Subcutaneous Fat Necrosis of the Newborn Presenting as a Large Plaque With Lobulated Cystic Areas

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GOAL

To understand the presentation and treatment of subcutaneous fat necrosis of the newborn (SFN)

OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Recognize the clinical presentation of SFN.
2. Explain the differential diagnosis of SFN.
3. Describe the link between SFN and hypercalcemia.

CME Test on page 174.

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Subcutaneous fat necrosis of the newborn (SFN) usually occurs in the first few weeks of life in full-term infants and presents as indurated, distinct nodules with a predilection for the cheeks, shoulders, back, buttocks, and proximal extremities. Most cases are related to some form of fetal distress, including obstetric trauma. Some cases are associated with hypercalcemia. We report a case of SFN with an unusual clinical presentation, complicated by hypercalcemia.

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Case Report

A 6-week-old healthy Hispanic girl presented to our clinic with a 10×11-cm irregular, diamond-shaped, fluid-filled, slightly erythematous cystic plaque over the central portion of her back. The infant was born after a difficult vaginal delivery, and the perinatal period was complicated by meconium aspiration and Erb's palsy of the left upper extremity. She was otherwise healthy, had a good appetite, and was discharged from the hospital at the usual time. When the infant was 4 days old, the mother noted an asymptomatic reddish purple discoloration on her back.

Results from a total body skin examination revealed a 10×11-cm slightly violaceous, lobulated cystic plaque, with several easily compressible cystic dilations filled with fluid over the central

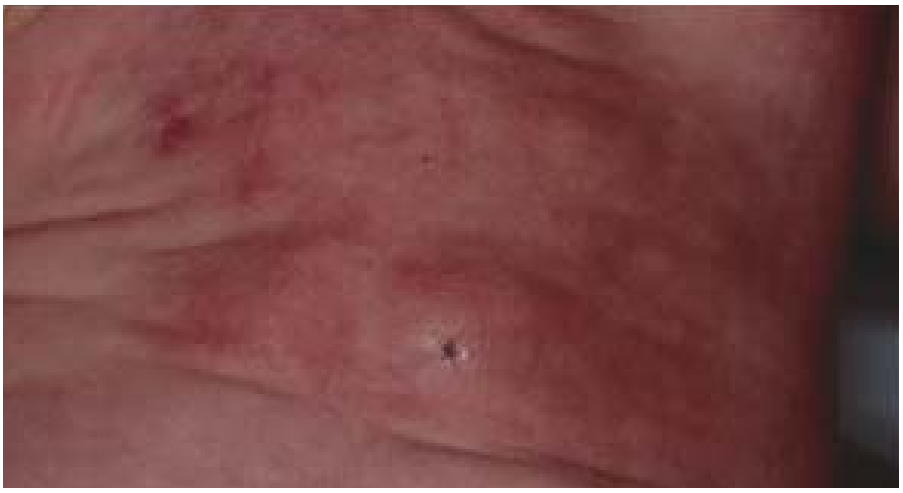

Figure 1. A large erythematous, lobulated cystic plaque involving the center of the back.

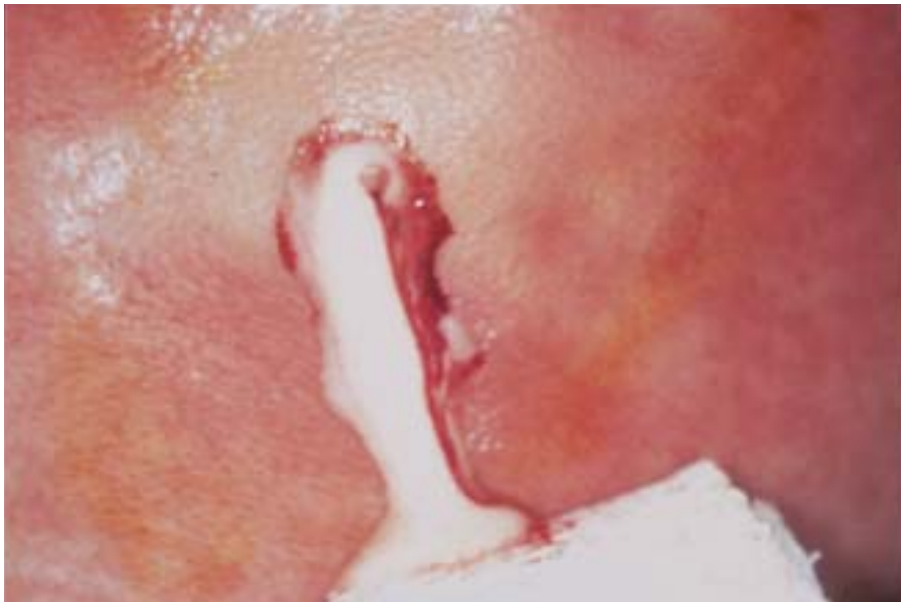

Figure 2. Drainage of exuberant, thick white fluid from the biopsy site.

portion of the back (Figure 1). The findings from cardiovascular and neurologic examinations were normal. The left upper extremity was held in adducted position, with the arm flexed and the fingers held in a fisted position secondary to birth trauma. There were no abdominal masses or hepatosplenomegaly.

The clinical impression was a vascular malformation, possibly lymphatic or mixed lymphatic and venous. Performance of a 4-mm punch biopsy resulted in the drainage of a thick white fluid from the biopsy site (Figure 2). Results of a histologic examination revealed granulomatous inflammation with numerous histiocytes and multinucleated giant cells in the subcutaneous fat (Figure 3). Radially arranged, needle-shaped clefts were seen within multinucleated giant cells (Figure 4). Five days after the initial biopsy, several milliliters of a thick, chalky white material were drained from the lesion via an incision made with a 2-mm punch. The affected areas on the back healed slowly and left residual mild hyperpigmentation and lipoatrophic scarring.

Starting at the age of 6 weeks, because of the potential risk of late-onset hypercalcemia, the levels of serum calcium and phosphorus were evaluated weekly and were elevated at 10.7 mg/dL (reference range, 8.8–10.2 mg/dL) and 6.0 mg/dL (reference range, 3.1–4.5 mg/dL), respectively. The level of parathyroid hormone was 8 pg/mL (reference range, 12–72 pg/mL). The calcium levels slowly

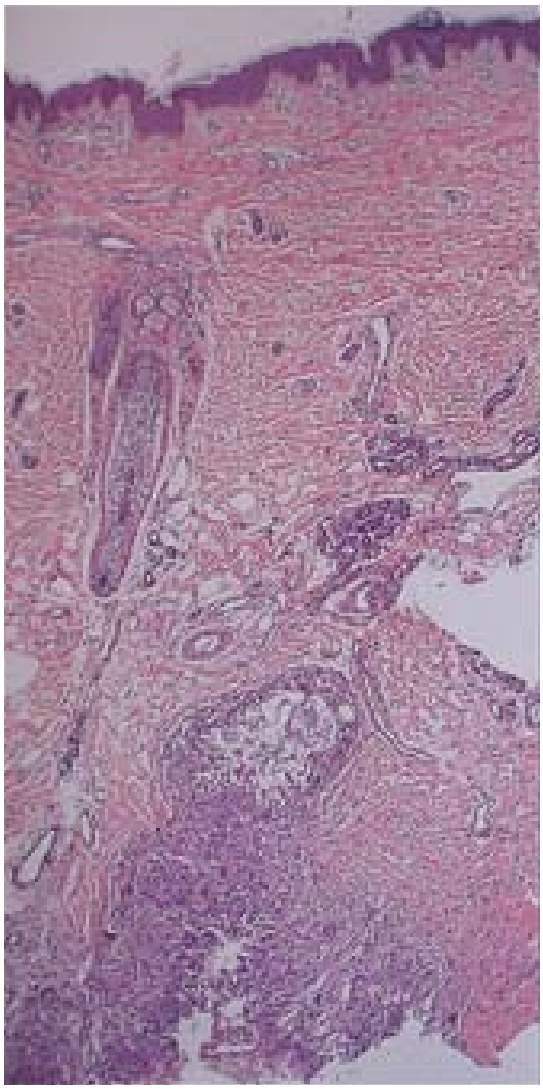

Figure 3. An area of granulomatous inflammation in the deep dermis and subcutaneous fat (H&E, original magnification ×4).

increased until, at age 11 weeks, the calcium peaked at 11.1 mg/dL and the phosphorus at 7.6 mg/dL. Even though the infant did not exhibit signs or symptoms of hypercalcemia, persistent and increasing levels of serum calcium became a concern, and she was given a 5-day course of oral prednisolone (1 mg/kg per day). One week later, repeat serum calcium and phosphorus levels were 11.0 mg/dL and 6.5 mg/dL, respectively. Without further intervention, levels of serum calcium and phosphorus continued to decrease over the ensuing month.

Comment

Subcutaneous fat necrosis of the newborn (SFN) was first described clinically by Cruse[1] in 1875 and initially was called *scleroderma of the newborn infant*. SFN is usually a self-limited disorder of unknown etiology, but if hypercalcemia is associated, the mortality rate is 15%.[2,3] Most infants are born at full term, but many have a history of prenatal or perinatal complications.[3] Suggested etiologies include fetal factors (eg, a primary defect in subcutaneous fat, birth hypoxia, local trauma, hypothermia) and maternal factors (eg, gestational diabetes, preeclampsia, maternal exposure to cocaine or calcium antagonists).[2,4] The most frequently recognized possible etiologic factors in a series of 11 cases were birth asphyxia and meconium aspiration.[4]

Clinically, SFN presents as erythematous to violaceous, firm, well-circumscribed, subcutaneous nodules that usually appear shortly after birth but sometimes within weeks.[2] The most commonly involved sites are those with the thickest fat deposition: buttocks, thighs, trunk, shoulders, face, and arms.[2] The lesions usually resolve spontaneously without scarring over the following few months.[2,5] Rarely, however, they may become fluctuant and soft, discharge oily material, and be associated with scar formation.[5] In our case, copious chalky white liquid was drained from the fluctuant nodules. Hypercalcemia may develop, but the subcutaneous nodules usually precede it by weeks or months. In a series of 11 cases of SFN, 28% showed hypercalcemia.[4] Most neonates with SFN and hypercalcemia show failure to thrive, but some die because of sequelae of hypercalcemia.

Histologically, abundant histiocytes and foreign body giant cells with needle-shaped clefts are seen in the panniculus.[2] Adipocytes at different stages of degeneration are present. In addition, calcific deposits may occur within the areas of fat necrosis.[2]

The differential diagnosis for SFN includes sclerema neonatorum (SN); poststeroid panniculitis; and, at least in our patient, a vascular malformation. SN usually occurs in premature, undernourished infants, often suffering from a serious illness, and affects the skin in a diffuse fashion, as opposed to a more localized area in SFN.[6,7] SN appears shortly after birth, within the first week of life, usually following an uncomplicated delivery.[2] The skin becomes hard with a waxy appearance on the lower legs that progresses to the trunk, upper limbs, and face.[2] The palms, soles, and genital area are spared.[2] The outcome usually is fatal, with a mortality rate of 75%, often due to sepsis.[2] Histologically in SN, expanded fat lobules and thickened fibrous septae are present.[2] Lipocytes contain the characteristic needle-shaped clefts in a radial or starburst pattern.[7] Unlike in SFN, there is little or no associated inflammation.[2,5]

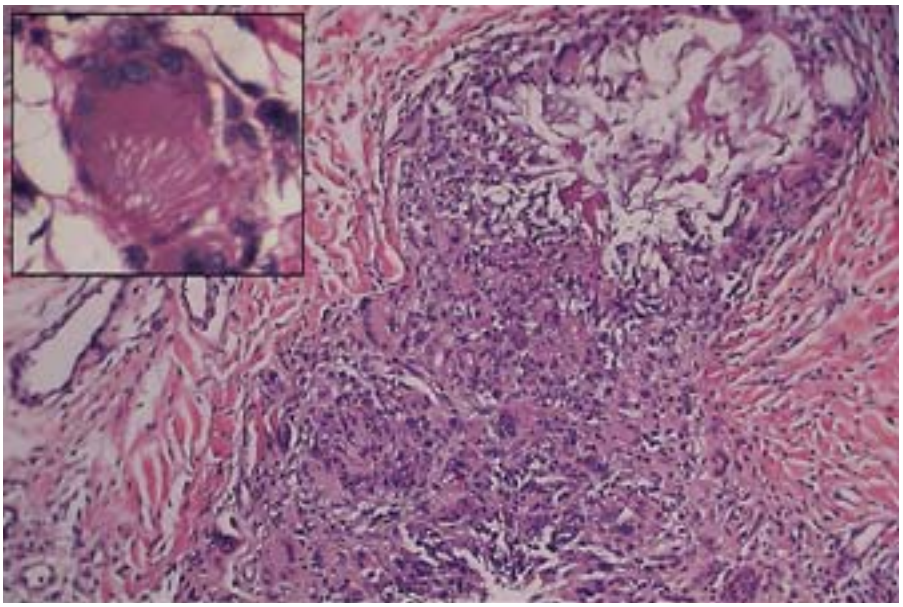

Figure 4. Higher magnification reveals granulomatous inflammation with numerous histiocytes and multinucleated giant cells in the subcutaneous fat (H&E, original magnification ×10). Inset: radially arranged, needle-shaped clefts within a multinucleated giant cell (H&E, original magnification ×40).

Poststeroid panniculitis is an extremely rare form of panniculitis that is seen in young children days after the rapid discontinuation of high doses of oral corticosteroids.[8] Painful or pruritic subcutaneous nodules develop on the face (especially cheeks), trunk, and arms, and resolve without scarring over a period of weeks to months.[8] Histologic examination reveals needle-shaped clefts occurring within both macrophages and adipocytes, similar to SFN.[8]

An association between SFN and hypercalcemia was first reported in 1956.[9] Most neonates with SFN show signs and symptoms of hypercalcemia, with failure to thrive as the most frequent clinical feature.[2] Irritability, fever, vomiting, hypotonia, seizures, and death may follow.[2] The cause of hypercalcemia in infants with SFN is unknown. Potential causes include increased prostaglandin activity, parathyroid hormone levels, vitamin D sensitivity, and calcium release from necrotic adipose tissue.[2,10-12] The most likely explanation is unregulated production of 1,25-dihydroxyvitamin D by macrophages participating in the granulomatous inflammatory process.[12] Hypercalcemia may be caused by increased intestinal absorption of calcium caused by elevated levels of 1,25-dihydroxyvitamin D.[13]

Treatment of patients with hypercalcemia may require vitamin D restriction, low-calcium diet, adequate hydration, and a calcium-wasting diuretic such as furosemide.[11,14] Oral prednisone effectively lowers serum calcium levels by interfering with metabolism of vitamin D to the active form 1,25-dihydroxyvitamin D.[2] Prednisone also may inhibit production of this metabolite by macrophages involved in the granulomatous inflammatory process.[2]

For patients with SFN and no hypercalcemia, periodic monitoring for signs and symptoms of hypercalcemia and periodic testing of calcium levels are indicated.[2] Because hypercalcemia can occur 1 to 4 months after the onset of skin lesions, it is important to educate the child's parents to recognize the signs and symptoms of hypercalcemia and report them promptly to their clinician.[2]

Unusual clinical presentations of SFN have been described. Hernandez-Martin et al[15] described congenital SFN with ulceration. Varan et al[16] described a neonate presenting immediately after birth with severe anemia, pallor, and difficulties in breathing who developed SFN on the fourth postnatal day.

Our case of SFN is unusual in that it presented as a large plaque with multiple fluctuating cystic areas on the back of a 6-week-old infant. Drainage of the area, as performed successfully in our case, could be considered an optional treatment to facilitate resolution of fluctuant lesions. In our patient, asymptomatic hypercalcemia did not occur until 11 weeks of age and responded to a short course of oral glucocorticoid. Not unexpectedly, the diffuse and previously prominent cystic lesion was barely perceptible.

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