A papulonodule on the nasolabial fold

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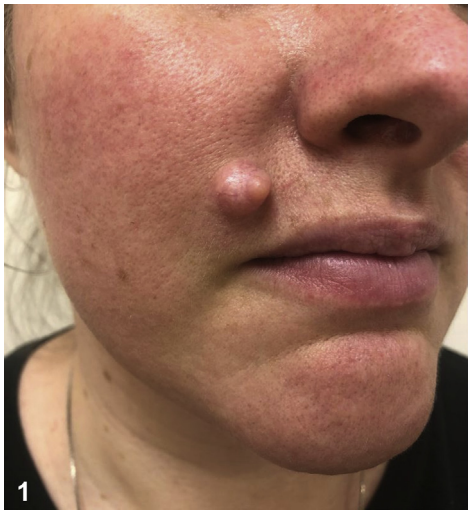

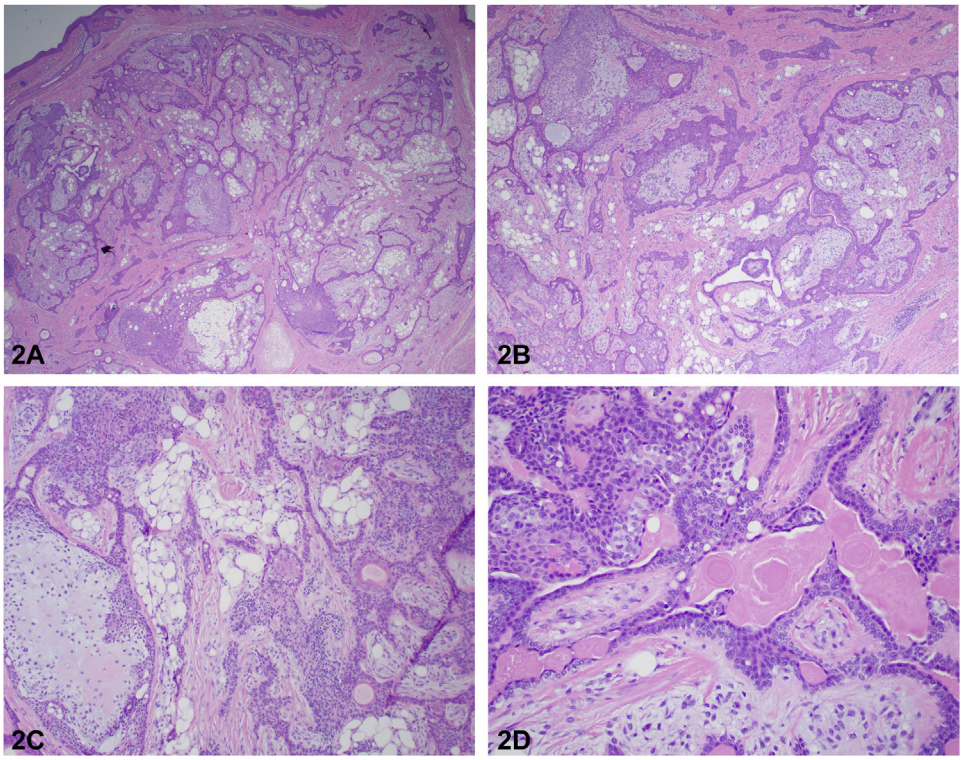

A 32-year-old woman presented with a painful papulonodule on her right nasolabial fold. She reported first noticing the lesion 9 years ago, and it had exhibited slow growth since the onset. Physical exam revealed a 1.5-cm, round, firm, skin-colored, exophytic papulonodule with overlying telangiectasias (Fig 1). A narrow margin excision was performed.

Histologically, the lesion was a multi-lobulated dermal tumor composed of islands and cords of epithelial cells and a variably cellular mesenchymal component (Fig 2, *A* and *B*). Focal chondroid differentiation and clusters of adipocytes were noted (Fig 2, *C*). The epithelial cells were basaloid with ductule formation. Neither cytologic atypia nor mitotic activity was observed (Fig 2, *D*).

Question 1: What is the most likely diagnosis?

A) Pleomorphic adenoma of the salivary gland

B) Dermatofibrosarcoma protuberans

C) Chondroid syringoma (mixed tumor)

D) Malignant chondroid syringoma

E) Hidradenoma papilliferum

Answers:

A) Pleomorphic adenoma of the salivary gland — Incorrect. This benign salivary gland neoplasm most commonly arises within the parotid gland or the major or minor salivary glands. It is typically slow-growing and painless. Histopathology demonstrates triphasic morphology composed of epithelial ductal cells, myoepithelial cells, and a chondromyxoid stroma.[1]

B) Dermatofibrosarcoma protuberans — Incorrect. This locally aggressive mesenchymal neoplasm of fibroblastic differentiation rarely presents on the face. It consists of a nodular cutaneous mass, exhibits slow growth over several years, and is characterized histologically by a dermal collection of spindle cells in a "storiform" whorled pattern. Infiltration of the subcutis in a "honey-comb" pattern is characteristic.

C) Chondroid syringoma (mixed tumor) — Correct. This benign and rare neoplasm typically presents as a slow-growing, isolated, firm, 0.5-3.0-cm, intradermal or subcutaneous nodule on the head or neck. Histology demonstrates a biphasic tumor with an epithelial component (forming islands, cords, tubuloalveolar, or ductal structures with eccrine or apocrine differentiation) and a mesenchymal component (which may contain a myxoid, chondroid, fibrous, fatty, or hyalinized matrix).[2,3]

D) Malignant chondroid syringomas — Incorrect. This malignant counterpart to chondroid syringoma tends to present on the trunk and extremities. It too contains epithelial and mesenchymal components on histopathology, but mitoses, nuclear atypia, pleomorphism, necrosis, and lymphatic invasion suggest a malignant process.[4]

E) Hidradenoma papilliferum — Incorrect. This is a rare benign adnexal neoplasm. It typically presents as a slow-growing nodule in the anogenital region of female patients. Histopathology reveals papillary fronds, apocrine ducts with decapitation secretion, and fibrous stroma.

Question 2: What are the best next steps in terms of treatment?

A) No further treatment is needed

B) Re-excision with a 4-mm margin of normal skin

C) Re-excision plus computed tomography of the head and neck

D) Mohs micrographic surgery

E) Referral to oromaxillofacial surgery

Answers:

A) No further treatment is needed — Correct. The standard treatment for chondroid syringoma is enucleation or simple excision, and the risk of recurrence is minimal.[2,3] Our patient did not receive any additional treatment after the initial excisional biopsy and remains free of disease at six months follow-up.

B) Re-excision with a 4-mm margin of normal skin — Incorrect. Re-excision with definitive margins is the recommended treatment for pleomorphic adenoma of the salivary gland, not for chondroid syringoma. Pleomorphic adenoma of the salivary gland has a tendency of local recurrence if incompletely excised as well as the potential for malignant transformation.[1]

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C) Re-excision plus computed tomography of the head and neck — Incorrect. The risk of recurrence and the metastatic potential are very low for chondroid syringomas.[2,3]

D) Mohs micrographic surgery — Incorrect. Mohs micrographic surgery is the treatment of choice for resectable cases of dermatofibrosarcoma protuberans. A benign appendageal tumor with a low risk of recurrence would not require 100% margin assessment.

E) Referral to oromaxillofacial surgery — Incorrect. While the referral to oromaxillofacial surgery may be warranted with extension of a neoplasm into the oral cavity, this well-circumscribed dermal tumor would not warrant such a referral.

Question 3: Rearrangements involving which gene are commonly found in this neoplasm?

A) *COL1A1* (collagen type I, alpha 1 chain)

B) *USP6* (ubiquitin-specific protease 6)

C) *NEMO* (NF-κB essential modulator)

D) *PLAG1* (pleomorphic adenoma gene 1)

E) *TCR*γ (T-cell receptor gamma)

Answers:

A) *COL1A1* (collagen type I, alpha 1 chain) — Incorrect. *COL1A1* forms a fusion protein with platelet-derived growth factor beta in cases of dermatofibrosarcoma protuberans.

B) *USP6* (ubiquitin-specific protease 6) — Incorrect. *USP6* rearrangement and subsequent activation are thought to play a role in the development of nodular fasciitis.

C) *NEMO* (NF-κB essential modulator) — Incorrect. Mutations in *NEMO* are thought to play a role in the development of incontinentia pigmenti.

D) *PLAG1* (pleomorphic adenoma gene 1) — Correct. PLAG1 codes for a zinc finger transcription factor. Over production via gene rearrangement has been implicated in the development of multiple neoplasms of myoepithelial origin, including cutaneous myoepitheliomas, salivary mixed tumors, and chondroid syringomas.[5]

E) *TCR*γ (T-cell receptor gamma) — Incorrect. *TCR*γ mutations and rearrangements play a role in the development of lymphoproliferative diseases, namely, T-cell lymphoma.

Conflicts of interest

None disclosed.

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