

## Visual Treats in Dermatology

# Beaded papules of eyelids

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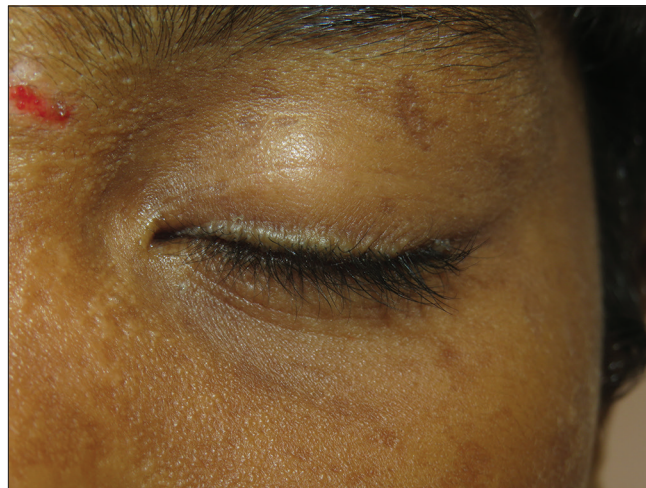
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**Quick Response Code:**



A 12-year-old adolescent boy presented with multiple skin colored papules of both eyelids since 5 years of age [Figure 1]. He also complained of verrucous plaques over the extensors of both elbow and knee joints associated with occasional itching. He was born of third degree consanguineous marriage with negative family history in other siblings. His mother gave the history of fluid filled vesicles during early childhood which healed with scarring. Examination revealed the presence of beaded papules arranged linearly along the margin of both eyelids symmetrically consistent with moniliform blepharosis evident on dermoscopy [Figure 2]. There was also the presence of pock like scarring over the face and extremities on a background of waxy thickened skin. Hoarseness of voice was present which was confirmed by direct laryngoscopy showing thickened vocal cords. There were no neurological manifestations except for paroxysms of anger episodes. A skin biopsy from the lesion revealed the presence of homogenous hyaline eosinophilic material in the dermis staining positively for periodic acid Schiff stain thus confirming a diagnosis of lipoid proteinosis.

Lipoid proteinosis also referred to as Hyalinosis cutis et mucosae or Urbach Wiethe disease is an autosomal recessive disorder characterized by a defect in the gene ECM1 coding for extracellular matrix glycoproteins important for structural support and scaffold for the dermis and basement membrane. Skin manifestations can occur as early as infancy in the form of blistering. The skin then becomes thickened and waxy with verrucous and psoriasiform plaques mainly over the



**Figure 1:** Multiple beaded papules arranged linearly over the margin of the eyelid (moniliform blepharosis).

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**Figure 2:** Dermoscopy highlighting the linearly arranged closely set beaded monomorphic papules (DermLite DL4, Polarized mode,  $\times 10$ ).

frictional sites. Similar thickening can also occur in the vocal cords leading to progressive hoarseness of voice. Neurological manifestations range from seizures to behavioral disorders and

neuropsychiatric disturbances. A computed tomogram of the brain reveals bean-shaped temporal calcifications bilaterally. A skin biopsy can be done to rule out other differentials such as amyloidosis, porphyria, and scleromyxedema.<sup>[1]</sup>

#### **Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent.

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#### **Conflicts of interest**

There are no conflicts of interest.

#### **REFERENCE**

1. Bhattacharjee R, Chatterjee D, Vinay K. Lipoid proteinosis. *JAMA Dermatol* 2018;154:1479-80.

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