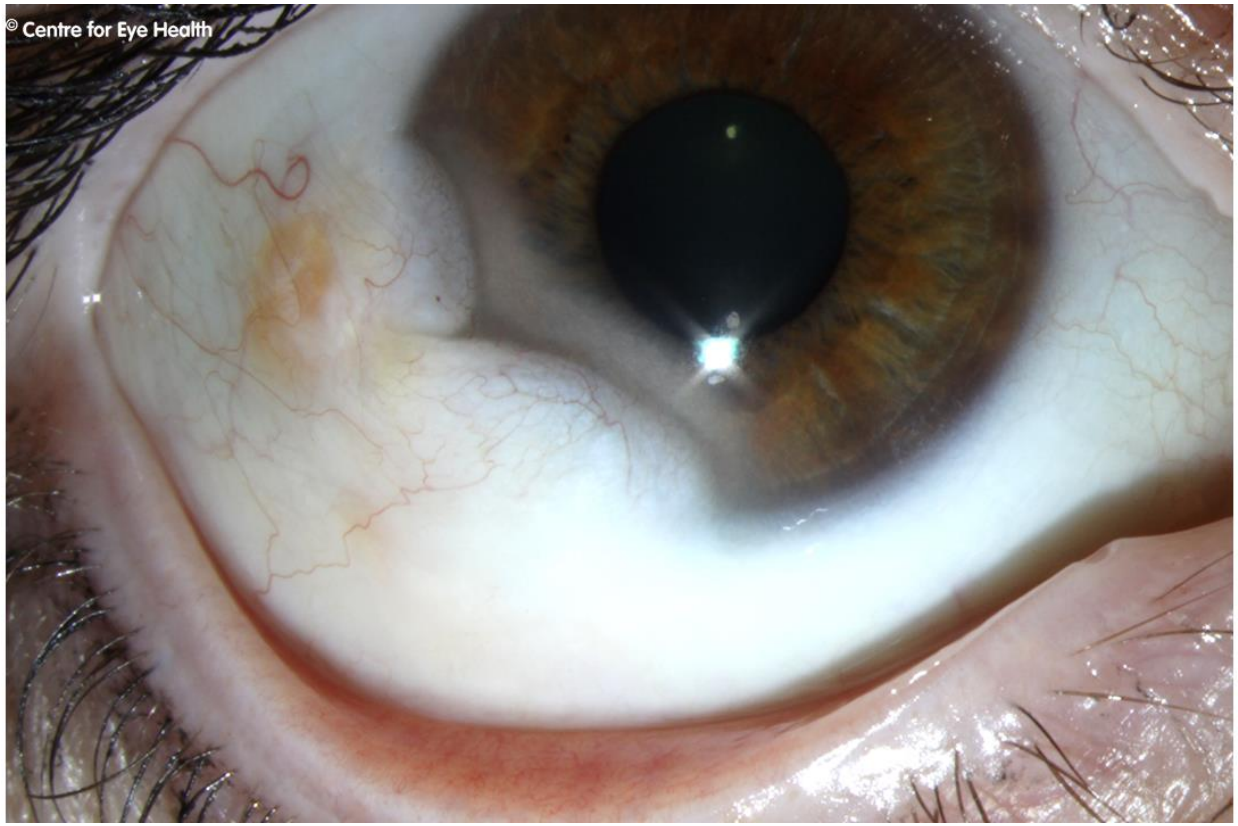
CFEH facebook Cases

CFEH Facebook Case #121

A 53 year old female presented for examination of a conjunctival lesion that has been present since birth. Shen mentions this eye has always been weaker and undertook patching of the eye for a short time at 8 years of age. BCVA in the right eye is 6/19 with a correction of +4.00/-2.00 x 80. Can you identify the lesion seen below and what systemic associations might this condition have?



Centre for Eye Health Education
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ANSWER

This is a limbal dermoid (epibulbar dermoid) which is a benign, smooth white lesion typically found infero-temporally on the globe or limbus. They may be unilateral or bilateral and are believed to arise due to an embryonic anomaly rather than having a genetic cause. Limbal dermoids contain choristomatous tissue (tissue that is not normally found at that site).

Limbal dermoids are usually graded according to the extent of tissue involvement:

Grade I: superficial corneal involvement, less than 5mm diameter and localized to the limbus.
Grade II: larger lesions covering most of the cornea, extending deep into the stroma but not involving Descemet's membrane
Grade III: involvement of the entire cornea and anterior chamber

Grade I lesions usually show slow growth over time and cause oblique astigmatism as they flatten the adjacent cornea. This may cause amblyopia such as that seen in our patient.

Interestingly, upon questioning our patient mentioned that she was born with only one kidney. This increases the likelihood that she has Goldenhar syndrome (oculo-ariculo-vertebral spectrum). This is a developmental malformation associated with limbal dermoids, auricular malformations and skeletal malformations. It typically affects one side of the body and additional manifestations can include congenital heart defects, renal defects (hypoplasia or failure of an organ to develop at all during embryonic growth), or central nervous system malformations (hydrocephalus, intracranial lipomas, cranial nerve dysgenesis).

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