

Keratoacanthoma of the conjunctiva complicating xeroderma pigmentosum

Sir,

We wish to report a case of keratoacanthoma of the conjunctiva in a patient of xeroderma pigmentosum for its rarity. Xeroderma pigmentosum is a rare autosomal recessive disorder characterized by photosensitivity, pigmentary changes, premature skin ageing and various neoplastic disorders. The underlying defect is abnormal DNA excision repair. In 20% of the cases the patients have normal excision repair, but the post-replicative repair is defective.^[1] A variety of neoplastic conditions like basal cell carcinoma, squamous cell carcinoma, malignant melanoma and angiosarcoma have been reported along with this condition. They predominantly occur on the sun exposed area of the body and are thought to be resulting from UV-induced mutations and immunosuppression.^[2]

A 17-year-old female presented with dry, scaly and wrinkled skin all over the body. She also had hyperpigmented macules and scars. All these lesions were present since early infancy and were more pronounced on the sun exposed parts. She also complained of an asymptomatic growth on the nasal aspect of the bulbar conjunctiva of the left eye since 4 weeks. The growth was rapidly increasing in size. There

was a history of photosensitivity since childhood. No history of trauma or foreign body in the eye could be elicited. She had two other siblings with a similar condition.

On examination, she was found to have freckling interspersed with atrophic patches over the face and extremities. There was conjunctival congestion in the left eye. A pale pink glistening swelling around 12×10×8 mm in dimension was seen along the nasal aspect of the bulbar conjunctiva [Figure 1]. The surface of the swelling had a small hyperkeratotic area at the center. The rest of the anterior segment of the eye revealed no abnormality. Fundus examination was normal. B-Scan ultrasonography did not reveal any posterior segment abnormality. All routine hematological and biochemical investigations were within normal range. A CT scan of the orbit and brain was unremarkable.

The lesion was excised under peribulbar anesthesia and was sent for histopathological examination. Histopathological study of the specimen revealed a central acanthotic epithelium surrounded by a collarette of differentiated squamous epithelium. However, normal conjunctival epithelium could not be found at the edge due to poor slide preparation.

The diagnosis of keratoacanthoma was established by the short clinical history, morphological features and a sharp outline between the tumor and the stroma. An immunohistochemistry study could not be performed



Figure 1: A pale pink nodule on bulbar conjunctiva

because this facility was not available.

A keratoacanthoma most frequently presents as a rapidly growing lesion on the sun-exposed area of the body. It may also involve the eyelids. It has frequently been reported in association with xeroderma pigmentosum.^[3] However, keratoacanthoma of the conjunctiva is a very rare condition and until 2001, only twelve cases had been reported.^[4] Conjunctival keratoacanthoma in association with xeroderma pigmentosum is still rarer. The mean age of previously reported cases was 40.75 years.^[4] However, in our case, the presence of xeroderma pigmentosum is probably responsible for the younger age of onset. In previously reported cases, there was a male preponderance and five of them reported a foreign material entering the eye; there was no such history in our patient.^[4] Involvement of the nasal aspect of the conjunctiva in our patient was similar to the report of Perdigo et al,^[5] whereas in most other cases, the temporal aspect was involved.

Keratoacanthoma of the conjunctiva in a patient of xeroderma pigmentosum requires early and proper diagnosis. It can be differentiated from squamous cell carcinoma by the rapidity of its growth and suggestive histopathological features.

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