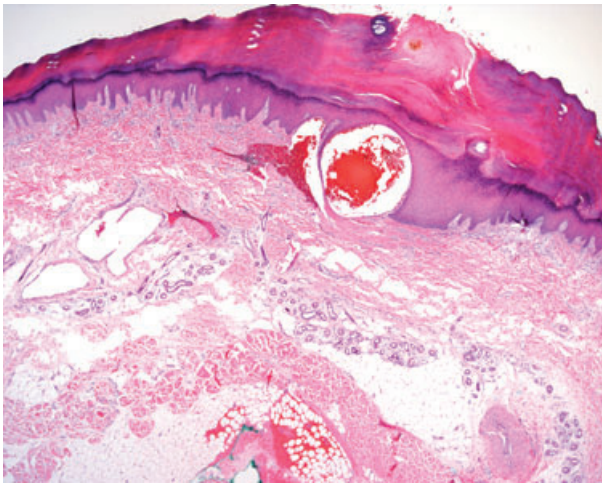

Figure 2. H&E section (2×).

equally affected. Approximately 150 cases have been reported in the literature. The differential diagnosis includes familial cutaneous and mucosal venous malformation syndrome, familial glomangiomatosis, diffuse neonatal hemangiomatosis, and Maffuci syndrome (1).

The most common site of visceral involvement is the gastrointestinal tract, most commonly the small intestine, documented by upper endoscopy, colonoscopy, or magnetic resonance imaging (MRI). Significant complications of gastrointestinal involvement include gastrointestinal hemorrhage, intussusception, volvulus, internal hemorrhage, and infarction (2).

Case reports of other organ system involvement include the central nervous system, liver, kidney, bladder, heart, thyroid, and spleen. However, these latter sites of involvement are far less common than the gastrointestinal tract (3). The initial evaluation of blue rubber bleb nevus syndrome includes a full history and physical examination, complete blood cell count, stool for blood, and appropriate investigations of involved organ systems (4,5). The coexistence of GI and central nervous system (CNS) involvement of blue rubber bleb nevus syndrome has been rarely reported. The patient continues to be followed by pediatric dermatology, pediatric gastroenterology, and pediatric neurology. Recently, she was seen by a pediatric oncologist and is being considered for systemic therapy with propranolol and bevacizumab as potential inhibitors of these vascular proliferations based on their experience with other vascular neoplasms.

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Postvaccination Morphea Profunda in a Child

Abstract: We report a new case of postvaccination morphea profunda (MP) in a child and discuss its different clinical presentations, prognosis, and therapy and its relationship with "solitary morphea profunda." A 2-year-old healthy girl presented with an induration of the anterior aspect of the left thigh of 9 months duration. The lesion had appeared 3 months after a third dose of diphtheria-tetanus-pertussis vaccine. Cutaneous examination showed an induration of 7 × 7 cm with an "orange peel" texture after pinching the skin. Histologic examination confirmed the diagnosis of MP. Systemic steroids (1 mg/kg/day) led to the stabilization of the lesion. After 4 months of treatment, we began the concomitant use of oral methotrexate (10 mg/wk) for 2 months. Methotrexate was then continued alone for 10 months, leading to a significant regression of the induration with no relapse.

Few cases of morphea profunda (MP) occurring at the site of intramuscular vaccine injection have been reported. Herein, we report a new case in a child.

CASE REPORT

A 2-year-old girl presented with an asymptomatic induration of the left thigh of 9 months duration. She had been correctly vaccinated. The last vaccine was the third dose of diphtheria-tetanus-pertussis (DTP) that had been intramuscularly administrated on the left thigh 3 months before the appearance of skin induration.

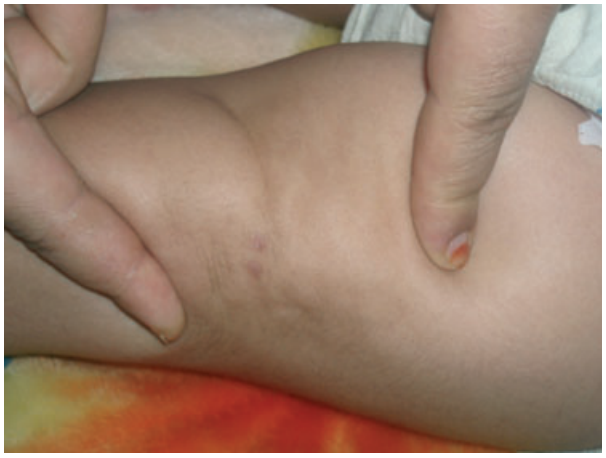

Figure 1. Skin-colored induration of 7 × 7 cm of the anterior aspect of the left thigh with an "orange peel" texture, more evident after pinching the skin.

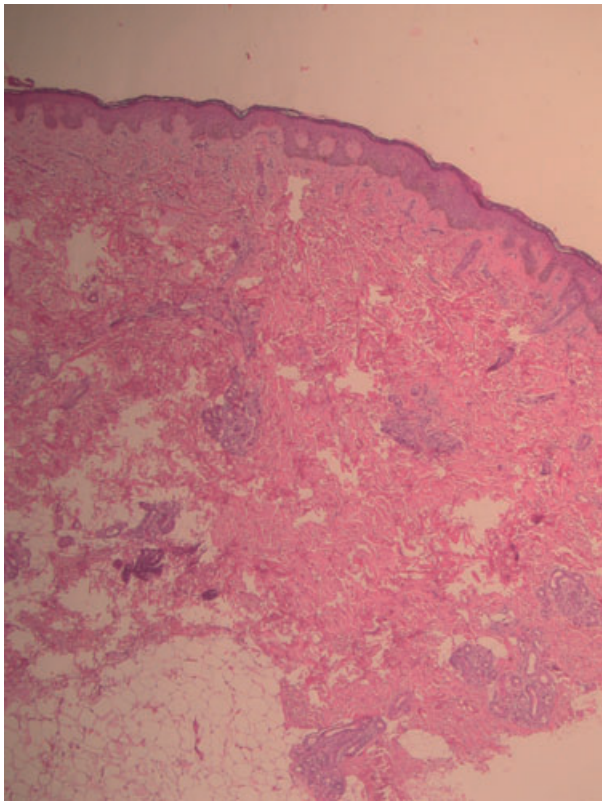

Figure 2. Normal epidermis with large, thick, hyalinized collagen bundles of the entire dermis and subcutaneous adipose tissue. The sclerosis appears to compress the eccrine sudoral glands.

Cutaneous examination showed, on the anterior aspect of the left thigh, a skin-colored induration of 7 × 7 cm with an "orange peel" texture that was more evident after pinching the skin (Fig. 1). Left knee mobility was preserved. The remaining physical examination was normal. Histologic examination of a cutaneous biopsy showed, under a normal epidermis, a diffuse sclerosis with large, thick, hyalinized collagen bundles of the entire dermis and subcutaneous adipose tissue. The sclerosis compressed the eccrine sudoral glands (Fig. 2). Histologic features, together with the clinical aspect, were consistent with the diagnosis of MP. Biological (hemogram, serum chemistry) and immunological tests (antinuclear and anti-Scl70 antibodies) were within normal limits or negative. Borrelia serology was negative. Prednisolone 1 mg/kg per day was started (15 mg/day), leading to a stabilization of the lesion. After 4 months of treatment, we began the concomitant use of oral methotrexate (15 mg/m^2/week, 10 mg/wk). She was then weaned off oral steroids over a 2-month period while methotrexate was maintained for 10 months, leading to significant regression of the induration with no relapse after 6 months of follow-up.

DISCUSSION

Postvaccination MP is rarely reported in the literature (1–4). It seems to occur only in predisposed patients. Its pathogenesis implicates immune response against specific and nonspecific antigens but also other phenomena secondary to trauma itself (2). Incriminated vaccines were DTP, measles-mumps-rubella, hepatitis B, tetanus, and influenza (1–4). Khelifa et al (4) reported a case of atrophic solitary MP of the shoulder; they named it "primary" despite its occurrence at a site of previous influenza vaccine injection. In this report, the clinical aspect consisted of atrophy with a cupuliform depression, different from the classically described aspect of solitary MP that is a solitary sclerotic plaque, sometimes associated with changes in skin pigmentation or texture, of the paraspinal region and sometimes of the upper buttock. An intramuscular injection, ignored or forgotten by the patient, may be the cause of its appearance in the latter location. Taking into account all the available reported cases, we can consider that all cases of MP, regardless of the clinical aspect (orange peel-like or atrophic) can be included into the so-called solitary MP, being vaccine or trauma induced or not. Treatment includes local potent steroids, systemic steroids (0.66–1 mg/kg/day) with or without methotrexate (15 mg/m^2/week, maximum 20 mg) (5). The possibility of progression, with risk of joint impairment, generalized lesions, or even systemic sclerosis, justifies aggressive therapy, especially combined oral steroids (for 3 months) with methotrexate (for 12 months or until treatment failure) (1–3,5). The early use of methotrexate can hasten steroid weaning and limit steroid side effects (5).

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Profuse Congenital Familial Milia with Absent Dermatoglyphics (Basan's Syndrome): Description of a New Family

Abstract: **Milia are common, small, keratin-containing cysts frequently seen in all age groups. They may arise spontaneously or develop after a variety of stimuli. They can be found alone or as part of syndromes. We present a female neonate born not only with profuse facial milia, but also with acral bullae and absent dermatoglyphics. Similar features were seen in several members of her family. These findings correspond to the syndrome known as Basan's syndrome, a rare autosomal-dominant inherited dermatosis characterized by profuse congenital milia, transient neonatal acral bullae, and absence of dermatoglyphics.**

Milia are common, small, keratin-containing cysts. They can be classified as primary milia when they arise spontaneously or secondary milia when they arise as a response to a variety of stimuli. Lesions can be single, few, or profuse. They can be isolated or associated with other clinical findings (1).

We present the case of a female neonate born with profuse facial milia, acral erosions secondary to bullae, and absent dermatoglyphics. At least four members of her family history had similar characteristics. Because of the clinical findings in the proband and her family, the diagnosis of Basan's syndrome was made.

REPORT

Dermatologic evaluation was requested for a 1-day-old girl. She was born at full term by vaginal delivery after an uneventful pregnancy.

On physical examination, she presented with profuse milial cysts over her chin, malar region, forehead, and nose (Fig. 1). Her palms and soles had extremely fragile skin, leading to a few palmo-plantar blisters and erosions (Fig. 2). Absence of palmo-plantar dermatoglyphics was also noticed (Fig. 3). Fingernails and toenails were excessively rounded, and she had bilateral syndactyly of the second and third toes. Her ears and palate were normal in morphology.

A thorough family history revealed that dermatoglyphics were also absent in her father (Fig. 4), her grandfather, and her two aunts, one of whom had also been born with profuse facial milia. The four also had painful calluses on the palms and soles and short

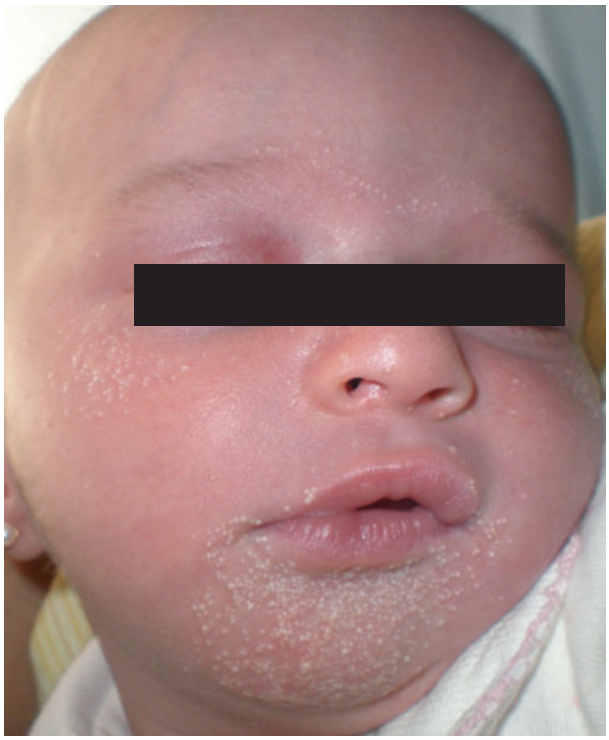

Figure 1. Profuse facial milia. Numerous lesions especially over the chin and malar region at birth.