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Panniculitis in Children

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KEYWORDS

• Panniculitis • Children • Newborn • Fat necrosis • Nodules

The panniculitides are a group of heterogeneous inflammatory diseases that involve the subcutaneous fat. 1,2 Different pathomechanisms may result in inflammation of the subcutaneous fatty tissue. Because most cases of panniculitis show the same clinical appearance, a histopathologic study is usually required for a correct diagnosis. The histopathologic study of panniculitis is often difficult, however, because of a limited ability of the fatty tissue to display multiple patterns of disease. Furthermore, the need for big size biopsy specimens and the nonspecific histopathologic findings usually found in late-stage lesions of panniculitis make it even more difficult to reach a final diagnosis. A good clinicopathologic correlation and a skilful dermatopathologist are very useful tools for the identification of the causes of panniculitis.

The panniculitides in children are not a frequent consultation. Indeed, besides erythema nodosum (EN), which is by far the most common form of panniculitis, there exist merely anecdotal reports of most of the well-known panniculitides of adults. Some types of panniculitis, such as subcutaneous fat necrosis (SFN), appear only in children, but even those disorders are rarely seen. Most types of panniculitis have been reported to occur in children, however, and a diagnostic work-up must be performed in every case to ascertain a definite cause, especially considering that some of them may bear an uncertain prognosis or may even be fatal if untreated. In the absence of specific diagnostic algorithms for childhood panniculitis, clinicians must rest on the diagnostic procedures usually performed in adults. A skin biopsy is the best tool for the diagnosis of panniculitis, and clinicians must be wise in the election of the best time and site to obtain a skin sample of enough size and depth to permit an accurate diagnosis.

There are as many classifications of panniculitis as textbooks and articles. The most useful are

those based on the classical histopathologic distinction between lobular and septal panniculitis. An etiologic classification is more desirable, however, from a clinical point of view (Box 1).

In this article the panniculitides are thoroughly reviewed, and virtually all types of panniculitis are treated elsewhere. The focus is on the specific panniculitides of children. Discussed also are the pediatric aspects of the main types of panniculitides commonly seen in adults.

SPECIFIC PANNICULITIS OF CHILDREN Fat Necrosis of the Newborn

SFN of the newborn is an uncommon, benign panniculitis usually found in full-term infants. The onset of the disease may range throughout the neonatal period, and both early and late onset cases have been reported.

Clinically, it is characterized by multiple indurate, nonpitting plaques or nodules with or without erythema on the cheeks, buttocks, thighs, back, and extremities (Fig. 1).^{3–5} The lesions tend to spare the anterior trunk. The affected areas may be mobile over the underlying tissues. The overlying skin may show some inflammation, leading to a red or brown hue, or a completely normal appearance (see Fig. 1). The lesions are better detected by palpation of the nodules; these may be discrete or may be rather confluent, especially over the back. Many lesions may become fluctuant with liquefied fat, which may eventually discharge. Mild atrophy or depression of the skin may be noticed after resolution of the inflammatory nodules.

The cause for this disorder is unknown. In many cases perinatal complications are recorded, such as Rh factor incompatibility, meconium aspiration, umbilical cord prolapse, asphyxia, seizures, congenital heart disease, intestinal perforation, hypothermia, sepsis, anemia, gestational diabetes,

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Box 1 Classification of the panniculitis of children

Specific panniculitis of children

Cold panniculitis

Subcutaneous fat necrosis of the newborn

Sclerema neonatorum

Poststeroid panniculitis

Adult-type panniculitis appearing in children

Erythema nodosum

Enzymatic panniculitis

 α_1 -Antitrypsin panniculitis

Pancreatic disease

Infectious panniculitis

Bacterial

Mycobacterial

Fungal

Panniculitis in connective tissue disease

Lupus panniculitis

Deep morphea

Dermatomyositis

Polyarteritis nodosa

Granulomatous panniculitis

Subcutaneous granuloma annulare

Deep sarcoidosis

Physical panniculitis

Injections, iatrogenic, factitial

Extravasation

Blunt trauma

Malignant panniculitis

Histiocytic cytophagic panniculitis

Subcutaneous panniculitic T-cell lymphoma

Edematous, scarring vasculitic panniculitis (hydroa-like lymphoma)

Idiopathic panniculitides

Idiopathic lipoatrophic panniculitis (including connective tissue panniculitis, panniculitis associated with autoimmune disorders, lipophagic panniculitis of childhood, atrophic connective tissue panniculitis of the ankles, and recurrent lobular panniculitis)

Eosinophilic panniculitis (not a specific disease)

preeclampsia, or maternal use or abuse of drugs.^{3,6} SFN has been reported independently of the modalities of delivery (vaginal versus caesarean section), and an obstetric trauma does



Fig. 1. Subcutaneous fat necrosis of the newborn: erythematous indurate nodule on the external aspect of the left arm.

not seem to be a major cause of the disorder.6 Perinatal asphyxia and meconium aspiration seem to be the most crucial factors for the disorder. In less than 5% of cases SFN appears in otherwise normal newborns. The pathogenesis of SFN might be related to the subcutaneous fat composition in neonates, with a relatively high concentration of saturated fatty acids in respect to adult fat, rich in unsaturated fatty acids. Saturated fatty acids have a higher melting point at 64°C, which confers a greater propensity of neonatal fat to undergo crystallization under cold stress, with consequent adipocyte necrosis.7 Other proposed mechanisms include immaturity of the enzymatic systems involved in fatty acids metabolism and hypoxic injury to subcutaneous fat.3

Histopathologically, SFN shows a mostly lobular panniculitis, with a dense inflammatory infiltrate composed of lymphocytes, histiocytes, lipophages, multinucleated giant cells, and sometimes eosinophils interspersed among the adipocytes of the fat lobule.² Many adipocytes are replaced by cells with finely eosinophilic granular cytoplasm that contain narrow needle-shaped clefts radially arranged, which represent crystals of triglycerides of the adipocytes (Fig. 2). Some of these clefts may also be seen within the cytoplasm of multinucleated giant cells. Although the needle-shaped crystals within the cytoplasm of adipocytes and histiocytes are quite characteristic of SFN, they may not be seen, and similar crystals have been described in cases of poststeroid panniculitis and sclerema neonatorum (SN) (see later). In late-stage lesions there is septal fibrosis and areas of calcification may appear within the fat lobule.

The course of SFN is usually uncomplicated and spontaneous resolution over several weeks is expected. SFN may sometimes be complicated, however, with metabolic disbalances that may

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