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CASE REPORT

Sclerema Neonatorum in a Full-Term Infant Showing Favorable Prognosis

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Sclerema neonatorum (SN) is a rare disease characterized by firm, indurated, waxy skin lesions extending throughout the body, sparing the fat-free soles, palms, and genitalia. The prognosis of SN is generally very poor, with a high fatality rate. We report the case of a full-term infant with delayed onset of SN showing good prognosis. A 4-month-old Korean male infant presented with diffuse waxy, sclerotic skin lesions on the whole body, sparing the face, nipples, and genital area, which began developing at 2 months of age. Histopathologic findings of the sclerotic skin lesions showed wide, fibrous intersecting bands in the subcutaneous fat tissue. Only sparse infiltration of lymphocytes and histiocytes was observed in the fat lobules and septa. Based on clinical presentation and histopathologic findings, he was diagnosed with SN. The patient survived with conservative care and had mild improvement of the skin lesions on his follow-up visit at 12 months of age. **(Ann Dermatol 29(6) 790~793, 2017)**

-Keywords-
Infant, Prognosis, Sclerema neonatorum

INTRODUCTION

Sclerema neonatorum (SN) is an extremely rare condition of infancy. SN is classified as a type of panniculitis that manifests as hardening of the skin and subcutaneous adipose tissue. The hardened skin and subcutaneous fat become adherent to the underlying muscle and bone to such extent that it hinders feeding and respiration and usually culminates in death[1]. Herein, we report a case of SN with delayed onset and without any comorbidity, showing good prognosis. As far as we known, this is the first reported case of SN in the Korean dermatologic literature.

CASE REPORT

A 4-month-old Korean male infant presented to the outpatient dermatology clinic with diffuse sclerotic skin on the whole body, which had been developing since the age of 2 months. Vomiting after feeding and mild respiratory difficulty accompanied the skin symptoms. Waxy, indurated skin with erythema was observed on the whole body, sparing the face, both nipples, and the genital area (Fig. 1). The patient was born at 40 weeks of gestation to a healthy mother by elective cesarean section. He weighed 2,780 g and was healthy without any congenital malformation or severe illness at birth. However, the infant was admitted to the neonatal intensive care unit for 4 days because of a rotavirus infection.

A skin biopsy was performed of a sclerotic skin lesion on the trunk. Histopathologic findings showed wide, fibrous, intersecting bands in the subcutaneous fat tissues (Fig. 2A). Fibrous bands extended to the deep portion along the septa of the subcutaneous fat tissue (Fig. 2B). Only sparse lymphohistiocytic infiltration was observed along the fat lobules and septa, showing a mixed pattern of panniculitis

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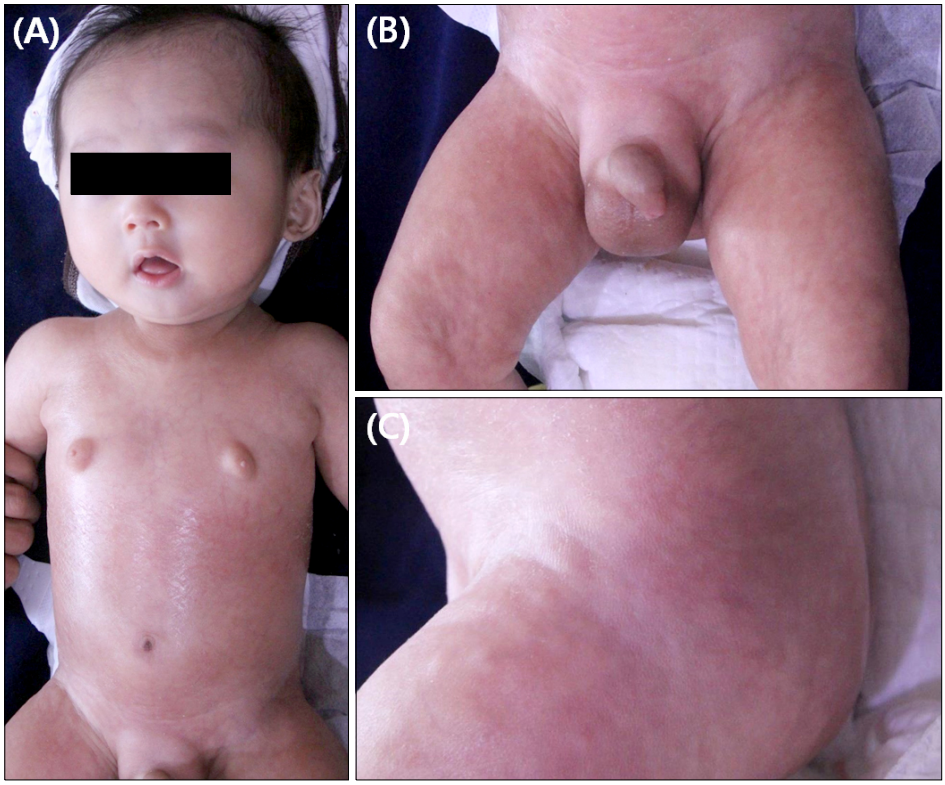

Fig. 1. Clinical presentation of the patient at 4 months of age. (A∼C) Diffuse sclerosis and subcutaneous indurations on the whole body, sparing the face, nipples, and genitalia.

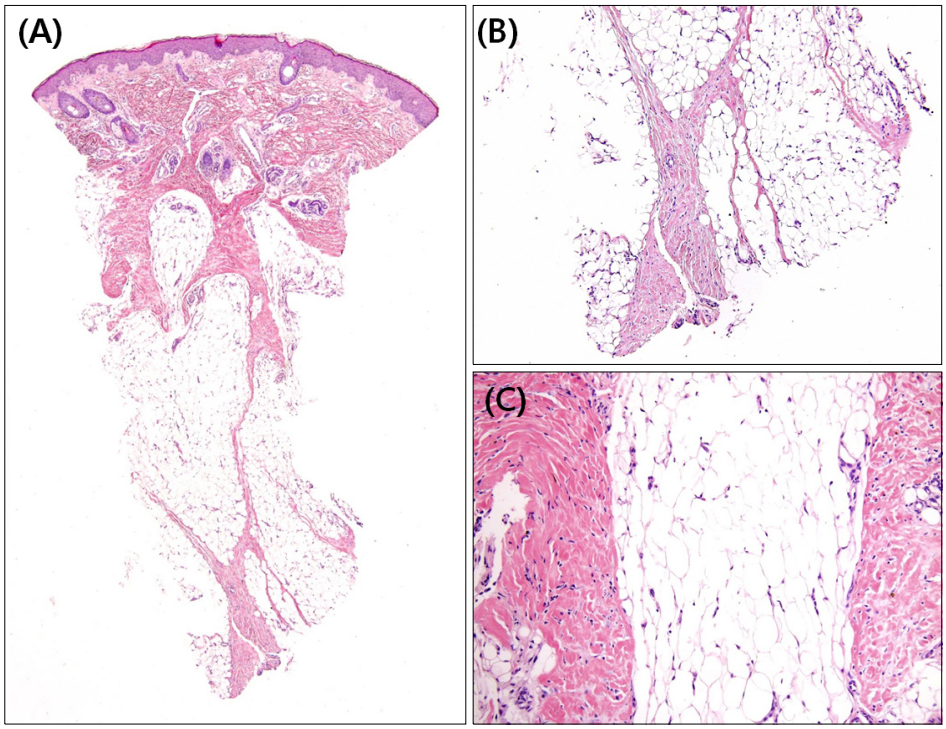

Fig. 2. Histologic findings. (A) Fibrous intersecting bands in the subcutaneous fat tissue (H&E, ×40), (B) Widened fibrous septa of subcutaneous fat tissue (H&E, ×100), (C) Sparse lymphohistiocytic infiltration along the fat lobules and septa showing a mixed pattern of panniculitis (H&E, ×200).

(Fig. 2C). The epidermis and papillary dermis were normal, and compact packing of collagen fibers was seen at the lower part of the reticular dermis. On the basis of clinical and histologic findings, SN was diagnosed.

Moisturizer was applied without any use of systemic medications. On his follow-up visit at 12 months of age, the patient showed delayed growth, calculated as 0.3 percentile of the growth curve. However, he maintained a fa-

vorable condition, with improvement in gastrointestinal and respiratory symptoms, and the sclerotic skin lesions of both lower extremities had improved mildly.

DISCUSSION

SN is an extremely rare disease in the 21st century, and it has disappeared where advanced neonatal intensive care is available. It has mostly been reported in low birthweight, premature newborns in the setting of severe illness, including infections and heart disease[2]. Affected neonates have variety of underlying conditions associated with the process, including respiratory and gastrointestinal disease, sepsis, and congenital malformations[3-5]. SN has been shown to occur primarily in preterm neonates, more often in boys than in girls, and most often within the first week of life[1].

SN is characterized by the development of firm, indurated, waxy plaques on the thighs, buttocks, and trunk. The process may involve any area of the body except the fat-free soles, palms, and genitalia[1]. Involvement rapidly extends to the whole body, rendering the infant immobile and impairing feeding and respiration[6]. The histologic features of the sclerotic skin lesions are thickening of subcutaneous tissue due to an increase in the size of the fat cells and the presence of wide, intersecting fibrous bands[7]. The trabeculae forming the framework are broadened, and the fat spaces are diminished with only sparse inflammation[2]. Fine, needle-shaped crystals (Type A crystals) may be observed to arrange radially in adipocytes[8].

The prognosis of SN is very poor, with the fatality rate reported to be as high as 98% in the report of 51 preterm neonates with SN in the first 28 days of life[9]. Previous report subjected to SN patients with septicemia presents patient distribution as follows; 88.3% of the patients (54/60) born with low birth weight (<2,500 g), 65% of the patients (39/60) born in preterm period (<37 weeks), and 85% of the patients (51/60) with predisposing factors such as prolonged rupture of membranes, maternal infection, and birth asphyxia[10].

In this case, the infant was born full term with a normal birthweight, without any congenital malformation or severe illness. The patient had a late onset of diffuse sclerosis starting 2 months after birth. SN is typically seen in the first few days after birth but may develop as late as several weeks of life[1]. Histologic features showed wide, intersecting fibrous bands throughout the fat lobules, with sparse lobular and septal lymphohistiocytic infiltration. Needle-shaped crystals in adipocytes were not observed in our case, but these are reported to be nondiagnostic[7]. The possibility of subcutaneous fat necrosis of the newborn could be ruled out by skin involvement of the whole body and intersecting fibrous septa with only sparse inflammatory infiltration of lymphocytes and histiocytes.

Our patient presented with vomiting and mild respiratory difficulty, and he was conservatively treated under consultation with the pediatric department without any use of systemic steroids or exchange transfusion. On his follow-up visit at the age of 12 months, the baby showed delayed growth; however, the sclerotic skin lesions of his lower extremities had improved mildly, and his general condition was favorable. Our case of SN is interesting because of its favorable prognosis and the late onset of presentation in a healthy full-term baby. This is the first case of SN reported in the Korean literature, and our case supports the theory that the prognosis of SN would be better in full-term infants than in premature infants[11]. The skin is expected to return to normal without long-term sequelae in surviving infants[12,13].

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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