

Lightning Schisms Across Clear Retinal Skies: A Rare Presentation of Juvenile X-Linked Retinoschisis

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Abstract

Juvenile X-linked Retinoschisis (JXLRS) is a hereditary retinopathy that commonly presents with macular retinoschisis. Rarely in the literature, patients have been reported with findings of peripheral retinoschisis and no evidence of macular schisis or atrophy.

A 13-year-old boy presented with a history of gradually progressive diminution of vision in both eyes over two years. There was no history of trauma, vitreous hemorrhage, or retinal detachment. Best-corrected visual acuity was 6/12 in both eyes. Anterior segment examination was unremarkable.

Fundus examination of the right eye revealed extensive inferior peripheral retinoschisis with two superior operculated retinal holes, while the macula appeared normal. The left eye showed inferior peripheral retinoschisis with a retinal break, again sparing the macula. Spectral-domain optical coherence tomography confirmed normal foveal architecture without macular schisis in both eyes.

This report highlights an atypical presentation of JXLRS with isolated peripheral retinoschisis and preserved macular anatomy. Awareness of such variants is essential for accurate diagnosis and appropriate long-term monitoring for retinal breaks and detachment.

Key-words: Juvenile X-linked retinoschisis; Optical coherence tomography; Peripheral retinoschisis

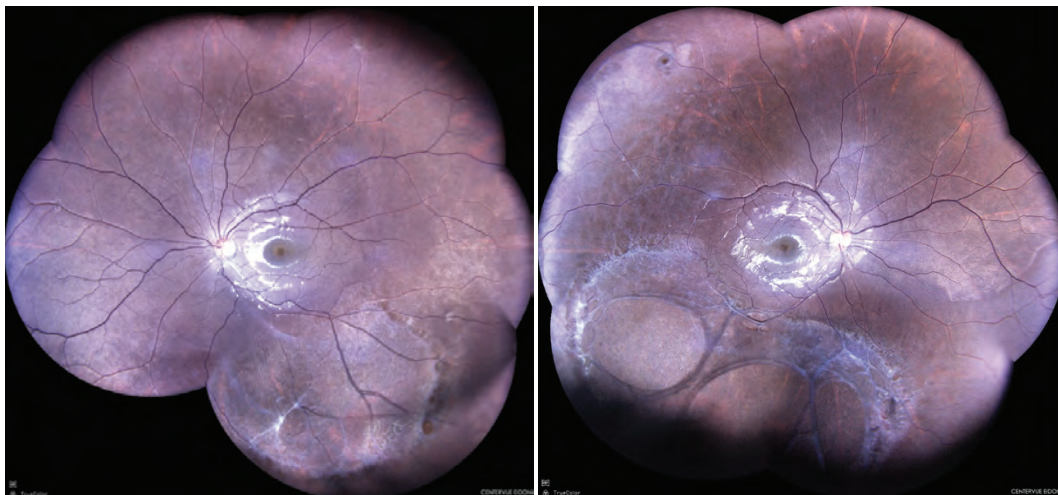


Figure 1: (a) Color fundus photograph showing characteristic spoke-wheel-like schitic cavities at the macula with peripheral retinal schisis. (b) Spectral-domain optical coherence tomography demonstrating multilayered schisis involving the inner nuclear and outer plexiform layers, consistent with juvenile X-linked retinoschisis.

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