Pedunculated Pink Papule on the Nose

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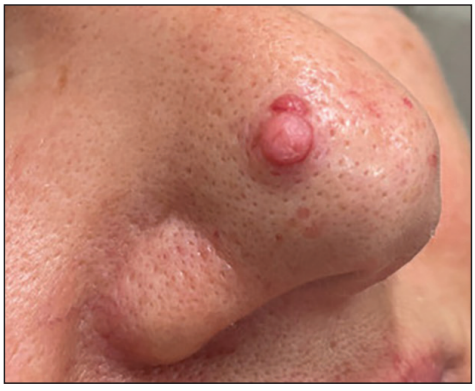

A 60-year-old woman presented to the dermatology department with a 6-mm, firm, pink, nonulcerated, nonmobile papule on the right nasal side wall of 1 year's duration. It had grown slowly and was asymptomatic with no tenderness or bleeding. No other skin lesions were noted on physical examination, and her medical history was otherwise unremarkable. A shave biopsy was performed.

WHAT'S YOUR **DIAGNOSIS?**

a. angiofibroma
b. angiomyolipoma
c. basal cell carcinoma
d. pedunculated lipofibroma
e. pyogenic granuloma

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THE **DIAGNOSIS:** Pedunculated Lipofibroma

Histopathology confirmed a pedunculated/polypoid lesion with intradermal lobules of adipocytes/mature adipose tissue admixed with connective tissue bundles and vascular ectasias. Overlying epidermal acanthosis with slight papillomatosis and hyperkeratosis was present (Figure 1). Masson trichrome staining highlighted admixed collagen bundles (Figure 2). Verhoeff–van Gieson staining showed marked reduction in elastic fibers (Figure 3). Immunostaining was negative for smooth muscle actin and desmin. A diagnosis of pedunculated lipofibroma on the nose was made based on both clinical and histopathologic findings.

Pedunculated lipofibroma (or solitary lipofibroma) is the solitary form of nevus lipomatosus cutaneous superficialis (NLCS).[7] First described by Hoffmann and Zurhelle[1] in 1921, NLCS is an uncommon benign hamartomatous cutaneous lesion/connective tissue nevus that also has a classic multiple form.[1-13] The etiology of NLCS remains unclear, but several theories have been proposed to explain its pathogenesis, including deposition of adipocytes secondary to degenerative changes in dermal connective tissue, focal/local heterotopic development of adipose tissue, and derivation from differentiating lipoblasts (preadipose tissue) originating from precursor vascular or perivascular cells.[2-13]

Pedunculated lipofibroma usually develops during the third to sixth decades of life and manifets as a single cutaneous lesion with a smooth surface, often on a non–pelvic girdle location.[7-13] No particular predilection sites are noted, with lesions reported on the arm, axilla, back, upper thigh, knee, and sole.[5,12] There are rare reports of this type of NLCS on the ear, scalp, forehead, or eyelid.[7-11]

In the classic form of NLCS, multiple cutaneous lesions are present at birth or develop within the first 2 to 3 decades of life.[2-6] Lesions consist of soft, nontender, pedunculated, flesh-colored or yellowish papules and nodules with a verrucoid or cerebriform surface that may later coalesce to form plaques.[2-6] Predilection sites include the pelvic girdle, buttocks, sacral and coccygeal regions, and upper posterior thighs, with a linear or zosteriform pattern of distribution.[2-6] Rarely, the classic form can arise in elderly patients and/or at an atypical anatomic location (eg, clitoris,[3] shoulder,[5] thorax,[5] abdomen[5]) and can demonstrate extension of lesions across the midline.[4] Rare cases of classic NLCS on the scalp[2] and face[3-6] have been reported, including lesions localized to the nose[3] and chin[4] and others extending from the right mandible to the neck[5] and right lower lip to the submandibular/posteriorateral cervical region.[6] In some cases, lesions clinically resemble plane xanthoma[4] and localized scleroderma.[6]

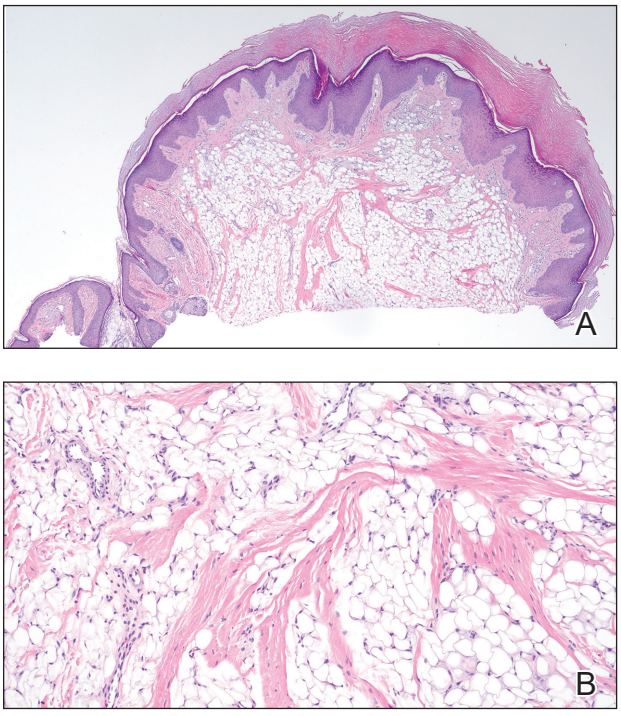

FIGURE 1. A, Histopathology demonstrated a pedunculated/polypoid lesion with intradermal lobules of mature adipose tissue, admixed with connective tissue bundles and vascular ectasias (H&E, original magnification ×20). B, Overlying epidermal acanthosis with slight papillomatosis and hyperkeratosis was noted (H&E, original magnification ×100).

Adotama et al[13] proposed a set of clinical features to differentiate classic NLCS, pedunculated lipofibroma (solitary NLCS), and fibroepithelial polyp with adipocytes (distinguished by their furrowed surface, hyperpigmentation, and anatomic predilection for the neck and axilla). Lesions are asymptomatic in both forms of NLCS.[2-13] Family history or predominant sex involvement have not been reported in either clinical type.[2-13] Reported associations with NLCS include a number of endocrinologic conditions including diabetes.[7] Other coexisting skin findings can include café-au-lait macules, leukodermic (white) spots, overlying hypertrichosis, comedolike alterations, angiokeratoma, hemangioma, and folliculosebaceous cystic hamartoma.[4] None of these were evident in our patient.

Lesions from both types of NLCS are indistinguishable on histopathology, characterized by the presence of a central core of ectopic mature adipocytes in the papillary/reticular dermis.[2-13] Additional light microscopic features (some seen in our case) have been described, including thickened collagen bundles, reduction of

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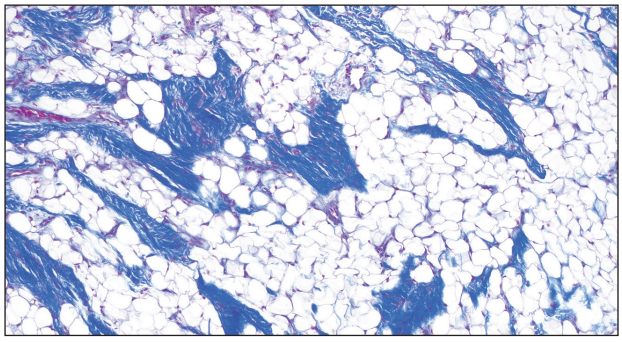

FIGURE 2. Admixed collagen bundles were highlighted (Masson Trichrome, original magnification ×100).

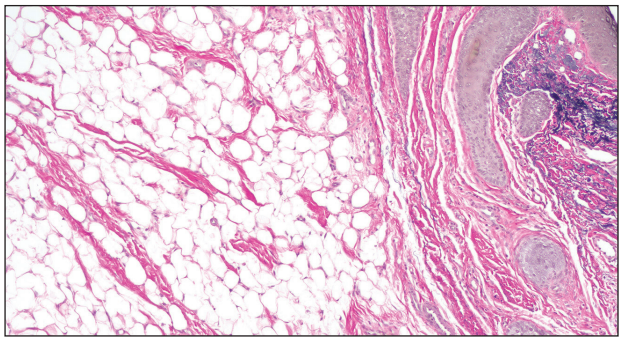

FIGURE 3. Marked reduction in elastic fibers was seen within the lesion, with adjacent background solar elastotic fibers on the right (Verhoeff-van Gieson, original magnification ×100).

elastic fibers, increased numbers of fibroblasts and/or mast cells, increased (small-vessel) vascularity, focal mucin deposition/myxoid degeneration, a mild perivascular lymphocytic infiltrate, attenuation of adnexal structures, and abnormalities of the epidermis (eg, surface ulceration).[2-13]

Prior to biopsy, the differential diagnosis in our patient included angiofibroma, pyogenic granuloma, and basal cell carcinoma given the exophytic, pink, papular appearance of the lesion; however, the histopathologic differential diagnosis included angiofibroma, angiomyolipoma, lymphangioma, nevus sebaceus, and spindle cell lipoma (SCL). In angiofibroma, a dermal proliferation of stellate fibroblasts, dilated blood vessels, and collagenous stroma are seen. Cutaneous angiomyolipoma demonstrates smooth muscle bundles in addition to thickened blood vessels and variable proportions of mature adipocytes. Lymphangioma is characterized by dilated lymph channels lined by flat endothelial cells. Nevus sebaceus shows superficial immature and abnormally formed pilosebaceous units, with epidermal papillomatosis.

Rare cases of SCL on the nose have been described.[14] Similar to pedunculated lipofibroma, reported examples demonstrate mature univacuolar adipocytes with thick collagen fibers and bland uniform spindle cells. Unlike the lesion seen in our patient, nasal SCL may be clinically mobile and typically is localized to the subcutaneous tissue, although dermal tumors also occur.[14] Variably reported histopathologic findings in nasal SCL include circumscription/encapsulation, spindle cells arranged in short fascicles with nuclear palisading, a myxoid/mucinous interstitial matrix, and/or multinucleated giant cells—all light microscopic features that were not identified in our case; however, variable proportions of adipocytic, fibrous, and myxoid components among reported examples of SCL on the nose[14] can make distinction from pedunculated lipofibroma difficult, as both are benign lipomatous tumor variants.

Clinically, pedunculated lipofibroma may be confused with more common benign cutaneous lesions and must be distinguished from other fibrolipomatous lesions on the nose. Specifically, the differential diagnosis includes benign cutaneous papillomas such as acrochordon, angiofibroma, melanocytic nevi, neurofibroma, nevus sebaceus, lymphangioma, and eccrine poroma.[7-13] These all can be readily excluded on histopathology. Pedunculated lipofibroma on the nose, as in our patient, must be distinguished from fibrolipoma[15] and dendritic myxofibrolipoma.[16] Fibrolipoma is a subcutaneous proliferation of mature adipose tissue and fibrous tissue and comprises 1.6% of all facial lipomas reported worldwide.[15] Dendritic myxofibrolipoma is a recently described benign soft-tissue tumor characterized by an admixture of mature adipose tissue, spindle and stellate cells, and an abundant myxoid stroma with prominent collagenization.[16]

Treatment of pedunculated lipofibroma on the nose is not indicated except for cosmetic reasons, in which case simple surgical excision would be considered satisfactory. Following biopsy, no further treatment was pursued in our patient.

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