SECTION EDITOR: MICHAEL E. MING, MD

Asymptomatic Labial Papules in a Teenager

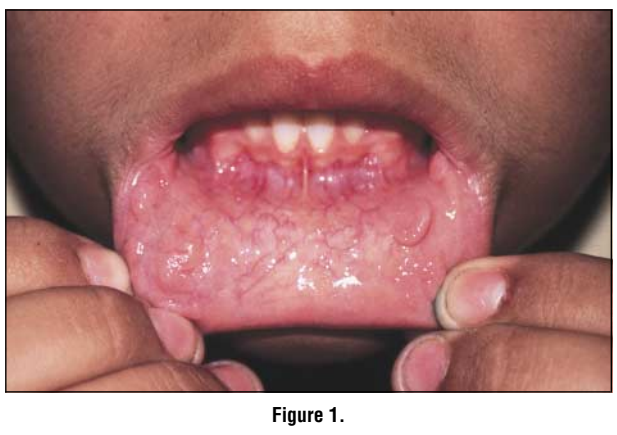
Carrie K. Guill, MD; Linda Y. Hwang, MD; Vivek Iyengar, MD; Sylvia Hsu, MD; Baylor College of Medicine, Houston, Tex

REPORT OF A CASE

A 12-year-old Native American girl presented with an 8-month history of asymptomatic growths on the mucosal aspect of her upper and lower lips. She was not sexually active and was otherwise healthy. There was no family history of similar lesions.

Physical examination revealed several nontender papules measuring 0.3 to 1.0 cm scattered over the labial mucosa (**Figure 1**). The rest of the oral mucosa, including the vermilion, buccal mucosa, and tongue, was spared. When stretched, the papules became nearly imperceptible. A shave biopsy specimen was obtained from one of the papules (**Figure 2** and **Figure 3**).

What is your diagnosis?



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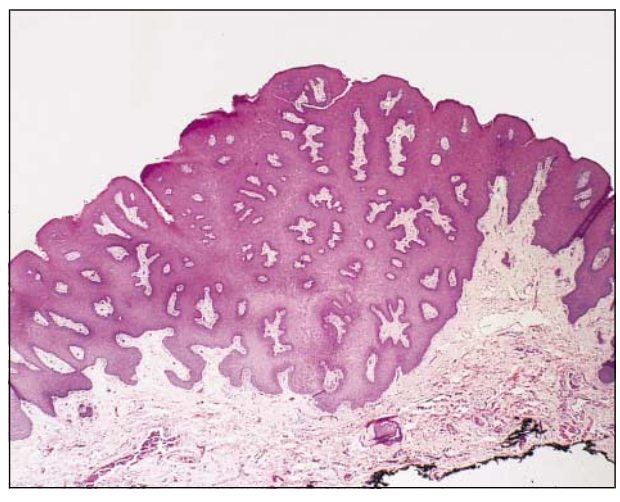

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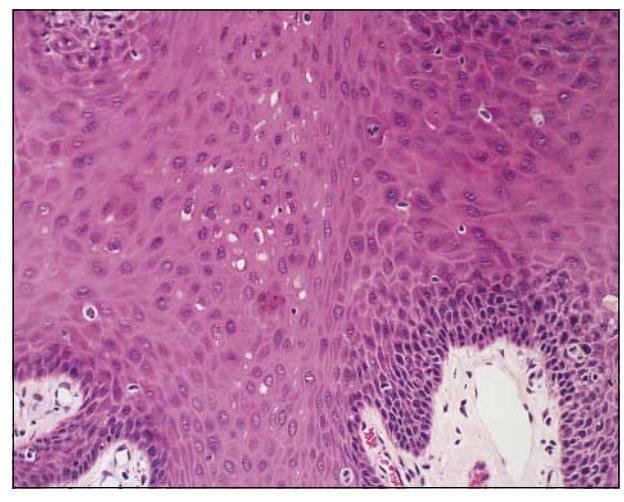


Figure 3.

Multilobated Abdominal Nodule

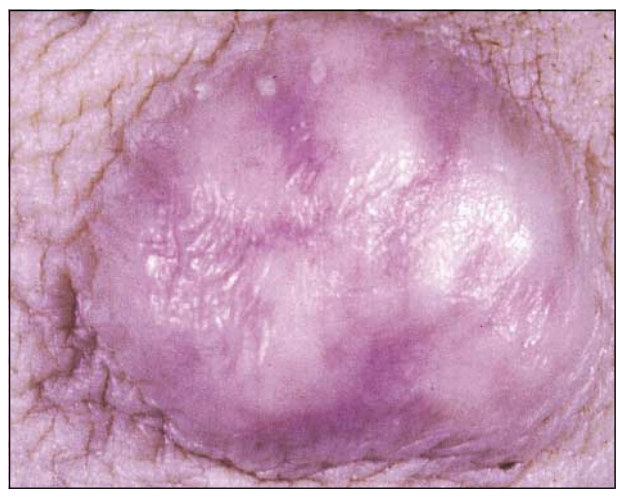
Lucia Pozo, MD; Enrique Jorquera, MD; Salvador J. Diaz-Cano, MD; St Bartholomew's and the Royal London School of Medicine and Dentistry, London, England (Drs Pozo and Diaz-Cano), and Clinica Sagrado Corazon, Seville, Spain (Dr Jorquera)

REPORT OF A CASE

A 56-year-old woman presented with an asymptomatic abdominal nodule. She stated that the lesion had originally appeared after trauma to the site long ago but that it had been growing slowly since then. Her medical history was remarkable for diabetes, which she controlled with oral medication.

Physical examination of the lower part of her abdomen showed a 3-cm soft skin-colored nodule with a nonulcerated and multilobated surface (**Figure 1** and **Figure 2**). No peripheral lymphadenopathy was palpable. The lesion was excised (**Figure 3** and **Figure 4**).

What is your diagnosis?



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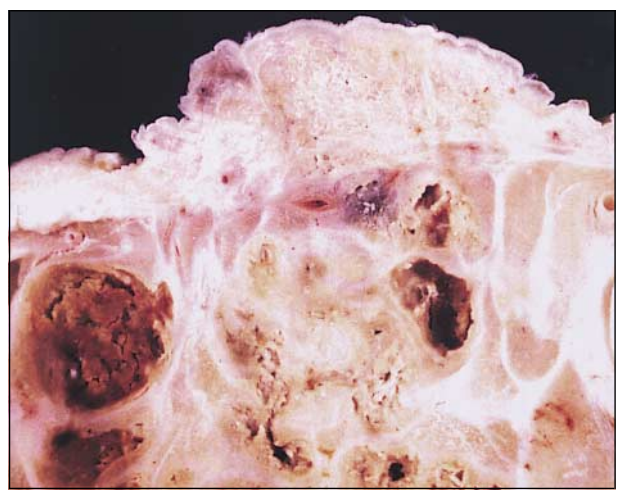

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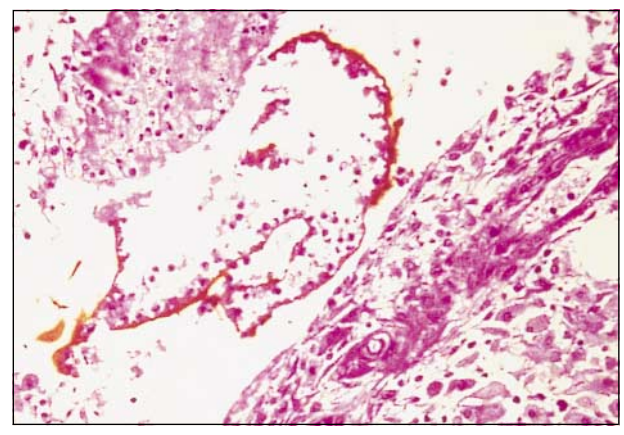

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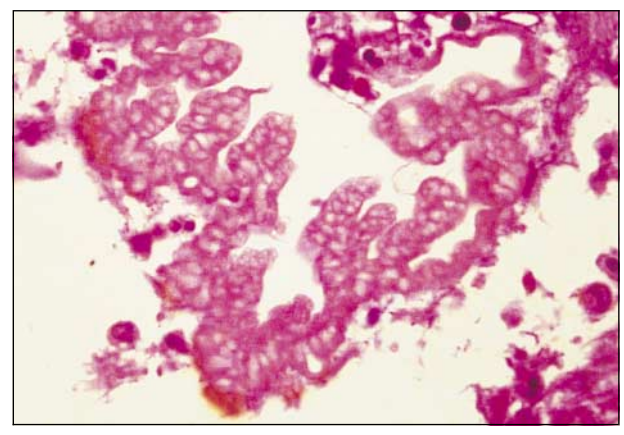


Figure 4.

Multiple Punctate Pits in a 27-Year-Old Woman

Julie E. Greenberg, MD; George Elgart, MD; Edward A. Gross, MD; University of Miami, Miami, Fla

REPORT OF A CASE

A 27-year-old woman presented with a lifelong history of a rash on the left palm of her hand. On the medial edge of the left hand and the left fifth finger, there was a linear patch of punctate keratotic papules (**Figure 1**). A punch biopsy specimen was obtained from the left hand and routinely stained with hematoxylin-eosin (**Figure 2**).

What is your diagnosis?

Figure 1.

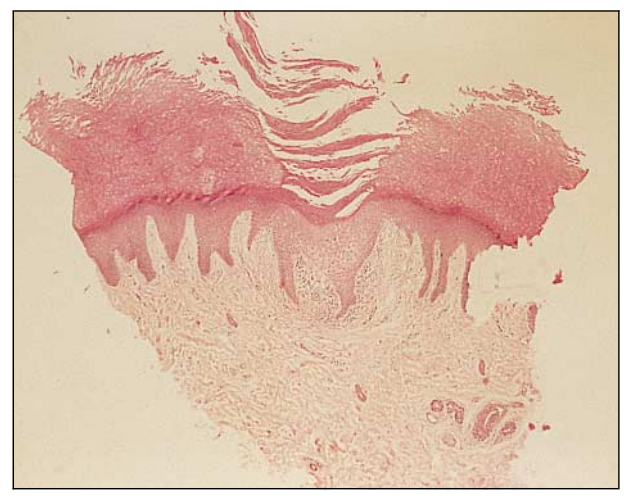


Figure 2.

Nodule on the Shoulder of a Young Woman

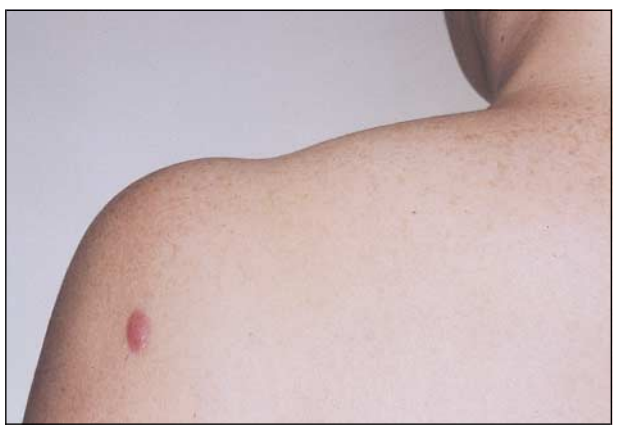
Simone Laube, MD, MRCP; Adam M. Boyde, MBBS, BSc; Richard J. Motley, MA, MD, MRCP; University Hospital of Wales, Cardiff

REPORT OF A CASE

A 36-year-old white woman presented with a 5-month history of an irritating lesion on her left shoulder. A crusted scab had arisen and fallen off, leaving her with a 1.5-cm, smooth, tender, intradermal nodule with no obvious surface abnormality (**Figure 1**).

The differential diagnosis included a keloid scar, adnexal tumor, and amelanotic melanoma. A biopsy specimen from the center of the lesion was obtained for histologic (**Figure 2**) and immunohistochemical staining with α-smooth muscle actin (**Figure 3**). The nodule remained exquisitely tender, and a complete excision was performed.

What is your diagnosis?



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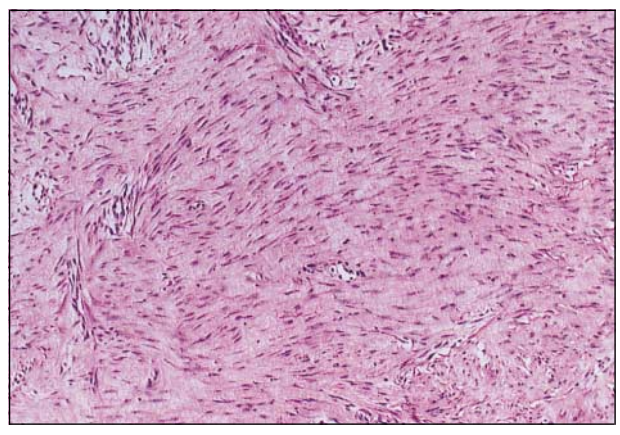

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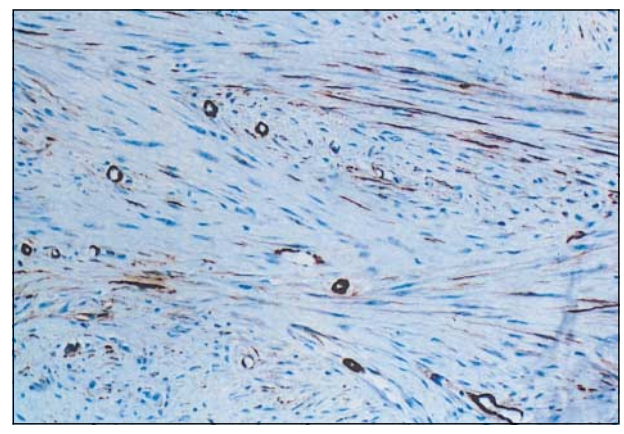

Figure 3.

Asymptomatic Labial Papules in a Teenager

Diagnosis: Focal epithelial hyperplasia (FEH [Heck disease]).

MICROSCOPIC FINDINGS

The biopsy specimen showed hyperplastic mucosal epithelium with acanthosis and anastomosing rete ridges. (Figure 2). There was slight pallor of the squamous epithelium and focal koilocytosis (Figure 3), but significant cellular atypia and mitoses were absent. There was no significant associated inflammation. The specimen was found to be positive for human papillomavirus (HPV) type 13 by polymerase chain reaction analysis.

DISCUSSION

Focal epithelial hyperplasia, or Heck disease, is a benign proliferative condition that most commonly affects the oral mucosa, typically in girls. It is characterized by multiple, asymptomatic, round, soft papules on the oral mucosa measuring a few millimeters in diameter.[1,2] The eruption affects the lower lip, upper lip, buccal mucosa, and tongue, in order of frequency.[3] The color of the lesions is that of the surrounding mucosa, and the papules may seem to disappear when the oral mucosa is stretched.[1]

Histopathologic examination is characterized by epithelial hyperplasia, with acanthosis of the superficial epithelium[4] and horizontal anastomosis of elongated and/or clubbed rete ridges.[5] Other findings may include pallor of the epithelium and focal koilocytosis,[1] focal parakeratosis, and slight perivascular, lymphocytic infiltrates in the superficial lamina propria.[5] Mitosoid figures, or cells showing mitosislike nuclear degeneration thought to represent arrested division, have been reported.[6]

Focal epithelial hyperplasia, which is most prevalent in Native Americans, Eskimos, and South Africans, has a predilection for female children. However, in Eskimos, FEH occurs more often in adults.[7] Possible contributing factors include genetic predisposition, a diet lacking in fresh produce, poor nutrition, crowded living conditions, and poor personal hygiene.[7] However, the most accepted theory of the cause of this condition is infection with HPV. In particular, HPV types 13 and 32 have been detected in these lesions by in situ hybridization,[8] a finding that has been confirmed on numerous occasions. However, detecting the virus in tissue is highly technique dependent, and as a result, HPV may escape detection in up to 30% of cases.[3]

The differential diagnosis of FEH includes condyloma acuminatum, verruca vulgaris, oral florid papillomatosis, biting papilloma, mucosal neuroma, white sponge nevus, and diffuse epithelial hyperplasia.[5] It is important to distinguish condylomas from FEH in young girls to avoid inaccurate implications of sexual transmission or abuse.[4]

The course of FEH is variable. Many lesions spontaneously resolve within a few months to 1 year. However, they may persist or recur over many years.[5] Possible treatment options include cryotherapy, topical eradicating agents such as podophyllum resin, surgical excision, and vitamins; however, these therapies yield inconsistent results. Carbon dioxide laser destruction has shown promising results for persistent lesions, with treated patients remaining free of lesions after 18 months.[9]

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Multilobated Abdominal Nodule

Diagnosis: Subcutaneous spherulocystic disease.

MICROSCOPIC FINDINGS AND CLINICAL COURSE

Gross examination of the specimen revealed a multicystic mass with a "Swiss cheese" appearance in the dermis and subcutaneous soft tissue. On microscopic examination, the lesion showed a mixed granulomatous inflammation surrounding cystic spaces that appeared to be lined by clusters of foamy histiocytes. The cystic spaces contained numerous membrane-encircled round bodies (spherules). The surrounding tissue showed occasional foreign body–type giant cells, refractive extracellular brown pigment, chronic inflammatory infiltrate, and fibrous reaction. There was no evidence of vasculitis or birefringent foreign bodies.

Diastase periodic acid–Schiff, Grocott methenamine silver, gram, Giemsa, and Ziehl-Neelsen stains were all negative for microorganisms. The extracellular pigment stained positive for hemoglobin (immunohistochemical demonstration) and hemosiderin ("brown bodies," Perls) and negative for melanin (Masson-Fontana) and bilirubin (Hall), confirming a previous hemorrhage.

The nodule was completely excised, and there has been no recurrence for more than 5 years.

DISCUSSION

Myospherulosis, currently named *spherulocytosis* or *spherulocystic disease*,[1,2] is a chronic granulomatous inflammation characterized by cystic structures.[3] The diagnosis depends on the histologic identification of the characteristic cystic spaces filled by erythrocyte aggregates and described as "partly filled bags of marbles."[1] There is a surrounding foreign body or foamy granulomatous reaction and variable number of lymphocytes, plasma cells, and eosinophils.

This condition has been described in 2 different settings. In Western countries, spherulocytosis usually affects the nose, paranasal sinuses, and middle ear after surgery or topical treatment with greasy vehicles, especially antibiotics.[4] In Africa, it presents as subcutaneous nodules on the limbs and buttocks and is usually related to previous trauma in those areas. Both settings involve the presence of hemorrhage in an inflamed tissue with high lipid content.[1] The membranes of the erythrocytes are altered by the oily substance, inducing clumping of red blood cells, described histologically as spherules, and finally eliciting a foreign body reaction or foamy histiocytic response.[5,6] It has been suggested that an altered immunologic response causes a granulomatous reaction against erythrocytes in cases involving hemorrhage and fat necrosis (eg, after surgery or trauma). Fat necrosis or the greasy vehicles of drugs (petrolatum, lanolin) provide the lipids required for spherulocystic disease. In this situation, spherulocystic disease could be considered an iatrogenic reaction to some ointments when they are applied directly to skin and mucosa.[6,7]

The differential diagnosis includes other granulomatous diseases, especially fungal infections, sarcoidosis, and foreign body granulomas. Special stains and cultures should be routinely used to identify microorganisms, granuloma features, and foreign bodies that will help in making the diagnosis. Extensive panniculitis, which can cause fat necrosis, must also be considered.

In summary, our patient had the typical clinical features of the African type of spherulocystic disease, a very rare presentation in Western Europe. Therefore, this condition should be included in the differential diagnosis of cutaneous nodules after local trauma occurs along with fat necrosis. Surgical excision has been effective in most patients with low-grade recurrences.[1]

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Multiple Punctate Pits in a 27-Year-Old Woman

Diagnosis: Porokeratotic eccrine ostial and dermal duct nevus (PEODDN).

MICROSCOPIC FINDINGS

There was a narrow dell in the epidermal surface, with a pattern of cornification identical to that seen in the cornoid lamella of porokeratosis. The stratum corneum formed a column of parakeratosis from the surface of the viable epidermis. This feature was limited to an area that appeared to be overlying the acrosyringium and was contiguous with it at the level of the granular layer of the surrounding skin. Thus, the findings appeared to overlie the ostium of the eccrine duct. There was an associated mild superficial dermal chronic infiltrate.

DISCUSSION

PEODDN is a rare entity that is composed of an eccrine hamartoma associated with cornoid lamellation. Abell and Read[1] first described PEODDN in 1980 in a 3-year-old girl who had linear keratotic papular lesions on the medial surface of her left foot. In 1979, Marsden et al[2] reported a similar case. Our case, like that of Abell and Read, was not associated with any other congenital abnormalities and followed the lines of Blaschko.

The lesions of PEODDN may present clinically either as palmoplantar papules that resemble comedones, with keratin plugs filling the central pits of these lesions, or as keratotic papules and plaques that resemble linear verrucous epidermal nevi on other areas.[2] Cornoid lamellae, which are exclusively associated with eccrine acrosyringia, are the histologic hallmark of PEODDN. Cases of PEODDN occurring on the upper and lower limbs, forehead,[3] axillae,[4] neck,[5] trunk,[3,5] and buttocks have also been reported.[6] Sassmannshausen et al[7] analyzed 24 previously reported cases and discovered a nearly equal sex ratio (male-female, 12:10, with 2 cases of unknown sex), an absence of a family history for PEODDN in all cases, 15 cases in which lesions were present from birth, and 23 cases in which the extremities were affected. The age range for appearance of the lesions is from birth to 60 years. Of 19 patients described by Leung et al,[5] 3 presented with bilateral lesions and 16 with unilateral lesions.

In 1992, Bergman et al[8] proposed that PEODDN is an abnormal keratinizing epidermal invagination transversed by an acrosyringium-like duct rather than by a dilated, porokeratotically plugged acrosyringium and dermal duct. This hypothesis was supported by the findings of immunohistochemical studies for carcinoembryonic antigen that were conducted in 1995[9] and 1996.[10] The differential diagnosis for PEODDN includes nevus comedonicus, punctate porokeratosis, linear epidermal nevus, spiny keratoderma, and linear porokeratosis.

PEODDN is an asymptomatic disease but in certain cases may be a cosmetic problem for the patient. Treatment options for PEODDN, which are similar to those for epidermal nevus, include topical steroids under occlusion, ultrapulsed carbon dioxide laser ablation, topical calcipotriol ointment, cryotherapy, cautery, and surgical excision.

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Nodule on the Shoulder of a Young Woman

Diagnosis: Dermatomyofibroma.

MICROSCOPIC FINDINGS

Histopathologic examination showed a well-circumscribed area located within the reticular dermis and composed of monomorphic spindle cells in intersecting fascicles that were aligned parallel to the epidermis. The spindle-shaped cells had plump nuclei and prominent nucleoli. Occasional mitoses were seen. Elastic fibers were absent. There was a patchy chronic inflammatory cell infiltrate in a perivascular distribution in the deep dermis. The lesion extended to the epidermis, which appeared atrophic but showed no specific features.

Immunocytochemical studies showed that the spindle cells were positive for vimentin, with focal positivity for α-smooth muscle actin. Stains were negative for S100 protein, HMB-45, HHF-35, CD34, desmin, and factor XIIIa.

DISCUSSION

Hügel[1] first described dermatomyofibroma in the German-language literature in 1991 using the term *plaqueförmige dermale Fibromatose* (plaquelike dermal fibromatosis). Kamino et al[2] reported the first 9 cases in the English-language literature in 1992. About 50 cases have been reported so far, usually as a firm plaque or nodule located on or around the shoulders, axillae, upper part of the arms, neck, or anterior abdominal wall in women. A few cases have been reported in males and children as young as 4 years.[3] Although the lesions are usually 1 to 2 cm in diameter, tumors of up to 8 cm have been described.[4] The tumor grows slowly, is usually asymptomatic, and does not recur after excision. Hügel et al[5] described a woman with multiple plaques involving an area the size of a palm on her right breast. The plaques had developed over a period of 40 years. The clinical and histologic differential diagnosis of dermatomyofibroma includes dermatofibroma, dermatofibrosarcoma protuberans, neurofibroma, piloleiomyoma, fibromatosis, and keloid.[3,5,6]

Microscopically, the tumor is a well-circumscribed but nonencapsulated proliferation of uniform, slender, spindle-shaped cells, which form well-defined intersecting fascicles in the reticular dermis that are arranged parallel to the skin surface. The lesion may extend into the upper subcutaneous fat tissue. Mitotic figures are rare. Thin collagen fibers separate the spindle-shaped cells. Elastic fibers are normally present and can appear thickened. Adnexal structures are spared. The epidermis shows mild acanthosis with elongation of the rete ridges.

Immunohistochemical studies are positive for nonspecific muscle actin and vimentin, but negative for desmin, factor XIIIa, S100 protein, and CD34.[7] In some cases, tumor cells have also been reported to be positive for α-smooth muscle actin.[7,8] On electron microscopy, the lesion is shown to comprise a mixture of fibroblasts and myofibroblasts.[9] Dermatomyofibroma is a rare, benign, soft tissue tumor with distinct clinical and histopathologic features.

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