A rare case of Ocular Surface Squamous Carcinoma

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Abstract

Purpose

This article presents a retrospective clinical case of an invasive keratinized squamous cell conjunctival carcinoma and the therapeutic attitude. A male patient, who has at his left eye a rapidly onset of a conjunctival tumor localized at temporal limbus, white colored, with feeder vessel, fixed with corneal invasion which causes ocular discomfort, epiphora and lagophthalmia.

Results

After ophthalmological investigation, the patient was successfully surgically treated.

Histopathological exam of the operatory piece reveals the final diagnosis of conjunctival squamous cell-carcinoma moderate differentiated. The short-term prognosis is favorable, but the long-term prognosis could be severe due to the high possibility of tumoral recurrence.

Conclusion

The histopathological exam was the gold standard for the positive diagnosis.

Keywords: conjunctival tumour, ocular surface squamous carcinoma, corneal extension

1. Introduction

Ocular surface squamous neoplasia includes a large group of pathological entities caused by dysplastic lesions of the conjunctival squamous epithelium. Ocular surface squamous carcinoma incidence is considered to be 0.13-1.9/100 000. The risk factors include ultraviolet light exposure – more frequently the tumor develops in equatorial region of the world at patients with light skin pigmentation. It is also associated with the mutations of a suppressor gene – p. 53, deficiency in DNA repair, HPV infection the subtypes 16, 18 and immunosuppression – HIV infection. [1, 2, 3].

Case report

A 78-year-old male from the country side was admitted in our department for a tumor in the left eye, developing near the inferior-temporal limbus associated with epiphora, lagophthalmia and ocular discomfort.

The patient's past medical history was an abdominal surgery for a hepatic cyst performed with an 7^{th} and 8^{th} segment liver excision, chronic obstructive pulmonary disease, chronic cardiac insufficiency with mitral valve insufficiency and arterial hypertension without any chronic treatment. The patient had no ocular history of trauma or other ocular disease in the left eye.

At the time of admission, his left eye presented a white tumor localized near the inferior – temporal limbus measuring 8/6/4 mm, fixed to the tissue, with feeder vessels and corneal extension causing epiphora, lagophthalmia and ocular discomfort. The mass lesion has been developing 4 weeks before the hospital admission.

Physical examination was normal, except a blood pressure value of 160/80 mmHg and abnormal heart murmurs due to the mitral regurgitation.

The ocular examination revealed a best corrected visual acuity (BCVA) of 0.4 OU with 4 spherical diopters correction for the right eye (RE) and 0,6 OU for the left eye (LE) with 2 spherical diopters correction. The intraocular pressure (IOP) for RE was 16 mmHg measured by applanation tonometry and for the LE, the IOP was appreciated digital to be normotonic. The refraction for the RE was + 5.25sf/- 0.50 cyl x100° D and for the LE +2,50Dsf/0,25 Cyl x26° D.

The slit lamp examination of the anterior segment for the both eyes highlighted cortical lens opacities and for the LE the conjunctival tumor which was located juxtalimbic extending from 3:00-5:00 hours with approximately dimensions: 8/6/4 mm, elevated shape, irregular surface, white appearance, fixed to the tissue beneath, vascular core, with temporal corneal extension about 3 mm. (Fig. 1)

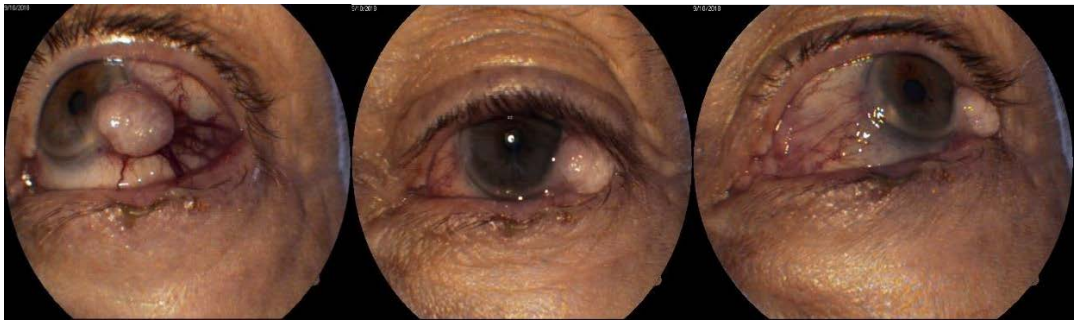

Fig. 1. Conjunctival tumor at LE

The ophthalmoscopy was performed for both eyes after the pharmaceutical mydriasis with 1% tropicamide and 10 % phenylephrine ophthalmic solutions. For the both eyes the optic nerve disc was well delimited with physiological excavation, retinal angiosclerosis and perimacular drusen at RE. (Fig. 2, Fig. 3)

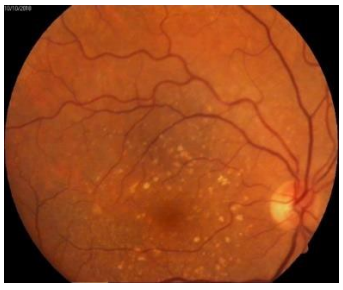

Fig. 2. Right eye fundus photography

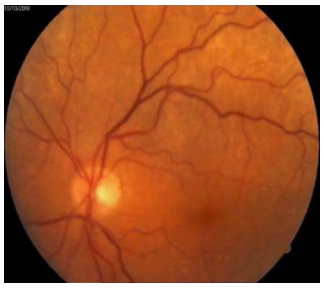

Fig. 3. Left eye fundus photography

The macular optical coherence tomography (OCT) highlighted in the RE multiple hyperreflective nodules present in the retinal pigmentary epithelium. (Fig. 4)

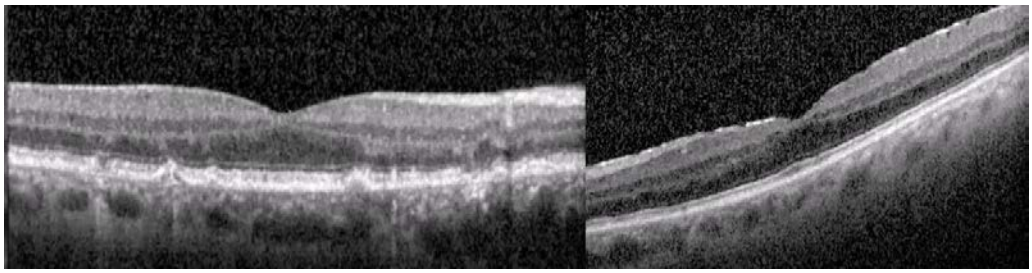

Fig. 4. Right eye and left eye – Macular optic coherence tomography

In order to detect any intraocular invasion of the conjunctival tumor, an ocular ultrasound examination for the LE was performed, but no pathological changes were discovered. (Fig. 5)

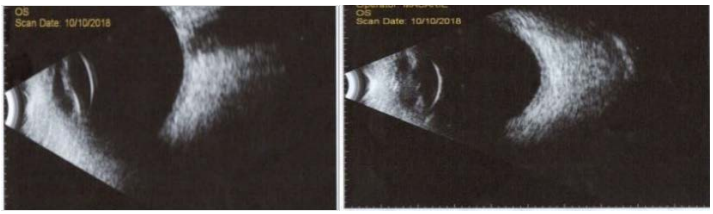

Fig. 5. Left eye ultrasonography

The blood tests had no pathological changes (complete blood count, erythrocyte sedimentation rate, biochemistry tests and HIV screening test).

After the clinical and auxiliary examinations, we determined that the patient had a LE conjunctival tumor with low hyperopia, and on the RE moderate hyperopia with age related macular degeneration. Both eyes had age related cortical cataract.

The treatment objectives were to remove the tumor in the LE and to repair the ocular surface during the same operation. The anesthetic substance injected retrobulbar was 2% lidocaine - 4 ml.

The surgical steps included tumor removal with diathermal hemostasis and finally the remaining wound was closed with a conjunctival autograph flap from the superior conjunctival sector with a 7.0 Vycril resorbable suture (Fig. 6)

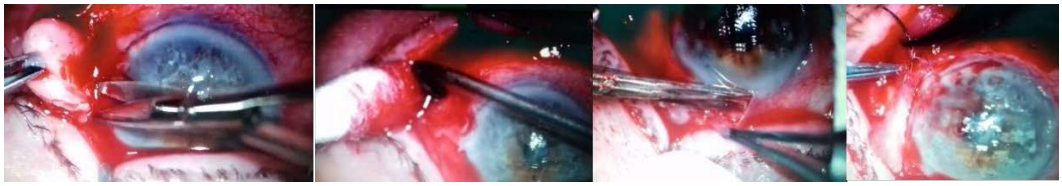

Fig. 6. Surgical steps: tumor removal, hemostasis, preparing the conjunctival flap and suture of the wound

The excised piece was histopathology examined. Macroscopically, the histopathological piece was 0,6/0,5/0,3 cm size, white and solid. Microscopically exam (Hematoxiline/Eosine) revealed stratified epithelial squamous conjunctival cell proliferation (abnormal mitosis) with intercellular bridges with focal dyskeratosis disposed in nests typical for squamous cell carcinoma – keratin pearls, in stromal layer infiltrating lymphocytes. (Fig. 7)

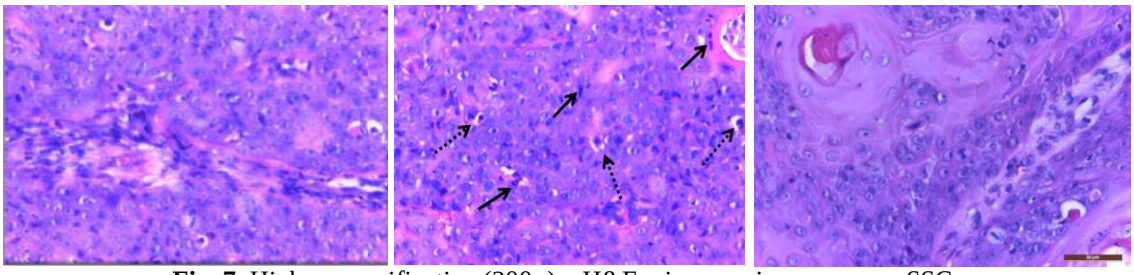

Fig. 7. Higher magnification (200μ) – H&E microscopic appearance SSC:
mitosis and apoptotic bodies (marked with⋯▶) – focal dyskeratosis – "keratin pearls"
(marked with⟶)

Taking into account the histopathological exam the positive diagnosis for the LE was an invasive conjunctival keratinized squamous cell carcinoma, moderately differentiated (G2).

For the differential diagnosis, the following were taken into account: the benign tumors (conjunctival papilloma, naevus, hemangioma, herniated orbital fat, xanthogranulloma, benign intraepithelial dyskeratosis), and also the conjunctival malign tumors like the pigmented or non-pigmented melanoma, lymphoma, Kaposi sarcoma. All these pathologies were excluded by the histopathological examination.

After surgery the patient followed a local treatment with Netilmycin and Dexamethasone 0.1%, five times per day for 4 weeks.

At 1 month at follow-up, the surgical wound was healed. (Fig. 7)

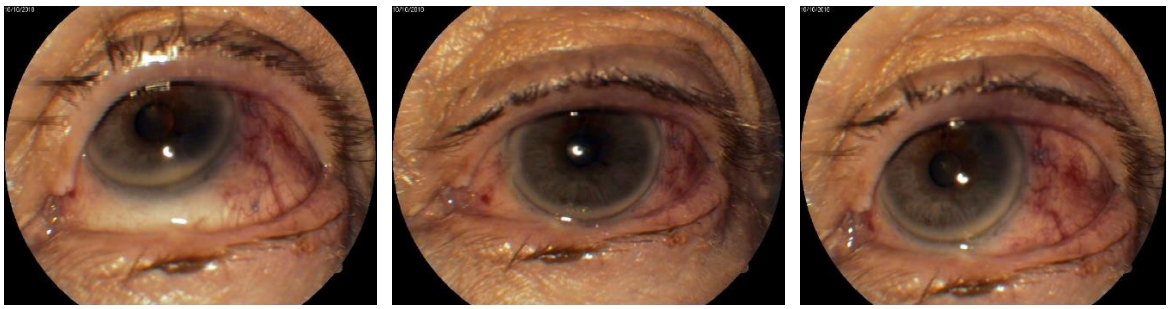

Fig. 7. Left eye – after surgery at 1 month

An anterior segment OCT was performed, showing a normal aspect of the cornea and conjunctiva of the left eye.

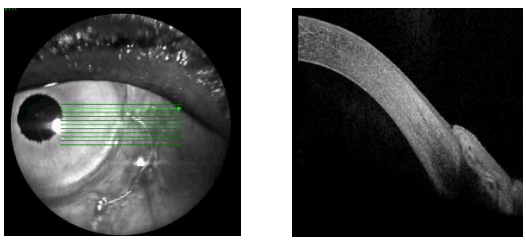

Fig. 8. Left eye anterior segment optical coherence tomography

The short-term prognosis for this case is good due to the clinical aspect in the follow-up examination, but the long-term prognosis is severe due to a high possibility of tumoral recurrence, because of the tumoral dimension and the aggressive corneal invasion. The particularities of this

are the fast progression of the tumor and also by the rare type of the conjunctival tumor (squamous cell carcinoma).

Discussions

The squamous cell carcinoma is included in the group of ocular surface squamous neoplasia and is a dysplastic lesion (DNA mutation) of the squamous epithelium of the conjunctiva.

The malignant property of this tumor is proportional with the infiltration. Commonly the tumor cells invade through the epithelial basement membrane. [5, 6] This type of tumor is classified by the degree of the epithelial and stromal infiltration. [2, 3, 5, 6] (Table 1)

Benign OSSN	Preinvasive OSSN	Invasive OSSN
Papilloma	Conjunctival/ Cornea intraepithelial neoplasms grades I- III	Squamous
Pseudotheliomatous hyperplasia		Mucoepidermoid
Benign hereditary intraepithelial dyskeratosis		

Table 1. Types of ocular surface squamous neoplasia classified by the malignant property

The ocular surface squamous carcinoma develops more frequently in male patients between 50-75 years, who are exposed to UV-B, chronic irritants like petroleum product exposure, smoking, or patients who have vitamin A deficiency, immunosuppression, xeroderma pigmentosum, HPV infection, HIV infection – in this case the risk of developing conjunctival malignant tumors increases 13 folds. In order to exclude HIV infection at this patient, the screening test was conducted, but it was negative. [3, 4] The majority of lesions develop in the nasal part of the bulbar conjunctiva, but in this case the conjunctival tumour was localized in the temporal part of the conjunctiva with corneal extension. The histopathologic exam is gold standard for establishing the diagnosis of any intraorbital or ocular tumour. [7]

Depending on the corneal extension, the recurrence of this type of tumours should be considered. The current literature considers the risk of relapse to be 12.9 %, and this figure could increase if the corneal extension exceeds 2 mm and the diameter of the tumour is greater than 5 mm. For this patient the diameter of the tumour was 6 mm and the corneal extension was 3 mm, for these reasons the patient is re-examined every 3 months. [8, 9, 10]

The recommended treatment is a complete excision of the tumour coupled with adjuvant therapy like cryotherapy, mitomycin, interferon α 2B applied during the surgery on the edge of the excision in order to induce an antiproliferative effect. The remaining wound is recommended to be closed using conjunctival autograph plasty in order to help the restoration of the ocular surface. For this reason, in this case a conjunctival autograph was used, the conjunctival flap was taken from the superior bulbar conjunctiva and sutured to the temporal defect. Moreover, this type of ocular surface carcinoma could develop secondary from tumors localized in the brain or sinus mucous membrane. [8, 9]

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responsibility for the integrity of the work as a whole, and have given final approval to the version to be published.

Disclosures

Stefanut Claudia,Scorobet Corina and Crisan Doinita have nothing to disclose.

Compliance with Ethics Guidelines

All procedures were followed in accordance with the responsible committee on human experimentation and with the Helsinki Declaration of 1975 and subsequent revisions. Informed consent was received from the patient involved in this case report.

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