Exophytic growth on nasal ala



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A 78-year-old woman presented with a painless mass on the left nasolabial fold causing visual impairment. She noted a similar prior growth on the left temporal aspect of the scalp, which subsequently "fell off" without any treatment rendered. Past medical, social, and surgical histories were unremarkable. On exam, there was an exophytic, flesh-colored nodular growth with overlying telangiectasias on the left nasal ala (Fig 1). A maxillofacial computerized tomography scan with contrast revealed a lobular exophytic, enhancing mass, contacting subpalpebral cutaneous tissues without any evidence of invasion or lymphadenopathy (Fig 2). An excision was performed, and histologic examination is shown in Fig 3.

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Question 1: What is the best diagnosis?

- A. Basal cell carcinoma
- **B.** Cylindroma
- **C.** Adenoid cystic carcinoma
- D. Spiradenoma
- E. Trichoepithelioma

Answers:

A. Basal cell carcinoma – Incorrect. Though the tumor is shiny in appearance with telangiectasias, basal cell carcinoma typically presents as a pearly papule or nodule with a smooth surface and raised peripheral borders. Histology would show basaloid nests with characteristic peripheral palisading, cleft formation around nests, and mucinous stroma.¹

B. Cylindroma – Correct. Though patients typically present with pink-to-red rubbery nodules, cylindroma remains the best diagnosis, given the solitary facial tumor with histology demonstrating basaloid islands within the dermis and subcutis in a pattern mirroring a jigsaw puzzle.²

C. Adenoid cystic carcinoma – Incorrect. This is a slow-growing neoplasm that rarely presents in the skin as a tender subcutaneous nodule. While basaloid cells are present on histology, characteristic tubular and cribriform structures are missing in this case.³

D. Spiradenoma – Incorrect. While the patient presented with a solitary benign adnexal tumor on her head, spiradenomas are composed histologically of well-circumscribed nodules composed of basaloid cells with admixed lymphocytes and hyaline deposits. It is not uncommon to observe overlapping histologic features of spiradenoma and cylindroma in a tumor, which is sometimes termed spiradenocylindroma. However, no features of spiradenoma are seen in this tumor.⁴

E. Trichoepithelioma – Incorrect. While solitary trichoepitheliomas occur on the face, especially around the nose, they typically present as skin-colored nodules or papules. Histologic examination would reveal a basaloid tumor of follicular differentiation, composed of nests and cords of basaloid cells with peripheral palisading. The tumor typically shows a fibrotic cellular stroma, and scattered papillary mesenchymal bodies may be found.¹

Question 2: This sporadic lesion occurs most commonly in which of the following settings?

A. Familial genetic mutations

- **B.** Female gender
- **C.** Following toxic exposure
- D. Secondary to prior trauma
- E. High solar damage

Answers:

A. Familial genetic mutations – Incorrect. Though cylindromas occur within the context of Brooke-Spiegler syndrome, which is an inherited autosomal dominant disease due to mutations in the CYLD gene,⁵ patients usually present in the second or third decades of life with multiple lesions along with trichoepitheliomas, spiradenomas, and lesions with overlapping features of cylindromas and spiradenomas (so-called spiradenocylindromas). Patients with isolated, or sporadic, cylindromas do not have the underlying CYLD gene mutation.^{2,5}

B. Female gender – Correct. Cylindromas are twice as common in females as in males and typically present in adults or elderly individuals.²

C. Following toxic exposure – Incorrect. Toxic exposure is not a risk factor for the development of cylindromas.

D. Secondary to prior trauma – Incorrect. Though trauma is a common etiology for dermatofibroma formation, it has not been reported as a risk factor for cylindroma formation.²

E. High solar damage – Incorrect. Though ultraviolet light exposure is a risk factor for several skin cancers, there is no known association with cutaneous adnexal tumors such as cylindromas.

Question 3: Which of the following is the *best* treatment option for this patient?

- **A.** Electrodessication
- B. Laser destruction
- **C.** Local radiotherapy
- **D.** Observation
- E. Surgical excision

Answers:

A. Electrodessication – Incorrect. Destruction of lesions with electrodessication is an option for cosmesis improvement, though not for a lesion of this size.⁵

B. Laser destruction – Incorrect. Successful treatment with several laser modalities, including

argon, carbon dioxide, and erbium:Yag plus carbon dioxide laser therapy, have been reported.⁵ However, a lesion of this size is not ideal for laser destruction.

C. Local radiotherapy – Incorrect. Radiotherapy is typically reserved for malignant processes as adjuvant therapy to excision. While cylindromatosis is a neoplastic process, such lesions are typically benign with low risk of malignant transformation.^{2,5}

D. Observation – Incorrect. Though often cosmetically unappealing, cylindromas are often benign and carry low risk of malignant transformation.^{2,5} Thus, observation is an option for patients who do not want to undergo a procedure. However, given the patient's vision impairment, surgical excision would be the best option.

E. Surgical excision – Correct. Surgical excision can be pursued for a lesion of this size and for cosmesis. Wide margins are typically not warranted,

given the benign nature of the lesions, but abnormal presentations such as in this case may be an indication for wide local exicision.^{2,5}

Conflicts of interest

None disclosed.

REFERENCES

- 1. Kang KJ, Seo BF, Shin J, Kwon H, Byeon JH, Jung SN. Trichoepithelioma misdiagnosed as basal cell carcinoma. *J Craniofac Surg.* 2019;30(3):e197-e199. https://doi.org/10.1097/S CS.000000000005089
- Jordão C, de Magalhães TC, Cuzzi T, Ramos-e-Silva M. Cylindroma: an update. Int J Dermatol. 2015;54(3):275-278. https://doi.org/10.1111/ijd.12600
- Ramakrishnan R, Chaudhry IH, Ramdial P, et al. Primary cutaneous adenoid cystic carcinoma: a clinicopathologic and immunohistochemical study of 27 cases. *Am J Surg Pathol.* 2013;37(10):1603-1611. https://doi.org/10.1097/PAS.0b013e318 299fcac
- Tran K, DeFelice T, Robinson M, Patel R, Sanchez M. Spiradenomas. Dermatol Online J. 2012;18(12):15.
- Kim C, Kovich Ol, Dosik J. Brooke-Spiegler syndrome. *Dermatol* Online J. 2007;13(1):10.