

Conjunctival Amelanotic Melanoma – A Case Report

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ABSTRACT

Conjunctival melanoma is a relatively rare malignancy. It is presented as pigmented nodule in any area of conjunctiva, amelanotic tumors are pink with smooth appearance. The authors describe an amelanotic melanoma of the conjunctiva in an 82-year-old female patient. Cytological, histopathological and immunohistochemical studies revealed an invasive amelanotic melanoma exhibiting S-100 and MART-1 positivity. The patient underwent surgical and chemotherapy treatment and three years after the initial treatment is in the terminal stage of metastatic disease. Absence of pigmentation delayed early clinical detection and treatment. Awareness of this nonpigmented melanoma is crucial for early recognition and appropriate management.

Key words: conjunctival amelanotic malignant melanoma, immunohistochemistry, S-100, MART-1

Introduction

Conjunctival melanoma accounts for about 2% of all ocular malignancies with incidence varying from 0.024–0.08 per 100,000^{1–7}. It is equally common to men and women, most prevalent in elderly and middle-aged, although 3 cases of malignant melanoma in children have been reported⁸.

The tumor may originate from Primary acquired melanosis – PAM (50–75% of cases), preexisting nevus (20–25% of cases) or as de novo lesion (25% of cases)^{9,10}.

Malignant melanoma is presented as pigmented nodule usually containing feeder vessels, amelanotic tumors are pink with smooth appearance. The tumor can occur in any area of conjunctiva including the tarsus, fornices, and caruncle, but the most common site is the perilimbal interpalpebral bulbar conjunctiva. Those not on the bulbar surface appear to behave more aggressively.

Histopathologically melanomas show infiltration of subepithelial stroma with atypical pigmented cells with prominent nucleoli and abundant cytoplasm and atypical mitotic figures. For further analysis immunohistochemical stains for S-100 protein, HMB-45 and Melan A (MART-1) may be used^{11–13}.

The treatment of conjunctival melanoma is surgical, with complete removal of the tumor, combined with cryotherapy and chemotherapy if necessary^{14,15}. Exenteration of the orbit is used with large tumors¹⁶.

Materials and Methods

Cytological, histopathological and immunohistochemical studies of aspiration and incisional biopsy of the conjunctival tumor in an 82-year-old white woman.

Case report

An 82-year-old female patient was referred to our department for surgical treatment of tarsal conjunctival tumor of her left eye. The patient reported reddening and itching of her left eye for the last 4 months for which she was treated with topical antibiotics and corticosteroids. One month prior to admittance she noticed swelling of the inner side of the eyelids. Biomicroscopy showed nodular tumor of tarsal conjunctiva sizing approximately 2×3×3 cm (Figure 1).



Fig. 1. Intraoperative picture of conjunctival amelanotic melanoma.

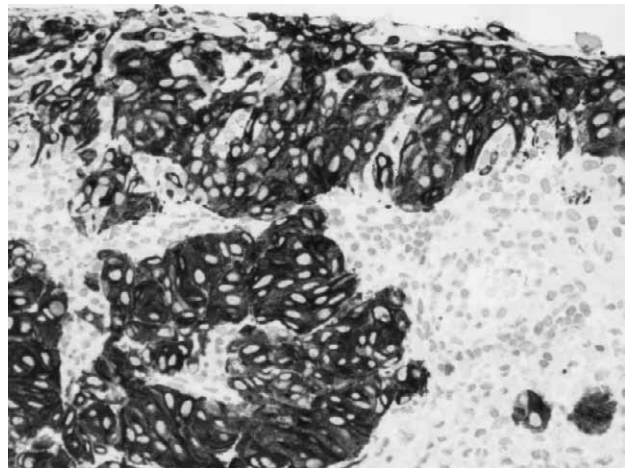


Fig. 2. Immunohistochemical stains exhibiting S-100 and MART-1 positivity.

Results

Cytological examination of aspiration biopsy revealed dense mass of malignant cells highly suspicious of spindle cell melanoma. Further surgical excision and immunohistopathologic examination revealed malignant cells exhibiting S-100 and MART-1 positivity (Figure 2) and CK negativity (Figure 3). CT scans revealed no retrobulbar infiltration.

One month later exenteration of the orbit was performed due to recidival tumor.

Four months after the initial treatment the patient underwent radical neck dissection with histopathologic verification of melanoma metastases in left submandibular lymph nodes.

Three years after the initial treatment the patient is in the terminal stage of disease with multiple cerebral metastases (Figure 4).

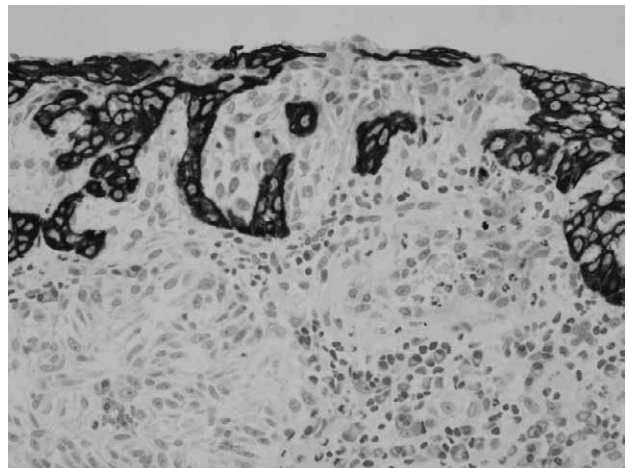


Fig. 3. Immunohistochemical stains exhibiting CK negativity.

Discussion and Conclusion

Conjunctival malignant melanoma is a potentially deadly tumor. The overall mortality rate from conjunctival melanoma is about 25%^{17,18}. Typically, metastases first develop in submandibular and parotid lymph nodes. Unfavorable prognostic factors are conjunctival site, orbital or scleral invasion, full-thickness intraepithelial spread, and involvement of the eyelid skin margin¹⁹.

One research found surgical technique of tumor management to be possibly related to tumor metastases and death²⁰.

Amelanotic malignant melanoma is often associated with a poorer prognosis. The lack of pigmentation makes clinical diagnosis virtually impossible, and diagnosis can only be established histopathologically. Awareness of this nonpigmented variety of melanoma is crucial for early recognition and appropriate management.

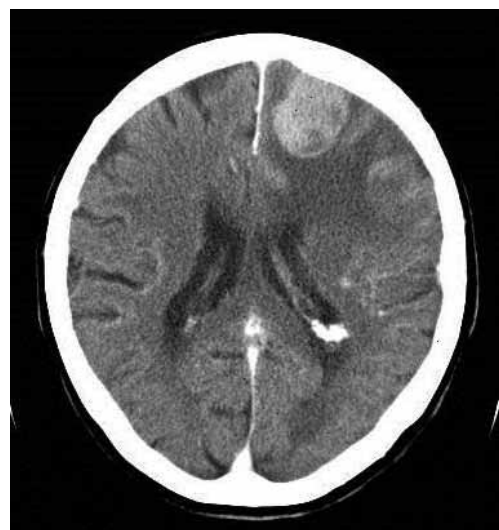


Fig. 4. CT scan showing multiple cerebral metastases.

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AMELANOTIČNI MELANOM KONJUKTIVE – PRIKAZ SLUČAJA

SAŽETAK

Melanom konjunktive je relativno rijedak malignom. Najčešće se manifestira kao pigmentirani čvorić na bilo kojem dijelu konjunktive; amelanotični tumori su ružičaste boje i glatke površine. Autori rada opisuju slučaj amelanotičnog melanoma konjunktive 82-godišnje žene. Citološkom, histopatološkom i imunohistokemijskom analizom utvrđen je invazivni amelanotični melanom pozitivan na S-100 i MART-1 markere. Pacijentica je prošla kirurško i kemoterapijsko liječenje, te se tri godine nakon početnog tretmana nalazi u terminalnoj fazi proširene metastatske bolesti. Izostanak pigmenta kod melanoma odgodio je rano otkrivanje i liječenje. Svjesnost na postojanje nepigmentiranog melanoma je ključna za rano postavljanje dijagnoze i primjereno liječenje.