

A case of syringocystadenoma papilliferum of eyelid with literature review

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Syringocystadenoma papilliferum can rarely affect eyelid skin. The lesion is frequently misdiagnosed as basal cell carcinoma or cyst or squamous cell carcinoma. We are presenting a case that was clinically diagnosed as basal cell carcinoma of eyelid but was later histologically diagnosed as syringocystadenoma papilliferum.

Key words: Basal cell carcinoma, eyelid, syringocystadenoma papilliferum

Syringocystadenoma papilliferum is a rare benign hamartomatous adnexal tumor. Also, this rarely affects eyelid skin. Here, we are reporting a case of syringocystadenoma papilliferum.

Case Report

A 58-year-old male presented with a lesion on the medial aspect of left lower eyelid near the punctum. He had a small elevated nodular swelling at the margin of the eyelid for the past two decades. There was a gradual increase in the size of the swelling for past 4 years, however, during past 6 months it became more visible associated with discomfort, itching, and a noticeable mass.

Physical examination revealed a solitary 2 cm × 2 cm size brownish firm fleshy growth with hyperkeratotic surface covered with crusts in some areas, involving the skin of the left lower eyelid [Fig. 1]. The lower lid margin was involved but not beyond the lash line. It did not involve the underlying deeper tissues also and there was no regional lymphadenopathy. The remainder of ocular and general physical examination was unremarkable.

The mass was excised along with a 3 mm margin of healthy skin, and the resultant skin defect was repaired with a median frontoglabellar pedicle skin flap. The final pathologic diagnosis of excision biopsy specimen confirmed syringocystadenoma papilliferum.



Figure 1: Fleshy growth with hyperkeratotic surface covered with crusts in some areas, involving the skin of the left lower eyelid

Pathologic findings

On gross examination, skin covered tissue measuring 1.3 cm × 1.1 cm × 1 cm. This showed a nodular lesion 0.7 cm in its greatest axis 0.3 cm away from the nearest peripheral surgical cut margins.

On microscopic examination, section showed a nodular lesion lined by epidermis with varying degree of papillomatosis [Fig. 2]. Cystic invaginations were seen extending from epidermis into dermis with numerous papillary projections. These were lined by two rows of cells. The luminal row consisted of columnar cells with evidence of active “decapitation” secretion and outer row of cells composed of small cuboidal cells. Plasmacytic infiltrates were seen in the papillary cores. There was no evidence of malignancy in the biopsy specimen.

Discussion

Syringocystadenoma papilliferum is a rare benign tumor that is believed to be derived from the apocrine or the eccrine sweat glands. In a recent report, immunohistochemistry proved the apocrine nature of the benign, noncystic lesion by virtue of its nuclear androgen receptor and cytoplasmic gross-cystic disease fluid protein-15 positivity, along with its smooth muscle actin-positive myoepithelial layer.^[1] This is predominantly a childhood tumor; however presentation at a later stage like our case has been described in literature.^[2] In half of those who are affected, it is present at birth, and in a further 15–30%, the tumor develops before puberty.^[3]

Syringocystadenoma papilliferum rarely affects eyelid skin. The lesion is frequently misdiagnosed as basal cell carcinoma or cyst or squamous cell carcinoma. In a series of 14 patients,

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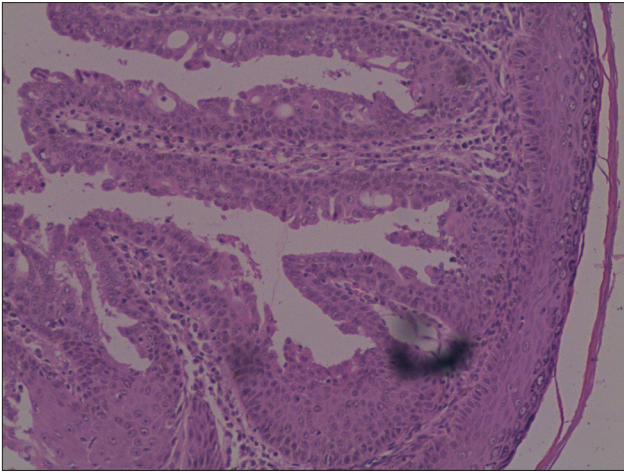


Figure 2: Papillary lesions lined by two layers of cells (H and E, $\times 10$)

most cases had a preoperative diagnosis of basal cell carcinoma or cyst. None of the lesions was associated with a malignant neoplasm.^[4] Syringocystadenoma papilliferum of the eyelid can be associated with other benign lesions. Most lesions are not clinically distinctive and require biopsy for diagnosis. This is unlike other benign eyelid lesions, where histopathological diagnosis confirms clinical diagnosis in 95.9% cases.^[5] Tumor is usually described as a skin colored to pink, hairless, firm plaque of grouped nodules or as a solitary nodule.^[6] Cauliflower like, verrucous, papillary, hyperkeratotic, or sometimes moist fleshy excrescences have also been described. Some tumors may show central umbilications.^[7] Most of the lesions develop and enlarge slowly, although a few can increase to significantly within a short period. Also, the lesion can develop ulceration and secondary infection.

The tumor has varied clinical presentations. The plaque type that presents a hairless area of the scalp is commonly associated with a sebaceous nevus of jadassohn. In about one-third of the case, syringocystadenoma papilliferum is associated with a nevus sebaceous.^[8] Appearance of the lesion in the face and neck region is seen in the linear type; however, a solitary nodular type shows predilection for the trunk. A presentation with multiple lesions is rare.

Syringocystadenocarcinoma papilliferum is a malignant counterpart of syringocystadenoma papilliferum.^[9]

The diagnosis is clinically suspected and histologically confirmed. Ulceration or a rapid enlargement of an existing tumor is indicative of a malignant transformation. We were suspicious in the index case because of the recent increase of size. In about one-tenth of cases of syringocystadenoma papilliferum, basal cell carcinoma can secondarily develop.^[6] Squamous cell carcinoma may also develop, but much less frequently. Because of this, surgical excision is the treatment of choice. In our case, excision of the tumor was done followed by repair of skin defect with median frontoglabellar pedicle skin flap. He was asymptomatic at 12 months follow-up.

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Conflicts of interest

There are no conflicts of interest.

References

1. Jakobiec FA, Rai R, Lefebvre DR. Papillary hidradenoma of the eyelid margin: Clinical and immunohistochemical observations further supporting an apocrine rather than an eccrine origin. *Surv Ophthalmol* 2014;59:540-7.
2. Rao VA, Kamath GG, Kumar A. An unusual case of syringocystadenoma papilliferum of the eyelid. *Indian J Ophthalmol* 1996;44:168-9.
3. Karg E, Korom I, Varga E, Ban G, Turi S. Congenital syringocystadenoma papilliferum. *Pediatr Dermatol* 2008;25:132-3.
4. Barbarino S, McCormick SA, Lauer SA, Milman T. Syringocystadenoma papilliferum of the eyelid. *Ophthal Plast Reconstr Surg* 2009;25:185-8.
5. Al-Faky YH. Epidemiology of benign eyelid lesions in patients presenting to a teaching hospital. *Saudi J Ophthalmol* 2012;26:211-6.
6. Rook A, Wilkinson DS, Ebling FJ. *Textbook of Dermatology*. Pondicherry: Blackwell; 1986. p. 177.
7. Helwig EB, Hackney VC. Syringadenoma papilliferum; lesions with and without naevus sebaceous and basal cell carcinoma. *AMA Arch Derm Syphilol* 1955;71:361-72.
8. Pinkus H. Life history of naevus syringadenomatosus papilliferus. *AMA Arch Derm Syphilol* 1954;69:305-22.
9. Ishida-Yamamoto A, Sato K, Wada T, Takahashi H, Iizuka H. Syringocystadenocarcinoma papilliferum: Case report and immunohistochemical comparison with its benign counterpart. *J Am Acad Dermatol* 2001;45:755-9.