Case Report

Conjunctival melanoma in a child: A clinicopathological report

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Abstract

We report a case of a 16 years old Asian Indian boy who presented with a large brownish lesion measuring 20 × 12 mm on the temporal conjunctive in his right eye. Anterior segment optical coherence tomography revealed cystic spaces without scleral involvement. The patient underwent conjunctival excisional biopsy using “no touch” technique with double freeze-thaw cryotherapy to underside of the adjacent conjunctival margins. Excision involved 4 mm of the surrounding apparently normal conjunctiva. Absolute alcohol epitheliectomy was done at the limbus and surrounding 2 mm of cornea to devitalize residual atypical melanocytes if any. Histopathology confirmed diagnosis of conjunctival melanoma. We started the patient on topical mitomycin C 0.04% with one weekly on and off cycles postoperatively. No recurrence was noted after nine months follow up.

Keywords: Conjunctival melanoma, Asian child, Anterior segment optical coherence tomography, Melanoma histopathology

Introduction

Conjunctival melanoma is a rare but potentially lethal neoplasm. It mostly affects middle aged and elderly. Conjunctival melanocytic lesions are commonly seen in Caucasian population (89%) and least common in Asian Indians (<1%).1 Children are rarely affected by the disease with very few case reports in literature. Conjunctival nevi are common in children but malignant transformation is rare. We hereby, report a case of a large conjunctival melanoma presenting in a young Asian Indian male child.

Case report

An Indian boy in his mid to late teens presented with a large brownish lesion in the right eye. The lesion was present since childhood and was initially pin-head sized and showed progressive increase over the last 2 years. There was no history of surgical intervention in the past. The pigmented lesion involved almost entire temporal aspect of the bulbar conjunctiva. Fornices and lateral canthus were spared (Fig. 1A). The lesion measured approximately 20 × 12 mm, extending from the limbus up to the lateral canthus. There were multiple cystic spaces on the surface of the lesion. The anterior segment optical coherence tomography (AS-OCT) revealed cystic spaces and sclera appeared to be spared (Fig. 1B). We considered differential diagnosis of conjunctival nevus or melanoma. The patient underwent conjunctival excisional biopsy by ‘no touch’ technique with cautery of scleral bed with double freeze-thaw cryotherapy to the underside of the adjacent conjunctival margins. The bare sclera was covered by conjunctival advancement. Four millimeters of the surrounding conjunctiva was excised. Absolute alcohol epitheliectomy was done at the limbus and surrounding 2 mm of cornea to devitalize residual atypical melanocytes. Histopathology revealed intact conjunctival epithelium. Basal layer showed junctional activity the tumor cells present in the...
subepithelial area within the substantia propria were arranged in sheets and nodules, with presence of melanin pigment in tumor cells. There was presence of early subepithelial infiltration by the tumor cells and melanin pigment production with cystic changes. On higher magnification, there was presence of tumor infiltration in the subepithelium showing dominantly round to oval cells and occasionally spindle shaped cells. The nuclei had hyperchromatic and pleomorphic morphology with intranuclear inclusions and moderate to scanty eosinophilic cytoplasm. There were frequent atypical mitotic figures. The tumor cells were seen arranged around cystic spaces lined by flat benign epithelium (Fig. 1C). Immunohistochemical stains like HMB-45 stained the cytoplasm and cell membrane (Fig. 1D), Melan A stain was also positive. The Ki-67 immunohistochemical test was strongly positive indicating moderate to high proliferation index. All the features confirmed diagnosis of conjunctival melanoma.

The patient’s thorough general physical examination along with metastatic work up including Positron Emission Tomography (PET) scan was unremarkable. We started the patient on topical mitomycin C 0.04% with one weekly on and off cycles. On nine months follow up, no recurrence was seen.

Discussion

Our case had a conjunctival nevus which progressed to a large conjunctival mass later confirmed to be a melanoma on histopathology. Literature revealed very few cases of conjunctival melanoma presenting in children.1 Conjunctival nevi in children are relatively common, indicating that a vast majority do not progress to melanoma. Alkatan et al.4 have described the entity Inflamed Juvenile conjunctival nevi (IJCN) in adolescents associated with vernal keratoconjunctivitis. In their study, 48% of compound nevus constituted of IJCN, the most common location being the bulbar conjunctiva. The authors concluded that some of the IJCN are excised because of suspicious of malignancy based on the change in appearance or increasing size. These changes are actually attributed to the presence of inflammation, thus ophthalmologists should be aware of the entity of IJCN in order to avoid misinterpretation of these findings as being indicative of malignancy.

Strempel et al.5 reported 3 cases of conjunctival melanoma. One is of a 16 year old Caucasian girl with history of conjunctival mass excision at 11 years of age. The lesion was situated in the right eye temporal to limbus. Five years later she presented with mass in the lacrimal sac region. Histopathologically, it was detected to be melanoma with generalised systemic spread. However, no local recurrence occurred. A second case was of a 3 year old Turkish boy with brownish mass in temporal bulbar conjunctiva, noted at 4 months of age. Later, massive growth occurred over 4 years. No recurrence was noted on follow up. Two more isolated case reports of a 9 year old white girl, who underwent conjunctival excision of a black pigmented lesion in inferior palpebral conjunctiva which was histopathologically proven to be melanoma. Another 9 year old Mexican boy had a yellowish nodule at limbus, which on excision was histopathologically proven as conjunctival melanoma. A brief summary of all the reported cases of conjunctival melanoma in children is given in Table 1. Histopathological findings such
Table 1. Literature review of cases of conjunctival melanoma in children.

| Authors/Year of publication | Age/sex | Site | Initial diagnosis | Treatment | Lymph node metastasis | Metastasis | Brachythrapy/chemotherapy |
|-----------------------------|---------|------|------------------|-----------|-----------------------|------------|--------------------------|
| 1 Croxatto et al.2 (1987)   | 11 years/Male | Limbus | Compound nevus | Exenteration | Cervical | Yes (extensive metastasis) | Systemic chemotherapy |
| 2 McDonell et al.3 (1989)   | 12 years/Male | Limbus | Melanoma | Excisional biopsy | Yes | Parotid gland | Systemic Chemotherapy |
| 3 Stempel et al.5 (1999)    | 14 years/Female | Temporal Bulbar | Active melanocytic lesion | Excisional biopsy | - | Orbit and Lacrimal sac | Systemic chemotherapy |
| 4 Stempel et al.9 (1999)    | 3 years/Male | Temporal Bulbar | Melanocytic lesion with uncertain etiology | Excisional biopsy | None | - | None |
| 5 Stempel et al.5 (1999)    | 4 years/Male | Temporal Bulbar | Melanoma | Excisional biopsy | None | - | - |
| 6 Brownstein et al.2 (2006) | 9 years/Female | Lower Palpebral conjunctiva | Junctional nevus/compound nevus | Lower eyelid resection | None | None | None |
| 7 Brownstein et al.2 (2006) | 4 years/Male | Lower Palpebral conjunctiva | Melanoma | Excisional biopsy | None | None | None |
| 8 Masaoudi A et al. (2013)  | 10 years/Male | Bulbar conjunctiva | Melanoma | Excisional biopsy | Pre-auricular | Parotid gland | Plaque brachythrapy and systemic chemotherapy |
| 9 Yangzes et al. (2015)     | 16 years/Male | Temporal conjunctiva | Melanoma | Excisional biopsy | None | None | None |

Fig. 2. The panel of the photomicrographs showing the different aspects of the tumor. (A) The intraepithelial lesion with early sub-epithelial infiltration by the tumor cells and melanin pigment production. One of the smaller cysts could be included in this photomicrograph. (Hematoxyline eosin, 500×); (B) the infiltrating tumor cells located in the sub-epithelium showing dominantly round to oval cells occasionally spindle in shape. The nuclei are hyperchromatic and pleomorphic with intranuclear inclusions and moderate to scanty eosinophilic cytoplasm. Majority of the tumor cells contain intracytoplasmic melanin pigment. (Hematoxyline eosin, 500×); (C) in addition to features described in B, some of the tumor cells along the infiltrating front have grossly pleomorphic bizarre nuclei. (Hematoxyline eosin, 500×); (D) photomicrograph of the tumor cells showing strong cytoplasmic positivity for melan A in immunohistochemistry staining. (peroxidise anti-peroxidase, 500×); (E) photomicrograph of the tumor cells showing nuclear positivity in Ki-67 immunohistochemistry staining. (peroxidise anti-peroxidase, ×1000) indicating moderate to high proliferative index.
as atypical melanocytic cells involving various epithelial levels, invasion of conjunctival stroma without maturation and atypical mitoses strongly point towards melanoma.1,2 Nevertheless, a long term follow up is required to ascertain the prognosis of conjunctival nevi and should be followed up for a long time and treated when suspicion of transformation to malignancy arises. This is so far, to the best of our knowledge, the first case report of an extensive conjunctival melanoma in an Asian Indian male child.

Conflict of interest

The authors declared that there is no conflict of interest.

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