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Case Report

Conjunctival Melanoma in a Patient of Dark Race

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Abstract

Conjunctival melanoma is a somewhat rare condition, relative to choroidal melanoma and just about 500 times less often than cutaneous melanoma. Data obtained from several reports revealed that the more incidence of conjunctival melanoma is among white population with rare reports among others. Our case is 65-year-old black race, presented with deep brownish to blackish separated areas in his left eye of four-year duration that increased in size during the last two months as the patient noticed. Hence, our presenting case is of great importance and its significance comes from both its occurrence in a dark race patient which is very rare relative to that of white race and also, its development on top benign looking melanocytic lesion. Ultrasonographic biomicroscopy (UBM) was performed which revealed that the thickness of tumor is 1.4 mm, and confirmed the sclera origin of the tumor and not from an extrascleral extension of a ciliary body melanoma. Histopathology revealed malignant conjunctival melanoma associated with primary melanosis with atypia. No metastatic deposits were present as revealed from results obtained from multiple investigation tools as histopathological examination of lymph nodes, bone scan, and computed tomography of the head, neck, chest, and abdomen. Surgical removal of the tumor, with applying cryotherapy to the normal-appearing conjunctiva surrounding the lesion was performed followed by adjuvant treatment in the form of chemotherapy and topical mitomycin. After the two-year of follow-up, the patient has no evidence of recurrence which was evident from the regular periodic investigations hat reflected on good general condition of our patient.

Keywords: Conjunctiva, Melanoma, Ocular tumor, Dark race

INTRODUCTION

Conjunctival melanoma is a somewhat rare condition, relative to choroidal melanoma and just about 500 times less often than cutaneous melanoma. Its incidence is 0.2 to 0.8 per million in Caucasian (Seregard, 1998; Jakobiec, 1980). Data obtained from several reports revealed that the more incidence of conjunctival melanoma is among white population with rare reports among others (Jakobiec, 1980; Yu et al., 2003). It is a potentially lethal neoplasm, with an average 10-year mortality rate of 30% (Brownstein et al, 1979). No sexual predilection was noted for conjunctival melanoma, and found mainly in the 4th to 7th decade of life, despite rare cases recorded in children (Tuomaala and Kivela, 2003; Paridaens et al., 1994; McDonnell et al., 1989). The perilimbal interpalpebral bulbar conjunctiva is the favorite site for conjunctival melanoma to develop (McDonnell et al., 1989). Up to date studies have pointed toward that like cutaneous melanoma, the incidence of conjunctival melanoma is increasing (Stratus, 2003).

Case presentation

A 65-year-old black man presented with deep brownish to blackish coalescent areas in his left eye of four-year duration that increased in size during the last two months as the patient noticed (Fig 1 A). On taking detailed history, this pigmented area started as light brownish

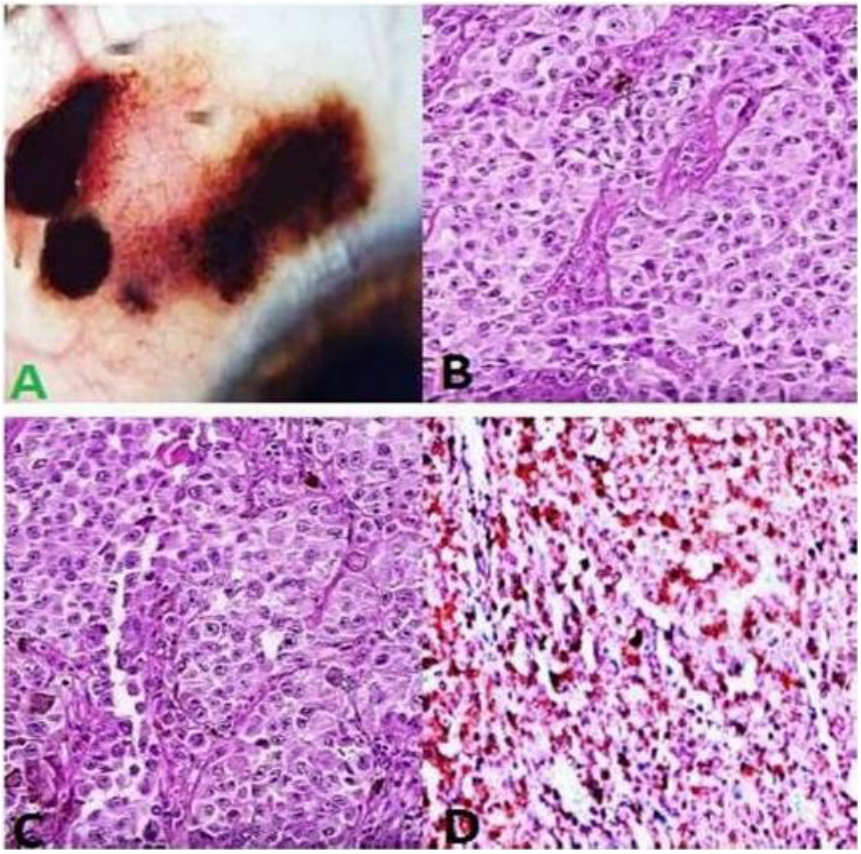

Figure 1. Multiple coalescent brownish to blackish slightly raised lesions are seen in (Fig A); Nests of atypical melanocytes extending down without normal maturation with multiple bizarre nuclei are seen. Some melanin pigment is seen in (Fig B) and (Fig C); the melanocytes show marked positive S-100 immunostain in (Fig D)

coalesce together in the last two months, not associated with pain or other symptoms. The patient gives history of diabetes for long time with history of trauma five years ago due to a blow on his left eye and the hemorrhagic brownish dots all over the conjunctiva were developed which underwent gradual improvement and the hemorrhagic dots disappeared completely after six months without any sequels to eye. No past history of ocular surgery or visual changes. Physical examination revealed blackish conjunctival raised lesions extending onto the peripheral limbus, and fixed to the underlying sclera. Tenderness of ipsilateral preauricular lymph node is marked, no palpale submandibular or cervical lymph nodes is present. Ear nose and throat (ENT) examination revealed otitis externa in the left side. Other systems are clinically free.

Ultrasonographic biomicroscopy (UBM) was performed which revealed that the thickness of tumor is 1.4 mm and confirmed the sclera origin of the tumor and the extrascleral extension of a ciliary body melanoma was completely excluded in which, in some instances, resembles a conjunctival melanoma in some cases.

Conjunctival excision biopsy was taken and sent for histopathalogical examination which revealed an initial diagnosis of malignant melanoma. Several routine preoperative investigations were done and the patient was subjected for an excisional scleral keratectomy followed by cryotherapy and alcohol application. The excised tissue was sent for histopathological examination (Fig 1 B, C) which revealed malignant conjunctival melanoma associated with primary melanosis with atypia, and the diagnosis was confirmed by strong positivity of the tumor to S100 immunostain (Fig 1D)

Surgical removal of the tumor, with applying cryotherapy to the normal-appearing conjunctiva surrounding the lesion was performed followed by adjuvant treatment in the form of chemotherapy and topical mitomycin. No metastatic deposits were present as revealed from results obtained from multiple investigation tools as histopathological examination of lymph nodes, bone scan, and computed tomography of the head, neck, chest, and abdomen. After the 2 years of follow-up, the patient has no evidence of recurrence which was evident from the regular periodic investigations which was evident by the good general condition of our patient.

DISCUSSION

The development of conjunctival melanoma in dark race man is extremely rare as reported in many studies done (Seregard, 1998; Jakobiec, 1980), this sequentially, reflects in our presenting case and renders it to be of great significance and therefore, the case must be highlighted and reported.

Conjunctival melanoma can happen in any conjunctival regions but the most common site affected is the perilimbal bulbar conjunctiva (Seregard, 1998). It arises from three clinical and histopathological settings; in concurrence with a nevus, de novo, and Primary acquired melanosis (PAM) with atypia (Paridaens et al., 1994a,b; Ho et al., 2007). Melanoma cells are larger than nevus cells and grow as individual cells or as irregular nests forming nodules. Characteristically, these cells have large nuclei with prominent eosinophilic nucleoli, and clumping of chromatin at the nuclear membrane. The presence of atypical melanocytes at the surface of the conjunctival epithelium is exclusive of malignancy (Esmaeli et al., 2012).

Not all conjunctival melanomas are pigmented; melanomas have no pigment can bear a resemblance to sebaceous gland and squamous cell carcinomas, lymphoid hyperplasia, papillomas, and even pterygia. Immuunohistochemistry staining using either the S-100 protein stain or homatropine methylbromide (HMB-45) antibody stain can support in diagnosis (Heegaard et al., 2000).

Accordingly, the tumor may arise on top of a benign nevus. In such cases, malignant melanoma is suspected when there are some morphologic changes occurring in the nevus as changes in shape, size, and color either lighting or darkness (Brownstein et al., 1979). Conversely, the vast majority of conjunctival nevi, do not progress to malignant melanoma. On the other hand, the conjunctival melanomas that develop de novo are very rare and have been reported in about 12% of patients. Area of PAM with atypia has a great risk for developing invasive malignant melanoma; it is encountered in about 75% in the pathogenesis and development of malignant melanoma. Thickening of the conjunctiva that harbor the area of PAM alleviate the diagnosis of the malignant melanoma in such cases (Folberg, 1996; Folberg et al., 1985a; Folberg et al., 1985b).

The corneal epithelium is commonly concerned when PAM with atypia is present in the limbal region (Tuomaala et al., 2002). Bowman's membrane come into view to afford a barrier to invasion beyond the subepithelial region in the majority of cases, but invasive growth into the cornea may occur. Corneal involvement is of great significance in scheming therapy, essentially with regard to being a risk factor for recurrent disease (Esmaeli et al., 2001; Nijhawan et al., 2004). In rare cases, conjunctival melanoma may extend directly into the globe or into the orbit (Tuomaala and Kivelä, 2004; Cohen et al., 2013). Clinical metastases usually take place first to the lymph nodes in just about 50% of patients with regional metastases (Esmaeli et al., 2001). Typically, the medial tumors are supposed to spread to the submandibular area and the lateral lesions to the preauricular region (Cook et al., 2002).

Ultimately, systemic dissemination to many body organs may occur, even though this often arises exclusive of prior clinical proof for involvement of regional lymph node. The lung, brain, liver, skin, bone, and the gastrointestinal tract are the most affected site for metastases (Esmaeli et al., 2001).

The histopathologic diagnosis of malignant melanoma of the conjunctiva necessitates identification of atypical melanocytes that may differ from apparent to tremendously subtle. The atypical melanocytes may show prominent nesting in the junctional region with pagetoid extension of single cell or groups of tumor cells into the overlying epithelium. The presence of epithelioid cells with pleomorphic nuclei, prominent nucleoli, atypical mitoses, and abundant cytoplasm make the diagnosis more evident (Jakobiec et al., 1989).

Invasion of tumor cells into the underlying substantia propria of the conjunctiva is diagnostic for invasive malignant melanoma particularly when there is failure of the maturation that is found in most nevi of superficial plump cells to more deeply situated spindle cells (Folberg et al., 1989). The thickness of the tumor, which is assessed by determining the distance from the epithelial surface to the deepest extent of the neoplasm in the substantia propria, is of major prognostic significance (Ho et al., 2007).

Mitomycin is a potent chemotherapeutic agent that inhibits fibroblasts and, therefore, diminishes scarring after glaucoma filtering surgery (Finger et al., 1998; Cunneen et al., 2009). Outpatient radiotherapy is indicated as needed in patients with conjunctival melanoma.

The surgical treatment of conjunctival melanoma is complete removal of the tumor, with applying cryotherapy to the normal-appearing conjunctiva surrounding the lesion. Excision of invasive conjuctival melanoma associated with adjunctive brachytherapy plus topical chemotherapy will alleviate the local tumor control and little ocular morbidity (Damato and Coupland, 2009). Exenteration of the orbit occasionally is compulsory for large melanomas that show orbital invasion. Its main indication is for local debulking of a tumor despite exenteration of the orbit is not linked to increased patient survival (Paridaens et al., 1994a), (Paridaens et al., 1994 b).

CONCLUSION

Conjunctival melanoma is very rare tumor relative to their counterpart in the ciliary body. Its incidence was reported

mainly in white race and tremendously rare in dark race. Owing to these data, our presenting case seems to be of great importance to be reported and highlighted.

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