SCLEREMA ADIPOSUM NEONATORUM

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SCLEREMA adiposum neonatorum is a disease of the first few weeks of life, characterized by widespread, patchy hardening of the subcutaneous tissue, the etiology of which is unknown. Infants suffering from this illness are usually premature, exhibit subnormal temperature, apathy, poor feeding and inability to gain weight resulting in rapid malnutrition and dehydration, and terminating in death within a few weeks.

Pathologically, this disease is characterized by a foreign-body granuloma showing wide fibrous branching trabeculae, neutral fat crystals, numerous proliferating blood vessels, and infiltration by foreign-body giant cells, lymphocytes, and an occasional polymorphonuclear leucocyte.

Since this entity is uncommon and because of close observation of a patient with a prolonged clinical course, including post-mortem examination, we are reporting the following case.

CASE REPORT

History.—A white male infant was born Sept. 11, 1946, to a 35-year-old mother who had a history of eight previous pregnancies including one abortion at the third month of gestation. The present pregnancy was full term and complicated by mild pre-eclampsia. Birth was spontaneous and the amniotic fluid was meconium-tinged. The mother's Wassermann was negative.

Clinical Examination.—The infant weighed 11 lbs., 8 ozs., and measured 21.5 inches. Scalp ecchymosis and moderate signs of moulding with overriding of the parietal bones, as well as marked caput at the vertex were present. The anterior fontanelle was not bulging. Supra- and infrasternal retraction were noted. Breath sounds were vesicular and transmitted throughout; heart tones were of good quality without murmurs; cry and color were fair. Sixteen hours after birth, clonic seizures of facial and arm muscles were present. Vitamin K and calcium gluconate therapy were administered.

On the second day, facial tremors and moderate spasticity of all extremities with marked flexion of the left wrist were noted. Cyanosis and regurgitation were prevalent, but responded to oxygen inhalation and gavage. A 16 oz. weight loss was present since birth.

During the next few days, several new features developed, such as cephalic cry, thrush, and a hard, subcutaneous mass in the left arm. A clysis of Lactate-Ringer's solution was ordered and the monilia infection cleared rapidly with 1 per cent aqueous gentian violet applied topically. A scalp infection followed, with several areas of oozing. A dry gangrenous slough developed in the occipital area, despite 10,000 units of penicillin every three hours intramuscularly and a 60 c.c. transfusion of whole blood. A low-grade fever and cyanosis persisted, but feeding definitely improved.

On September 22, several firm, indurated, subcutaneous areas were noted on the cheeks, back, volar surfaces of arms, calves, and abdomen. The flexion deformity of the left wrist had decreased since application of splints and the scalp was healing rapidly. A blood count showed 20,000 white blood cells

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per cubic millimeter, polymorphonuclears 70 per cent, lymphocytes 30 per cent, hemoglobin 14.5 Gm., and 4.8 million red blood cells.

X-ray examination at this time revealed moderate separation of all intracranial sutures, particularly in the basiocciput with no evidence of bony involvement of the upper extremities. No pulmonary pathology was evident.

A subdural tap was performed bilaterally but bloody fluid was not obtained. A spinal tap between the second and third lumbar vertebrae was unsuccessful. A cisternal puncture revealed clear, colorless, spinal fluid containing 5 lymphocytes per cubic millimeter, protein 74 mg. per cent, sugar 65 mg. per cent, with negative serology and colloidal gold, and which was sterile on culture.

Orthopedic consultation noted a "spastic contraction of all fingers of the left hand, with thumb markedly adducted and the wrist markedly flexed. The right hand is in flexion but extension of the fingers and wrist does occur.

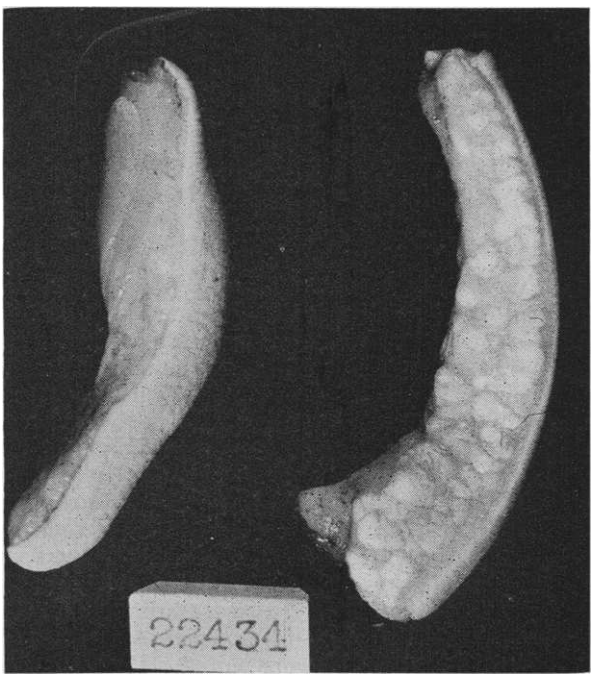

Fig. 1.—Section through entire thickness of skin and subcutaneous tissue. Note "nodular" pattern.

Diagnosis.—Cerebrospastic with flexion contracture of both wrists, more marked on left.

On October 22, a biopsy of one of the indurated areas on the calf was performed. The specimen consisted of skin and subcutaneous tissue which was moderately firm and white. The microscopic section of skin and subcutaneous tissue showed intact epidermis with normal morphology. The subcutaneous fat layer was traversed by thick, fibrous, collagenous trabeculae and areas of looser connective tissue, throughout which were numerous foreign-body giant cells containing from 5 to 30 nuclei. The diagnosis was sclerema adiposum neonatorum.

The infant began to vomit again and rapidly developed signs of dehydration. A pinhead-sized draining sinus was noted over the lesion of the left forearm. Subcutaneous clyses were administered. At this time the hemoglobin was 12.5

Gm. per cent, 4.2 million red blood cells, hematocrit 36.5 per cent and plasma proteins 5.5 gm. per cent. Urinalysis revealed an acid reaction, albumin one plus, sugar negative, microscopic negative, and Sulkowitch test was positive for calcium.

Since vomiting persisted despite formula changes consisting of Sobee and thickened cereal feedings with and without atropine, a gastrointestinal series seemed advisable. The x-ray report follows:

"Fractional study of the gastrointestinal tract within a period of five hours shows no intrinsic lesion visible in the stomach or duodenum. A slight delay of gastric evacuation is noted within three hours. In five hours most of the barium is distributed in the lower portion of the small intestine with a small gastric residue."

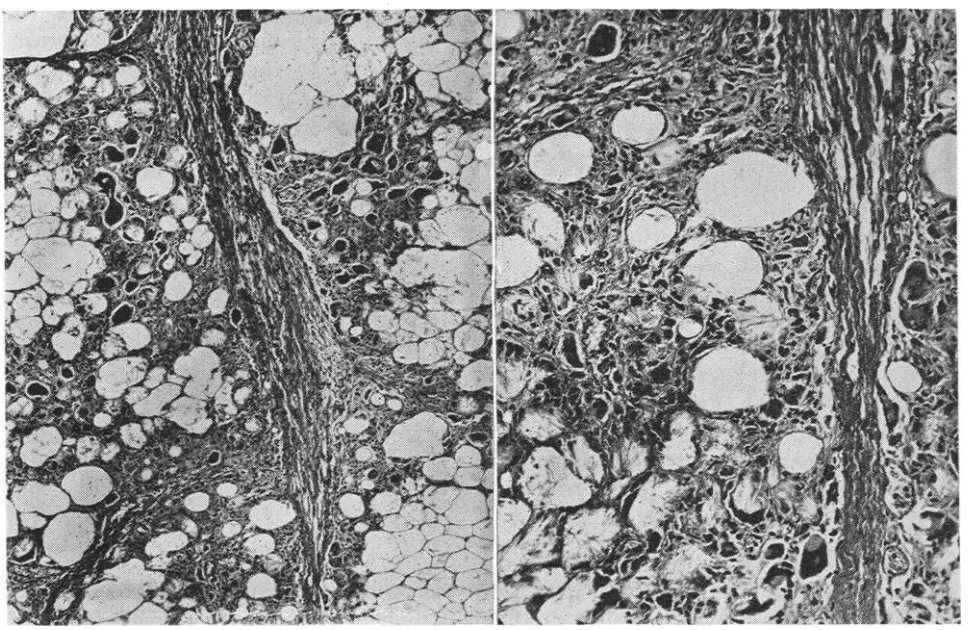

Fig. 2. Fig. 3.

Fig. 2.—Section through skin and subcutaneous tissue; stained with H. & E. (×100). Note thickening and branching of fibrous trabeculae and numerous foreign-body giant cells.

Fig. 3.—Same section as Fig. 2 with higher magnification (×440).

Vomiting and dehydration continued despite subcutaneous clyses and small blood and plasma transfusions. Diarrhea ensued, for which oral feedings were stopped and continuous intravenous fluids were given, including parenteral vitamin therapy. The diarrhea abated but a subnormal temperature occurred and was followed by coma and generalized fine inspiratory rales. A blood CO_2 combining power was 30 volumes per cent and urea 69 mg. per cent. The baby expired Nov. 14, 1946, sixty-four days following birth.

NECROPSY

At necropsy, twenty-one hours after death, the body was that of a white, 2-month-old male infant, measuring 22 inches and weighing approximately 12 lb. Several large, firm, subcutaneous, fibrotic patches were palpated, which were most marked on both cheeks, buttocks, thighs, right lower deltoid, flexion

surfaces of the forearms, and both calves. The thyroid was not palpable. The abdomen was moderately distended. Both testes were descended. The adipose tissue measured 3 mm. over the thorax and 6 mm. over the abdomen. The superficial layer was firm and divided into numerous distinct grayish brown lobules beneath which a deeper layer of white, gelatinous, transparent, glistening fat was noted. Many cutaneous nerves were distinctly seen within this portion of skin.

The peritoneal cavity contained about 100 c.c. of clear, straw-colored fluid with a few fibrin flakes. The stomach was moderately distended; the intestines were collapsed. The liver extended 4 cm. beneath the right costal margin in the midclavicular line, 1 cm. beneath the xiphoid process, and the left border was at the left costal margin in the midclavicular line. The urinary bladder was distended. The kidneys and ureters were in their usual position. The thoracic and abdominal cavities showed nothing unusual. The thymus was small and contained a moderate amount of surrounding fat. The brain showed an area of softening measuring 1 x 1 cm. in the gyri of the right temporal area just beneath the cortex.

Histologically, the epidermis and cutis showed normal morphology. The subcutaneous fat revealed numerous branching trabeculae composed of dense collagenous connective tissue, the borders of which were lined by young fibroblasts and giant cells and contained large proliferating blood vessels, some obliterated. At the junction of the subcutis and fat, there was a thin reactive band of fibroblasts, occasional lymphocytes and polymorphonuclears as well as numerous foreign-body giant cells containing five to thirty nuclei. Special fat stain (Sudan III) showed numerous neutral fat crystal in the adipose layer. When examined with a polarizing microscope, using crossed Nicol prisms, these crystals were seen to rotate the plane of polarized light and shined brilliantly throughout the section.

Microscopic examination of all the remaining organs revealed normal histology except the liver, which showed hydropic degeneration and cloudy swelling of the parenchyma with distention of the sinusoids.

COMMENT

A review of the literature reveals a marked confusion between the terms sclerema, pseudosclerema, and scleredema, which many authors use interchangeably. This concept is erroneous since each can be differentiated clinically and pathologically.

Sclerema is characterized by:

1. Onset: within the first two weeks of life.
2. Distribution of lesions: Parts with most fat involved such as cheeks, buttocks, back and extremities (thighs, calves, and forearms). The lesions of the extremities tend to follow the long axis.
3. Palpation: discrete, nontender, nonfluctuant, nonpitting, firm, indurated lesions, with irregular borders. The skin is not movable over the lesion.
4. Color: prevailing tint of skin.
5. Course of lesions: no regression or calcification; possible ulceration.
6. Systemic effects: subnormal temperature, gastrointestinal upset, dehydration, debility, and death after several weeks or months.

Pseudosclerema, or subcutaneous fat necrosis, is usually traumatic in origin with no uniform distribution, occurring at any age, without systemic effects, and characterized locally by tenderness, an overlying, freely movable skin,

with or without discoloration or ulceration. This is a benign condition which usually resolves; occasionally calcification results.

Scleredema (infantile) or edema neonatorum, is a particular variety of edema in which the skin is very hard but exhibits pitting, and is not adherent to the underlying subcutaneous tissue. The parts involved are usually the feet, calves, outer thighs, but occasionally the genitals and palms and soles are affected. These latter sites are always spared in sclerema adiposum.

Biopsy conclusively establishes the diagnosis.

The etiology of sclerema adiposum neonatorum is unknown, although many theories have been proposed. The most widely accepted explanation is that a deficiency of unsaturated fat exists in the adipose tissue, thereby causing it to solidify at a higher temperature than is needed for normal fat. This results in a foreign-body reaction of the tissues.

Although the literature emphasizes the prevalence of this disease in premature infants with an extremely rapid course (two to three weeks); the above case, on the contrary, is that of a large baby with a protracted illness.

SUMMARY

A case of sclerema adiposum neonatorum is presented, including necropsy. The terminology, etiology and differential diagnosis is discussed. Unlike most cases, this patient was unusually large and pursued a prolonged clinical course.

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