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SYRINGOCYSTADENOMA PAPILLIFERUM OF THE EYELID: CASE REPORT OF A RARE BENIGN TUMOUR

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ABSTRACT Syringocystadenoma papilliferum is a rare adnexal tumour which is thought to originate from apocrine or eccrine sweat glands. Although it typically involves the head and neck, eyelid involvement is extremely rare. It usually appears at birth or during infancy, but cases have also been reported in older age groups. It can be associated with pre-existing organoid nevus sebaceous and other adnexal tumours. Taking into consideration the rarity of this lesion in eyelid, the authors' aim to understand its clinical presentation, histopathology, treatment, prognosis and to add to the existing literature.

KEYWORDS : syringocystadenoma papilliferum, benign neoplasm, eyelid tumour.

Case Report

An 80 year old woman came with the complaint of a mass growing over the lower eyelid of the left eye since 6 months. She did not complain of pain. There was no history of discharge or bleed from the mass. She gave no history of trauma. She gave no history of swellings elsewhere in the body. Her general health was good.

On examination, a pedunculated mass was present on the left lower eyelid causing mechanical ectropion. It was at the junction of the medial 1/3rd and lateral 2/3rdof the lid, involving the margin. It measured 4x3x3cm in size. The mass was pink in colour, with superficial nodular appearance. No abnormal blood vessels were seen over the mass. It was non-tender, and firm in consistency. It did not bleed on touch. The base of the mass was not indurated, and underlying tarsal conjunctiva appeared normal. There was no loss of eyelashes in the involved lid segment. She had immature senile cataract. Rest of the ocular examination was unremarkable. Bilateral pre auricular, cervical and submandibular lymph nodes were not palpable. Her systemic examination was unremarkable. Our differential diagnoses included large lid wart, granuloma or malignancy like squamous cell carcinoma. The patient underwent conservative complete excisional biopsy for the mass under local anaesthesia. Intraoperatively the mass was shaved off the base. Underlying tarsus did not look abnormal. No sutures were required. Post-operatively, antibiotic eye ointment was prescribed for use over the wound for a period of 3 weeks. Histopathological examination of sections from the mass showed keratinised stratified squamous epithelial lining, invaginated at various places forming cysts. These cysts contained numerous papillae with basal cuboidal and apical columnar cells. There was dense inflammatory infiltrate composed of lymphocytes, plasma cells, eosinophils and histiocytes along with few congested blood vessels and areas of haemorrhages; all features suggestive of syringocystadenoma papilliferum.

On subsequent follow up for 3 months, there was no recurrence and the patient observed good cosmetic recovery.

Discussion

Syringocystadenoma papilleferum (SCAP) is a rare benign hamartomatous adnexal tumour which originates from the apocrine or the eccrine sweat glands. It is relatively a rare neoplasm, predominantly a childhood tumour. It usually presents at birth or develops before puberty but there are cases reported in adult age also. (1) Although it predominantly affects the skin of head and neck, SCAP is extremely rare in the eyelid, with only few documented cases to date. (2) Other uncommon sites of involvement include leg, genital area, lower abdomen, flank, etc (3–5) J Bakshi et all have reported a first case of SCAP on the pinna. (6)

It has a varied clinical presentation. It can appear as a solitary papule or a linear arrangement of several papules. With increasing size, a more prominent papillary configuration develops, and the surface can become scabbed. (4) The surface can be smooth, flat, papillomatous, or verrucous. Increase in their sizes, crustations, nodular or verrucous transformations are noted at puberty. (5) The plaque type which presents as a hairless area on the scalp, is commonly associated with a sebaceous nevus of Jadassohn. (1)

It arises de novo or is associated with nevus sebaceous. (7) Few lesions which are reported to be associated with syringocystadenoma papilliferum include viral warts, naevus sebaceous, linear naevus verrucosus, naevus comedonicus, apocrine poroma, apocrine hidrocystoma, tubulopapillary hidradenoma, hidradenoma papilliferum, papillary eccrine adenoma, verrucous carcinoma, apocrine acrosyringeal keratosis, poroma folliculare, linear naevus verrucosa, atypical fibroxanthoma, clear cell syringoma, basal cell epithelioma, sebaceous epithelioma, trichoepithelioma and verruca vulgaris. (5) Syringocystadenocarcinoma papilliferum is a malignant counterpart of syringocystadenoma papilliferum.(1) Basal Cell Carcinoma (BCC) development has been reported in upto 10% of the cases. (8)

Histopathology

Histopathology frequently shows multiple cystic invaginations in a background of fibrous tissue. The upper regions of the cystic invaginations are commonly lined by keratinizing squamous epithelial cells which are similar to those seen on the surface epidermis, whereas the lower region contains numerous papillary projections of variable shapes and sizes, which extend into the lumina of the invaginations. These papillary projections are the most characteristic pattern. The glandular epithelium is lined by two layers of cells, namely, tall, columnar cells with oval nuclei and faintly eosinophilic cytoplasm seen at the luminal surface and small cuboidal cells with round nuclei and scanty cytoplasm seen at the base. In some areas, the cells of the luminal layer are arranged in multiple layers and they form a lace-like pattern, resulting in multiple, small, tubular lumina. Decapitation secretion or “apical snouts” are often seen at the luminal surface. Another diagnostic feature of this neoplasm is the presence of a mononuclear inflammatory infiltrate which consists mainly of plasma

cells of IgG and IgA class in the fibrous tissue of the papillary projections, along with dilated capillaries. Diastase resistant Periodic acid-Schiff (PAS) positivity favours the apocrine differentiation of this tumour. Tumour cells also may show positivity for carcinoembryonic antigen (CEA) and protein-15. (7)

Treatment

The treatment for Syringocystadenoma papilliferum is excision biopsy, histopathological examination being the gold standard to confirm the diagnosis. Mohs micrographic surgery offers the advantages of tissue preservation with a high degree of surgical margin control.(9) Carbon dioxide lasers have gained importance for treatment of lesions at difficult anatomic sites, which were not suitable for surgery. (10)

Conflicts of interest

The authors declare no conflicts of interest.

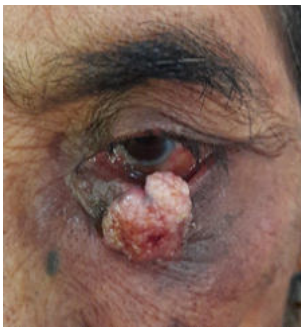

Figure 1: Left eye lower eyelid mass

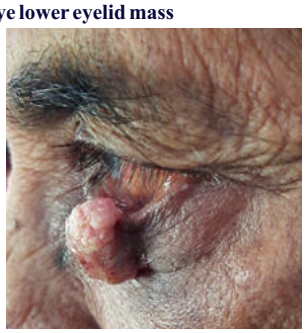

Figure 2: Lateral view

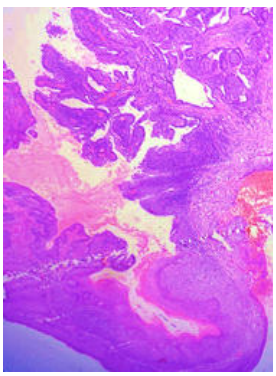

Figure 3: Histopathology showing cystic structure lined by Keratinized stratified squamous epithelium with papillary frond extending in the stroma (H&E, 4X)

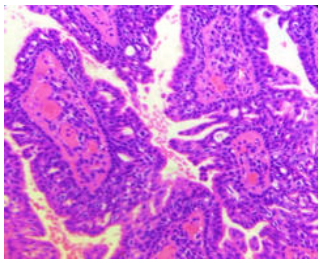

Figure 4: Histopathology showing papillae with double cell layers, inner layer is composed of columnar cells with secretions, outer layer is composed of cuboidal cells with papillary projection (H&E, 40X).

REFERENCES

1. Sangma MMB, Dasiah SD, Ramachandra Bhat V. Syringocystadenoma papilliferum of the scalp in an adult male - A case report. Journal of Clinical and Diagnostic Research. 2013;7(4):742–3.
2. Barbarino S, McCormick SA, Lauer SA, Milman T. Syringocystadenoma papilliferum of the eyelid. Ophthalmic Plast Reconstr Surg. 2009;25(3):185–8.
3. Nascimento BAM, Carneiro CMO, Carvalho AH, Bittencourt M de JS, Drago MG, Freitas LKM. Syringocystadenoma papilliferum in an unusual location. An Bras Dermatol. 2015;90(6):900–2.
4. Xu D, Bi T, Lan H, Yu W, Wang W, Cao F, et al. Syringocystadenoma papilliferum in the right lower abdomen: A case report and review of literature. Onco Targets Ther. 2013;6:233–6.
5. Agrawal R, Kumar P, Varshney R. Syringocystadenoma papilliferum: An unusual presentation. Journal of Clinical and Diagnostic Research. 2014;8(5):3–4.
6. Bakshi J, Parida PK. Syringocystadenoma papilliferum: report of first case on the pinna. J Laryngol Otol. 2006;120(12):939–42.
7. Malhotra P, Singh A, Ramesh V. Syringocystadenoma papilliferum on the thigh: An unusual location. Indian J Dermatol Venereol Leprol. 2009;75(2):170–2.
8. Elson B, Victor C. Papil iferum. 2015;
9. Chi CC, Tsai RY, Wang SH. Syringocystadenocarcinoma Papilliferum: Successfully Treated with Mohs Micrographic Surgery. Dermatologic Surgery. 2004;30(3):468–71.
10. Srivastava D, &. T. (2012). appendage tumors and hamartomas of the skin. In &. K. Goldsmith L.A., Fitzpatrick's Dermatology in General Medicine. McGraw Hill.