral circulation may impair the subcutaneous or muscular absorption. At this time there is little agreement regarding either the advisability of the dosage to be given. No conclusion is possible on the clinical picture without considering coincident factors, which less attenuates the mortality rate.

The occurrence of sclerema neonatorum in infants with normal clinical picture is rare by others. The fact that it is often associated with central nervous system and respiratory tract involvement suggests an underlying etiology such as anemia, hypothermia, infection, and metabolic disturbance. The history of the infant prior to the onset of sclerema should be reviewed carefully.

The occurrence of sclerema neonatorum in a single year at the University of Oklahoma Medical School is significant. This report is of interest because of the unusual occurrence of 3 cases in 1 year and the relatively good out-
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