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Concurrent Conjunctival Occular Surface Squamous Neoplasia (OSSN) and Primary Acquired Melanosis (PAM) with atypia -Report of a rare occurrence

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Abstract

Ocular Surface Squamous Neoplasia and Primary Acquired Melanosis are well known conjunctival lesions. However, their concurrence in a single focus has not been reported in the conjunctiva so far. We report one such rare occurrence in 40-year-old male presenting with fleshy, sessile, painless, pigmented lesion over bulbar conjunctiva of right eye. Histopathological examination of the excision biopsy revealed focal severe dysplasia in the conjunctival epithelium along with melanocytic hyperplasia in the basal layer with atypical melanocytes infiltrating the full thickness of epithelium, suggestive of concurrent Ocular Surface Squamous Neoplasia and Primary Acquired Melanosis with atypia.

Introduction

Ocular Surface Squamous Neoplasia (OSSN) OSSN is an encompassing term for pre-cancerous and cancerous epithelial lesions of the conjunctiva and cornea. It includes the spectrum of Dysplasia, Carcinoma in-situ (CIS) and Invasive SCC. Primary Acquired Melanosis (PAM) with atypia is a premalignant lesion of conjunctiva with high risk of progression to malignant melanoma respectively. Concurrent occurrence of the two conditions in a single focus of the conjunctiva has not been reported so far. We report the first case of such rare coexistence in the conjunctiva of a 40 years old male.

Case report

A 40-year-old male presented to ophthalmology OPD with a fleshy, sessile, painless, pigmented lesion over bulbar conjunctiva of right eye. The lesion was clinically diagnosed as nevus which was excised and sent to us for histopathological examination. Microscopic examination revealed fragments lined stratified squamous epithelium with interspersed mucous cells, but focally revealing increased thickness. In this focus thickened lining epithelium comprised of dual population of cells. (Figure 1) First population of cells comprised of keratinocytes revealing full thickness moderate to marked pleomorphism, nuclear atypia, mitotic figures at all levels

and focal dyskeratosis with preserved surface maturation. In addition, there was a second population of cells comprising of melanocytes with increased pigmentation of the basal layer and single and small clusters of melanocytes showing mild atypia reaching up to the superficial layer. (Figure 2) All these changes were confined to the conjunctival epithelium with no invasion in to the underlying stroma. The resection margins were also free. The histological findings were of Occular Surface Squamous Neoplasia (OSSN) and Primary Acquired Melanosis (PAM) with atypia.

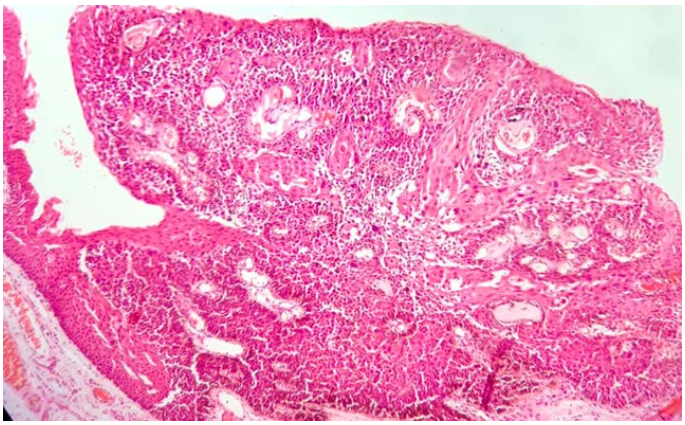

Figure 1: Focally hyperplastic conjunctival lining epithelium showing changes of both OSSN and PAM with atypia (H&E, 10x)

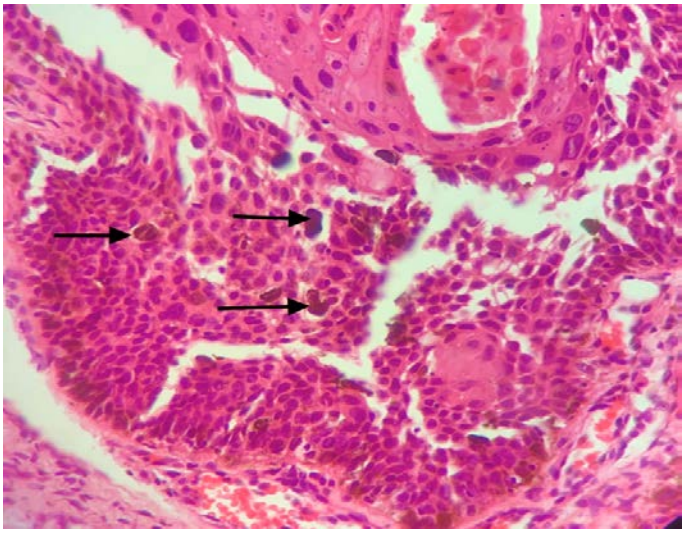

Figure 2: Higher magnification revealing atypical keratinocytes, melanocytes hyperplasia in basal layer and single atypical melanocytes (arrows) invading the conjunctival epithelium (H&E, 40x)

Discussion

Ocular surface squamous neoplasia (OSSN) comprises of a wide spectrum of corneal as well as conjunctival lesions ranging from mild dysplasia at one end of the spectrum and invasive squamous cell carcinoma on the other end. OSSN is predominantly seen in adults ranging in age from 4-96 years with an average age of presentation of 56 years.[1] Advanced age, male gender, exposure to solar ultraviolet radiation, infection with human papilloma virus (HPV), immunosuppression and infection with human immunodeficiency virus (HIV) are considered as risk actors which play an important role in the development of OSSN. It usually presents as a sessile, fleshy, elevated lesion adjacent to the limbus in the inter-palpebral region. The thickness of the lesion is not always an indication of invasive SCC. Even reasonably thick tumours tend to be confined within the epithelium. The presentation of CIN and invasive SCC is very similar thus making clinical differentiation difficult.[2]

Histology is the gold standard for the confirmation of suspected OSSN.[3] OSSN is classified depending upon the depth of dysplasia and integrity of the conjunctival basement membrane. Lesions limited to the epithelium are classified as CIN I–CIS, whereas tumours are classified as invasive when the basement membrane has been breached by nests of dysplastic cells. Histology typically shows thickened and disorganized epithelium with the cells having a high nuclear: cytoplasmic ratio with hyperchromatic nuclei. and mitotic figures above the basal epithelium. There is sometimes an abrupt transition between the normal and dysplastic epithelia. Surgery plus MMC(Mitomycin) (0.04%) 4 cycles, is the

most effective mode of treatment in these neoplasia[4]. The risk of recurrence depends on the involvement of surgical margins as well as on the histological grade, corneal location and size. After surgery carcinoma in situ can recur in >50% of patients even years later.[5] Recurrence rates are reported after topical chemotherapy.

Primary acquired melanosis (PAM) is a pigmented lesion of the conjunctiva that is flat, painless and non-cystic. PAM usually occurs unilaterally in 6th decade of life. And is more likely to occur in lightly pigmented individuals. PAM represents 11% of all conjunctival tumors and 21% of all conjunctival melanocytic lesions [6]

Primary Acquired Melanosis (PAM) is histopathologic ally divided into PAM with or without atypia. PAM without atypia is defined as pigmentation of the conjunctival epithelium with or without benign melanocytic hyperplasia. PAM with atypia is characterized by the presence of atypical melanocytic hyperplasia. Mild atypia is defined as atypical melanocytes confined to the basal layer of the epithelium; severe atypia is defined as atypical melanocytes that extend into the superficial non-basal portion of the epithelium and may contain epithelioid cells.[7] Atypia is determined by cytological features and growth patterns that are associated with malignant potential. Four types of atypical melanocytes include the small polyhedral cells, epithelioid cells, dendritic cells, and spindle cells. Polyhedral cells contain small, round nuclei with little cytoplasm. Epithelioid cells contain abundant eosinophilic cytoplasm. Spindle cells are aligned such that the long axis are parallel to the basement membrane. Dendritic cells are large cells with complex branching dendrites found along the basilar layer.[8]

PAM with atypia has a high chance of progression into melanoma while PAM without atypia has little chance of progressing to melanoma.[6] Biopsy and histopathologic examination allow determination of the presence or absence of atypia. Because PAM with atypia has significant risk of progression into melanoma, a potentially lethal tumor, surgical and medical intervention is warranted. Infiltration of tumor base/ tumor margins, scleral infiltration, intraocular and orbital tumor extension are considered as high-risk factors for OSSN [9]

The mainstay of treatment of PAM with atypia is wide excision of the lesion and cryotherapy of the borders of the lesions. Amniotic graft can be applied to the surgical site to facilitate healing. Topical chemotherapy, most commonly with mitomycin-c, can be used as adjuvant therapy in cases of diffuse lesion, positive surgical margins, large lesions that cannot be completely removed, or in cases of recurrent disease.[6] Interferon a-2b has shown promise as a medical management. A six-week trial showed that successive application of interferon a-2b resulted in shrinking of PAM. [10]

In our case the lesion was focal and confined to superficial conjunctival lining epithelium. There was no invasion in to sclera or deeper tissues and the resection margins were also free from tumor. Surgical site showed good healing and the patient has not developed any recurrence 6 months following incision.

In conclusion we report a rare concurrent occurrence of OSSN and PAM with atypia in a single conjunctival focus. Awareness regarding these entities are important as there is high risk of progression in to malignancy and patient should be kept under careful follow up.

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