Precalcaneal Congenital Fibrolipomatous Hamartoma

Hamartoma fibrolipomatoso precalcáneo congénito


Figure 2  Predominantly perivascular and interstitial inflammatory dermal neutrophilic infiltrate.

tant metastases of the breast cancer, except in 1 case where it was described in association with skin metastases; only 1 case has been reported in a patients receiving granulocyte-colony stimulating factor. Compared to classic Sweet syndrome, the systemic symptoms are mild (in some cases absent), neutrophilic leukocytosis is less common, and relapses are less prevalent. The other distinctive difference is that the lesions are restricted to the site of the lymphedema. Treatment with antibiotics appears to cure the lesions more rapidly than treatment with systemic corticosteroids or potassium iodide, and the disorder also responds well to oral anti-inflammatory drugs and high-potency topical corticosteroids.

The pathophysiology of this condition is not understood and a number of theories have been put forward, all of which posit a local disruption in cell trafficking due to inadequate lymphatic drainage caused by the lymphadenectomy and the radiation therapy. The hypothesis is that cytokines accumulate at the site of the lymphadenectomy attracting neutrophils to an area with reduced immunocompetence and thereby favoring local development of malignancies, infections, and immune disorders such as neutrophilic dermatosis.

The differential diagnosis should include infections such as cellulitis, erysipelas, folliculitis, and herpes zoster, as well as thrombophlebitis and recall phenomenon. Histology should rule out chronic radiodermatitis, carcinoma erysipeloides, and contact dermatitis. The clinical course, biopsy findings, and additional tests will all help to establish the definitive diagnosis.

In conclusion, we report a new case of neutrophilic dermatosis on the site of a lymphedema. This condition is a localized variant of classic Sweet syndrome with differences that include not only the location of the lesions but also a milder course with fewer systemic symptoms, fewer relapses, and a good response to oral antibiotics, anti-inflammatory drugs, and topical corticosteroids. Despite the large number of cases of breast cancer and of lymphedema arising as a complication of the different treatments used in these patients, few cases of neutrophilic dermatosis have been documented. This is probably due to confusion with other inflammatory or infectious diseases that are more common in this group of patients.

References


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To the Editor:

Precalcaneal congenital fibrolipomatous hamartoma (PCFH) is a rare and benign childhood skin disorder, with only a few cases reported in the literature. It has been referred to by a variety of names, including pedal papules in the newborn, congenital piezogenic-like papules, and bilateral congenital adipose plantar nodules.

We present the case of a 9-month-old girl, with no relevant personal or family history, whose family brought her to consultation for the presence of symmetric subcutaneous
nodular lesions on the plantar surface of both feet, just anterior to the heels (Fig. 1). The lesions had been present since birth. They had a soft consistency, were poorly circumscribed, and were not adherent to the superficial or deeper layers. The overlying skin was normal. They had a diameter of 1.5 cm at their widest point and appeared to be asymptomatic. There were no other evident abnormalities. Based on these clinical features, a diagnosis of PCFH was made. The lesions remained unchanged over more than 6 months of follow-up. During this time, the patient started to walk and there was no gait disturbance. Treatment was not considered necessary.

PCFH was first reported in 1990 by Larralde et al., who described the nodules as pedal papules in the newborn. In 1996, Larregue et al. called the disorder precalcaneal congenital fibrolipomatous hamartoma and this term has been used ever since. The literature contains only isolated case reports and small series. Although PCFH appears to be rare, it is probably underdiagnosed because of low awareness among clinicians and the benign nature of the nodules.

It is typically present at birth, but it can develop later. It has been reported to be slightly more common in males than in females.

Its pathogenesis is unknown. Early descriptions suggested that it might be due to incomplete regression of fetal tissue as fibrolipomatous fetal tissue in the area of the heel exhibits physiologic hypodermic hypertrophy. However, the fact that fetal adipose tissue has never been detected on histologic examination and that a higher incidence of PCFH has not been noted in preterm infants would seem to contradict this theory. Other possible causes that have been suggested include a congenital alteration in the fibroconnective trabecular network of the adipose tissue and a tissue overgrowth disorder.

PCFH tends to appear sporadically, although there have been reports of a familial association, with an apparently autosomal dominant pattern of inheritance. It has also been suggested that there might be an X-linked or mitochondrial inheritance pattern.

PCFH is characterized by the presence of soft, symmetric, mobile, subcutaneous nodular lesions that are generally located on the mid region of the soles of both feet, just anterior to the heel, although they can also extend onto the heel. On occasions, the nodules can be more prominent on one foot than on the other. The color of the overlying skin is normal. The nodules are asymptomatic and have not been reported to cause gait disturbance. Accordingly, they do not generally require treatment, but surgical excision is an option if they cause discomfort.

The natural history of PCFH is not well established. The nodules tend to increase in size as the child grows, and there have been reports of lesions persisting well into adulthood. However, because so few reports have been published on this relatively recently described disorder, little is known about clinical course or potential for spontaneous regression. There have been no reports to date of associated abnormalities or potential for malignant transformation.

Histology is generally not necessary to diagnose PCFH. Histologic findings include mature adipose tissue surrounded by collagen fibers of different thicknesses and normal elastic fibers. In addition, there may be mucin deposits at the periphery and within the fat lobules and an increased number of blood vessels without associated perivascular alterations. The above findings have been confirmed in ultrastructural studies.

The differential diagnosis should include piezogenic papules, which are generally found in adults and are caused by herniation of fat through the dermis following injury. Unlike PCHF nodules, piezogenic papules are typically multiple and are often painful, particularly on walking. Other types of neonatal nodular lesions should also be considered in the differential diagnosis. Examples are juvenile fibromatosis, lipomas, nevus lipomatosus, dermal hypoplasia, infantile hemangioma, congenital hemangioma, lymphatic malformation, or plexiform neurofibroma, all of which are typically unilateral. In most cases, these conditions can be distinguished by their clinical features. In equivocal cases, however, histologic examination, or even less invasive procedures such as transillumination or echo Doppler, can be helpful.

It is important to be familiar with this probably underdiagnosed disease and to inform parents that it is a harmless, asymptomatic condition that is not associated with other abnormalities and generally does not require further tests or aggressive treatments. Parents should also be informed that it might be hereditary, although the pattern of transmission has not yet been well established.

References


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