Sir,
A case of Hunter syndrome with bilateral retinal detachment

We present a case of Hunter syndrome (HS) with bilateral simultaneous rhegmatogenous retinal detachment, an association that to our knowledge has never been reported before.

Case report

A 32-year-old male with a history of HS was brought to the eye casualty with behaviour signifying visual loss. The patient had coarse facial features with a prominent forehead, flattened nasal bridge, and impaired mental development but no seizures or any associated ocular features. The diagnosis of HS had been confirmed by the absence of serum enzyme iduronate-2-sulphatase. Because of the difficulty in performing a complete ophthalmic examination, an examination under anaesthesia was carried out, which revealed bilateral rhegmatogenous retinal detachments with proliferative vitreoretinopathy (Figures 1 and 2). Bilateral immediate sequential 20G pars plana vitrectomies and inferior retinectomies with silicone oil tamponade resulted in successful retinal reattachment in both eyes.

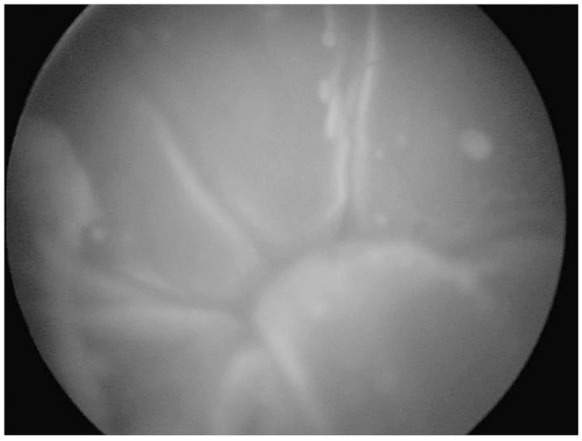

Figure 1 Retinal detachment in the right eye.

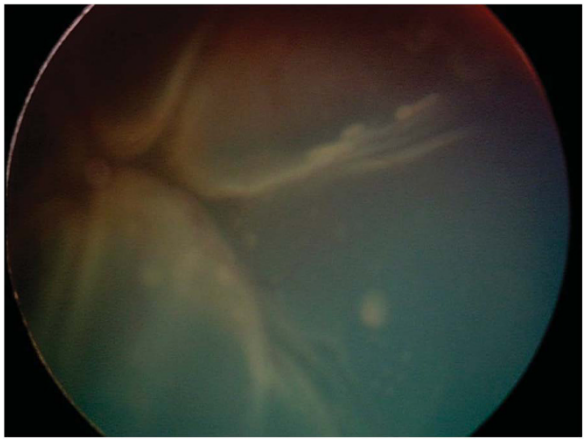

Figure 2 Retinal detachment in the left eye.

Discussion
First described by Charles Hunter in 1917, HS is an X-linked recessive type II mucopolysaccharidosis (MPS) characterized by deficient or absent enzyme iduronate-2-sulphatase.[1] Ocular manifestations of mucopolysaccharidoses include corneal opacification, glaucoma, pigmentary retinopathy, and optic atrophy due to deposition of glycosaminoglycans in various structures of the eye.[2] HS is distinct in the absence of corneal features, exhibits a wide variation in clinical severity, and generally presents with learning disability and seizures.[2]

The retinopathy in HS is due to deposition of glycosaminoglycans in the retinal pigment epithelium and interphotoreceptor matrix leading to progressive photoreceptor loss. High-resolution OCT of the retina in HS has shown marked thinning of the outer retina, especially the photoreceptor layer.[3] There have been reports of free-floating vitreous opacities, epiretinal membranes, and uveal effusion syndrome in HS.[4,5] Rhegmatogenous retinal detachment has never been reported in HS to the best of our knowledge. We postulate that retinal degeneration in HS may lead to thinning of the retina and there may be a risk of rhegmatogenous retinal detachment especially in the presence of vitreo-retinal interface abnormalities, which have been described.[4] In summary, we describe a case of HS with bilateral retinal detachments and conclude that a complete dilated retinal examination (under anaesthesia if required) should be carried out in such patients to exclude any vitreo-retinal anomalies.

Conflict of interest

The authors declare no conflict of interest.

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