SOLITARY TRICHOEPITHELIOMA– A CASE REPORT

Navnoor Singh[1], Nitin Batra [2], Rupali Chopra[3]

[1]Postgraduate Student, Department of Ophthalmology, Christian Medical College and Hospital, Ludhiana, Punjab.
[2]Professor and HOD, Department of Ophthalmology, Christian Medical College and Hospital, Ludhiana, Punjab.
[3]Professor, Department of Ophthalmology, Christian Medical College and Hospital, Ludhiana, Punjab.

HOW TO CITE THIS ARTICLE: Singh N, Batra N, Chopra R. Solitary trichoepithelioma – a case report. J. Evid. Based Med. Healthc. 2018; 5(5), 468-469. DOI: 10.18410/jebmh/2018/95

PRESENTATION OF CASE

An 80-year old man presented to our outpatient department with a solitary swelling on the lateral aspect of his right lower lid. The swelling had appeared 5 years ago and was progressively increasing in size and attained the present dimension of 2.0 x 1.0 centimeters. It was raised and firm in consistency with well-defined borders. There was no evidence of ulceration or intraepithelial erosion. The conjunctiva appeared uninvolved. The patient was otherwise healthy and there was no family history of skin lesions. The visual acuity was 6/24 in both eyes. Due to its blemishing appearance, the patient wished to have the tumour removed. It was excised with a small oval incision and primarily closed.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis for the swelling included fibrofolliculoma, trichofolliculoma, trichoepithelioma, desmoplastic trichoepithelioma, trichoblastoma, trichoadenoma, pilomatrixoma, basal cell carcinoma, trichilemmal carcinoma and pilomatrix carcinoma.

PATHOLOGICAL DISCUSSION

On gross examination, the swelling measured 2.5 x 1.4 x 0.5 centimeters. The overlying epidermis was intact. The tumour was reaching upto the lateral margins and base. Microscopic examination showed two types of cells - basaloid and ovoid cells. The basaloid cells were arranged in tiny cysts, some of them showing peripheral palisading pattern. Interspread between them were seen ovoid cells arranged in solid nests and grooves. There was an increase in mitosis and nuclear pleomorphism.

Trichoepitheliomas are benign tumors arising from the wall of the hair follicle. Formerly termed epithelioma adenoids cysticum of Brooke, it is a nodular and cystic tumour that is classified as a poorly differentiated hamartoma of the hair germ. It can occur as a solitary eyelid lesion or as multiple and bilateral facial or eyelid nodules.[1] It is more commonly seen as a solitary lesion which tends to occur in older individuals as an asymptomatic, flesh coloured to yellowish, firm papule that rarely ulcerates. It is not inherited and nodule is usually less than 2 centimeters in diameter.[2] These lesions sometimes have a morphea like appearance (sclerosing trichoepithelioma) and thus often can be confused with basal cell carcinoma, except that the latter is inexorably progressive, whereas the former can attain a relatively stationary character.[3]

Trichoepitheliomas occur in three varieties: solitary, multiple and desmoplastic.[4-6] The solitary and multiple forms share a common appearance on histology and are traditionally differentiated by their clinical picture.[4] Solitary trichoepitheliomas appear as isolated lesions and are not inherited. Multiple trichoepitheliomas are inherited in an autosomal dominant pattern and appear in groups of lesions. Multiple trichoepitheliomas can occur alone as multiple familial trichoepithelioma, or in association with cylindromas and syringomas as Spiegler-Brooke syndrome.[7] Our case is an example of solitary Trichoepithelioma, determined on the basis of the single eyelid lesion, negative family history and classical histopathological findings.

In addition to phenotypic differences, recent chromosomal studies show that multiple and solitary trichoepitheliomas differ genotypically. Multiple familial Trichoepitheliomas are associated with chromosome 9p21, but reports have found mutation in cylindromatosis tumor suppressor gene, found on chromosome 16q12q13 in most of cases.[7] Cases of solitary trichoepithelioma have not been linked to chromosome 9p21, but also one half of studied lesions show deletions on chromosome 9q22.3 at the patched gene that also has the potential to cause hereditary basal cell carcinoma.[8]

The third form of trichoepithelioma, the desmoplastic variant, differs from the solitary and multiple variants in gross appearance and on histology. Desmoplastic trichoepitheliomas have raised borders and non-ulcerating centers. Milia-like lesions sometimes surround them. Inheritance is uncommon, and there are no known specific genetic loci.[5]

This case also demonstrates an uncommon location, the eyelid, for a solitary trichoepithelioma. Aurora reported the first case of solitary eyelid Trichoepithelioma in 1974, after previously finding no cases in a series of 678 benign eyelid lesions.[6] Similarly, Gray and Helwig found only one case of solitary trichoepithelioma near the inner canthus of the right lower lid in 83 solitary trichoepitheliomas.[4] Simpson at al presented the largest study of eyelid trichoepitheliomas in 18 patients over a duration of more than 30 years. Males were found to be twice more predisposed than females. The upper lid was found to be twice more predisposed as

Financial or Other, Competing Interest: None.
Submission 03-01-2018, Peer Review 06-01-2018,
Acceptance 20-01-2018, Published 29-01-2018.
Corresponding Author:
Dr. Navnoor Singh,
Room No. 1012, Junior Doctor Hostel, Christian Medical College, Ludhiana, Punjab.
E-mail: navnoor.singh@gmail.com
DOI: 10.18410/jebmh/2018/95

compared to lower lids. All patients underwent excision however, lesion recurred in two of them after 10 and 12 years, respectively.[9] Ozdal et al found out that of the 228 benign adnexal tumours, 182 were diagnosed as apocrine or eccrine hydrocystoma (79.8%), 12 pilomatrixoma (5.3%), 12 syringoma (5.3%), 11 trichilemmoma (4.8%), 5 syringocystadenoma papilliferum (2.2%), 3 trichoepithelioma (1.3%) and 3 trichofolliculoma (1.3%).[10]

Basal cell carcinoma can be clinically and histologically difficult to differentiate from solitary trichoepithelioma. The trichoepitheliomas feature numerous keratinous cysts, lacy pattern of tumor cells, dense stroma, minimal inflammation, absent of minimum of abnormal hair follicles. As both tumours are basal keratin cysts and in some cases with no clear demarcation between them, it necessitates a clinical revaluation.[2] The presence of ulceration makes the clinician to error toward the diagnosis of either a BCC or sebaceous carcinoma. The differentiation of these tumours is possibly based on clinical examination and histological findings.[4]

Other benign tumours originating from the hair follicle are: (1) trichofolliculoma that has a central crater which emerges from white hairs; (2) trichilemmoma, a relatively commom verrucous tumour originating from follicular infundibula; (3) pilomatrixomas showing dark cells on histopatholgy. The last two tumours show no histological lobular architecture or manner of branching, such as Trichoepithelioma.[11]

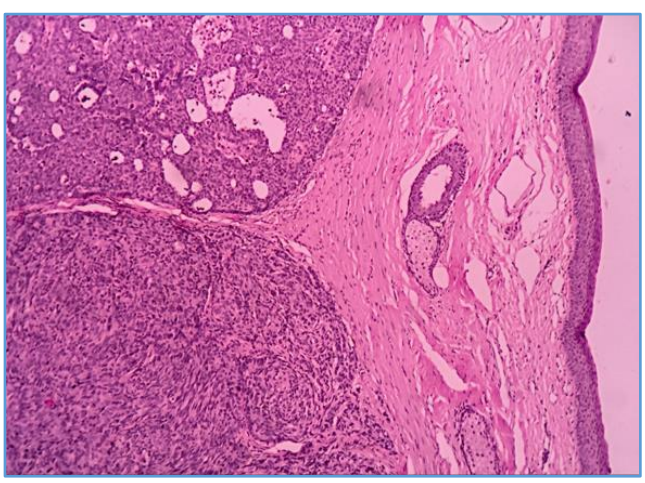

Figure 1. Showing Well Circumscribed Tumor with Intact Stratified Squamous Epithelium and Ovoid Cells Arranged in Solid Nests and Grooves

DISCUSSION OF MANAGEMENT

Most hair follicle tumors are benign in nature and can be removed with complete surgical excision. It is important to make a correct diagnosis after the excision especially in cases of malignant tumors. Supericial biopsy of the tumor is not recommended as it may not lead to a correct diagnosis. Deep and peripheral surgical resection of margins should be performed to assure complete excision of tumor. Patients with malignant tumors be followed up regularly for any recurrence or distant metastasis.

FINAL DIAGNOSIS

A diagnosis of solitary trichoepithelioma of the right lower lid was made for this patient. Trichoepithelioma is a nodular or cystic tumour that is classified as a poorly differentiated hamartoma of the hair germ. Histopathological examination is important to differentiate it from other tumours of the lid margin and helps in the treatment and prognosis of the patient.

REFERENCES

[1] Kirzhner M, Jakobiec FA, Borodic G. Desmoplastic trichoepithelioma: report of a unique periocular case. Ophthal Plast Reconstr Surg 2012;28:e121-e123.
[2] Zeligman I. Solitary trichoepithelioma. Arch Dermatol 1960;82:35-40.
[3] Mansour AM, Hidayat AA. Metastatic eyelid disease. Ophthalmology 1987;94(6):667-670.
[4] Gray HR, Helwig EB. Epithelioma adenoides cysticum and solitary trichoepithelioma. Arch Dermatol 1963;87:102-114.
[5] Carter JJ, Kaur MR, Hargitai B, et al. Congenital desmoplastic trichoepithelioma. Clin Exp Dermatol 2007;32(5):552-554.
[6] Aurora AL. Solitary trichoepithelioma of the eyelid. Indian J Ophthalmol 1974;22(2):32-33.
[7] Salhi A, Bornholdt D, Oeffner F, et al. Multiple familial trichoepithelioma caused by mutations in the cylindromatosis tumour suppressor gene. Cancer Res 2004;64(15):5113-5117.
[8] Matt D, Xin H, Vortmeyer AO, et al. Sporadic trichoepithelioma demonstrates deletions at 9q22.3. Arch Dermatol 2000;136(5):657-660.
[9] Simpson W, Garner A, Collin JR. Benign hair-follicle derived tumours in the differential diagnosis of basal cell carcinoma of the eyelids: a clinicopathological comparison. Br J Ophthalmol 1989;73(5):347-353.
[10] Ozdal PC, Callejo SA, Codere F, et al. Benign ocular adnexal tumours of apocrine, eccrine or hair follicle origin. Can J Ophthalmol 2003;38(5):357-363.
[11] Grzesinska MC. Two cases of trichoepithelioma. Post Dermatol Alergol 2006;23(4):198-202.