

Concurrent Best Disease and Multiple Evanescent White Dot Syndrome: A Rare Ocular Manifestation

Ana Luisa Souto Gandra
Julio Rezende de Andrade
Maria Paulina Viana Miquilino
Raquel Nezio de Caravalho
Neiffer Nunes Rabelo
Giovanna Vieira Moreira
Denise Marinho Pardini
Tereza Cristina Moreira Kanadani

Retina Instituto





Case Report

- 16 year-old french female with vision impairment in the left eye (OS).
- Previously diagnosed with bilateral Best Vitelliform Macular Dystrophy.
- VA: 20/40 and 20/200
- Fundoscopic examination: presence of yellowish vitelliform lesions in both eyes and yellow-white spots in the OS.



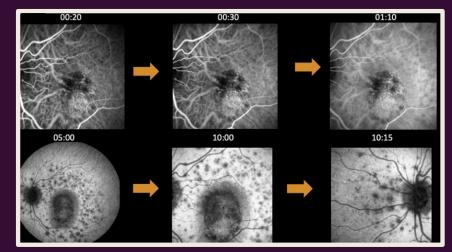


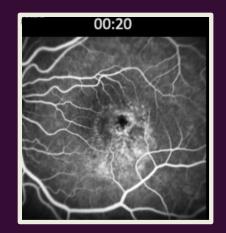


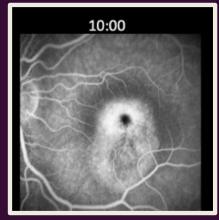


Indocyanine green angiography: hypofluorescence in the vitelliform lesions, with the white dots hypofluorescent in the OS.





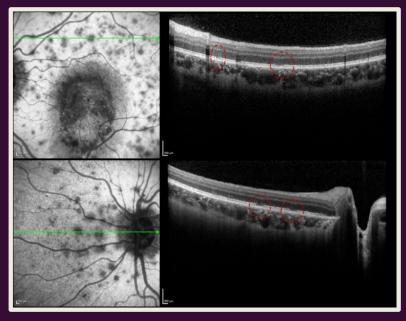


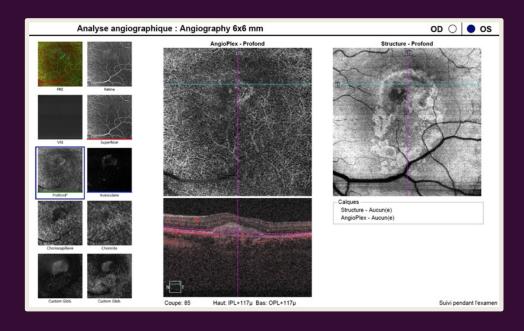


Fluorescein angiography: hyperfluorescence in the vitelliform deposits and impregnation of the optic disc in the OS.









- OCT: subfoveal hyperreflective material accumulation with ellipsoid zone (EZ) disruption in both eyes, with corresponding disruption of the EZ in areas of indocyanine hypofluorescent lesions.
- OCT-A was absencent of positive decorrelation signs \rightarrow expectant management.
- Systemic investigation was normal.
- VA improved to 20/100 in the OS with the disappearance of the white spots





Best Disease X MEWDS

• Best Disease is an inherited dystrophy characterized by bilateral yellow 'egg-yolk' appearance of the macula.





• Multiple Evanescent White Dot Syndrome (MEWDS) is a self-limiting inflammatory disease of the choriocapillaris, that presents with unilateral vision loss and yellow-white spots in the posterior pole to midperiphery.

- The co-occurrence of these conditions has been documented previously, with hypotheses suggesting a proinflammatory microenvironment induced by choroidal neovascularization in Best Disease as a potential trigger for MEWDS-like features.
- This unique case offers additional insights into the pathogenesis of MEWDS, broadening the spectrum of potential causative factors and elucidating potential interactions between distinct ocular diseases.

