Inferior Bitot's spot in a case of Crouzon syndrome: The environmental theory reaffirmed

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DESCRIPTION

A 35-year-old man, known case of Crouzon syndrome with bilateral proptosis presented to us. He had undergone tarsorrhaphy in one eye, and the fellow eye showed striking inferior scleral show with lagophthalmos. On torch light examination, multiple white translucent irregularities were seen on the inferior conjunctiva. On slit-lamp biomicroscopic examination, they appeared as dry, corrugated, foamy patches radiating from the limbus in a semicircular fashion (figure 1). On blinking, they stood prominently with interruption of the overlying tear film. He was a chronic alcoholic by nature. Taking all the contributory features into account, a clinical diagnosis of Bitot's spots was made, although the location of inferior conjunctiva was atypical.

Crouzon syndrome is a type of craniosynostosis, where there is retrusion of the inferior and lateral orbital margins, resulting in a short orbital floor. Proptosis is almost universal, and up to half of them may manifest exposure-related complications.[1] While conjunctivitis and keratitis are the most commonly studied exposure induced manifestations, we here describe for the first time, development of Bitot's spots in the exposed inferior conjunctiva of a case of Crouzon syndrome.

Ocular epithelial surface, like other body mucosal surfaces undergo metaplastic keratinisation in the presence of Vitamin A deficiency. When advanced, they become grossly visible as 'xerosis'. Conjunctival xerosis, an initial manifestation of xerophthalmia, are called the Bitot's spots. They appear as a dry, discrete, corrugated foamy patch, commonly triangular, with the apex pointing towards the outer canthus. They are most commonly seen in the temporal quadrant followed by the nasal. Variations in shape and location though rare, may exist.

Bitot's spots are primarily a collection of the desquamated keratin and commensal diptheroids (corynebacterium xerosis).[2] The latter is a gas forming bacteria which contributes to their foamy appearance. They become more prominent as the tear film retreats over it, manifesting 'like sandbanks at receding tide'. The development of Bitot's spot is also postulated to be related to exposure, as they are predominantly found in the exposed parts of the conjunctiva. This is because, in these regions, the above said accumulations are fairly protected from the effect of lid massage.[3] That is why they are most commonly found in the temporal followed by the nasal bulbar conjunctiva.

The aetiology of Bitot's spot is confusing, since they are sometimes found when there's no evidence of vitamin A deficiency, nor do they always disappear after vitamin A therapy. Especially in children over 6 years of age, it is likely due to a past or chronic suboptimal retinol levels. In adults, changes are more likely due to chronic ultraviolet exposure, smoke and dust, and should not be pathognomonic of vitamin A deficiency. The development and persistence of Bitot's spots are almost certainly related to exposure.[2]

In our case, as the exposure was primarily inferior, our patient may have manifested inferior Bitot's spots, thereby supporting the environmental theory. In literature, there has been a single previous report of inferior Bitot's spot, that occurred in a case of lower lid ectropion.[4]

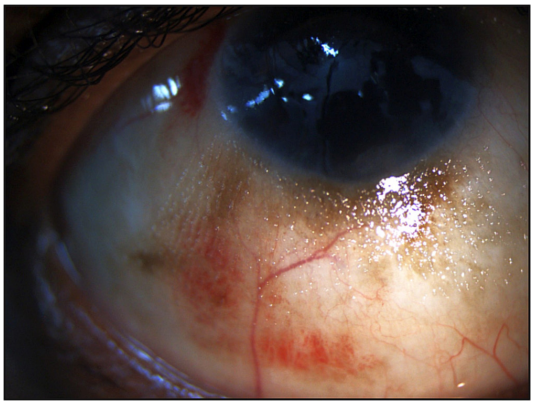

Figure 1 Slit-lamp biomicroscopic photograph showing right eye of a patient of Crouzon syndrome in upgaze, depicting dry, foamy Bitot's spot over the inferior conjunctiva, radiating from limbus in a semicircular fashion, appearing like 'sandbanks at the receding tide'.

Learning points

- Crouzon syndrome is a type of craniosynostosis which results in proptosis and exposure-related complications.
- Bitot's spot are an accumulation of desquamated keratin and commensal diphtheroid, accumulating in the exposed parts of the conjunctiva, commonly temporal.
- Crouzon's syndrome patients show significant inferior exposure, and hence may manifest inferior Bitot's spots.

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