a homozygous deletion of 8815 bp in both siblings with break points in introns 11 and 13 resulting in loss of exons 12 and 13 (Figure 2A and B). Further examination of the region showed a 34-bp repeat (differing by only 1 bp) flanking the break points, suggesting homologous recombination as the mechanism for generating this deletion. The predicted result of this deletion would be that exon 11 splices directly to exon 14 and remains in frame, resulting in a protein of 685 amino acids, compared with the wild-type protein of 829 amino acids (Figure 2E). This was confirmed using complementary DNA from patient and control lymphocytes and sequencing the products (Figure 2C and D).

Comment. Hypotrichosis associated with juvenile macular dystrophy is caused by mutations in CDH3, which encodes P-cadherin, a member of the classic cadherin family. We report here, to our knowledge for the first time, an intragenic deletion causing HJMD that results in the homozygous loss of exons 12 and 13 of CDH3. The resultant transcript has exon 11 spliced directly to exon 14, and although it remains in frame, the predicted protein loses extracellular calcium-binding domains 4 and 5 and part of the transmembrane domain and would therefore be nonfunctional.

Kjaer et al.6 have shown that mutations in CDH3 also cause ectodermal dysplasia, ectodactyly, and macular dystrophy (EEM; OMIM 225280); they also discuss possible mechanisms for this phenotypic variation. Both HJMD and EEM have very similar hair and retinal defects, but patients with EEM have additional limb abnormalities. Hypotrichosis associated with juvenile macular dystrophy is a rare condition with 1 previous report in the ophthalmology literature.7 This case highlights the importance of assessing patients with inherited retinal degeneration for extraocular features and the fact that HJMD should be considered if early-onset macular degeneration is diagnosed in the context of hypotrichosis.

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Bilateral Ophthalmic Artery Occlusions Due to Probable Varicella-Zoster Virus Vasculopathy

Varicella-zoster virus (VZV) vasculopathies account for almost one-third of arterial ischemic strokes in children.1 Visual complications are rare, with previous reports occurring secondary to unilateral central retinal artery2 or posterior ciliary artery3 occlusion. We describe the first case, to our knowledge, of an immunocompetent child who became blind due to bilateral ophthalmic artery occlusions secondary to probable VZV vasculopathy.

Report of a Case. A 6-year-old boy visited his local emergency department with a history of sudden painless bilateral visual loss. He was otherwise in good health apart from a history of chickenpox 8 weeks previously. His father had been treated for culture-negative mediastinal tuberculosis a year earlier. Initial examination in the emergency department showed visual acuity of counting fingers OU, a moderate bilateral panuveitis, and diffuse bilateral retinal edema with sheathing of both retinal arterioles and veins. The optic discs were not swollen. Neurological examination findings were otherwise normal. Initial management aimed to treat a possible tuberculosisis optic neuropathy and/or vasculopathy, using oral prednisolone, rifampin, pyrazinamide, and isoniazid. Initial investigations showed no abnormality on hematology and biochemistry tests of peripheral blood. Cytomegalovirus and VZV IgG were both detected on testing of serum, but polymerase chain reaction results for the respective DNA were negative. Evidence of tuberculosis infection was not found, with negative results on both enzyme-linked immunosorbent spot and Heat tests and
normal findings on chest radiography. Magnetic resonance imaging demonstrated no abnormal enhancement in the brain or chiasm, with normal optic nerve appearances. During the next week, visual acuity deteriorated to light perception OU. Panuveitis with retinal vascular sheathing (Figure 1A) persisted, with patchy areas of reperfused retina observed peripherally in the left eye on fluorescein angiography (Figure 1B). Doppler ultrasonography showed no detectable flow in central retinal artery or central retinal vein. Polymerase chain reaction results for a vitreous biopsy specimen were negative for both VZV DNA and tuberculosis. Electrophysiology showed a completely undetectable electroretinogram in both eyes, indicative of loss of outer retinal photoreceptor function and thus not in keeping with dysfunction confined to central retinal artery or vein circulation (Figure 2A). Magnetic resonance angiography showed a mostly normal cerebral vasculature but neither ophthalmic artery could be visualized (Figure 2B), in keeping with the electrophysiological data. Subsequently, VZV IgG was detected in cerebrospinal fluid, confirming VZV as the likely cause of the vasculopathy. Despite a course of systemic acyclovir and intravenous methylprednisolone, he maintains visual acuity of light perception OU.

Comment. Vasculopathy caused by VZV can occur after both reactivation (zoster) and primary VZV infection and may involve both large and small cerebral arteries. Visual loss, previously reported to affect only the posterior ciliary artery and central retinal artery unilaterally, is rare but devastating. Branch retinal arterial occlusions in an immunocompetent adult have been reported in association with posterior uveitis and the presence of VZV DNA detected in the vitreous by polymerase chain reaction. However, VZV IgG and VZV DNA were absent in the cerebrospinal fluid in that case and subsequent investigations confirmed isolated ocular involvement.

Detection of VZV IgG in the cerebrospinal fluid has been shown to be a more sensitive and specific marker for the diagnosis of VZV vasculopathy compared with detection of VZV DNA by polymerase chain reaction alone, where a negative result does not exclude the diagnosis. The diagnosis in the present case was challenging owing to the absence of cerebrospinal fluid pleocytosis (present in one-third of cases) and the presence of normal brain magnetic resonance imaging findings (unusual in VZV vasculopathy).

Prompt diagnosis of VZV vasculopathy is crucial, with an untreated mortality rate reported at 25%. Although ocular complications may be irreversible after several weeks, treatment with systemic acyclovir may prevent further neurological complications and loss of life.

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Idiopathic Pigmented Vitreous Cyst

Idiopathic floating vitreous cysts are very rare. They often give rise to intermittent blurring of vision. Only a few clinical and histopathological reports document features of such cysts, but the origin of these cysts is still a matter of debate. We describe the ophthalmic assessment of a cyst suspended in the anterior vitreous cavity of a 48-year-old patient who visited the Vasan Eye Care Hospital, New Delhi, India.

Report of a Case. A 48-year-old man had a mobile, insectlike floater in the central visual field of his left eye for 4 years. There was no history of trauma,