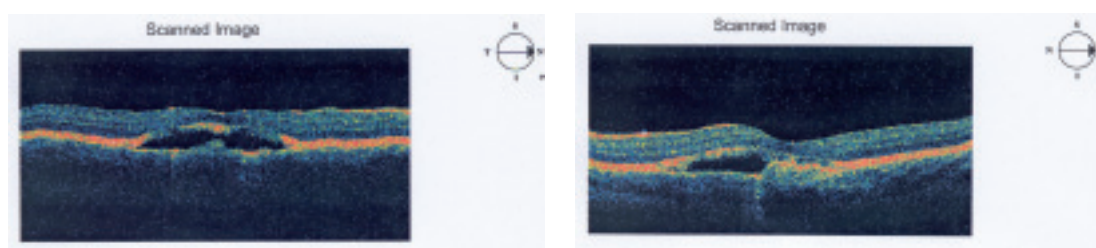

VITELLIFORM MACULAR DYSTROPHY

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PHOTOGRAPH 1

Vitelliform macular dystrophy (Best disease) diagnosed in a young girl born in 1989. She has a familial history of affected members: grand-father, uncle and cousins. EOG Arden ratio was 1.29 OD and 1.40 OS.

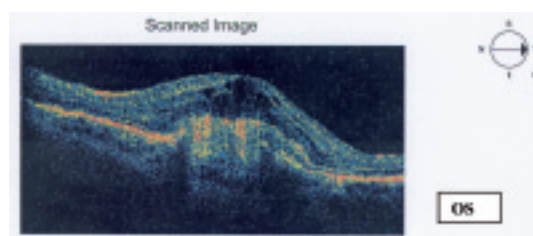
OCT (optic coherence tomography) showed the typical appearance of subretinal deposits with some liquefaction. (OD: marked resorption of material, leaving an optically empty space between the neurosensory retina and the retinal pigment epithelium (RPE); OS: closed to the optically empty space, hyperreflective area caused by the accumulation of material that elevates the RPE)



Because of anisometropia, she had in her young age a bad right vision. Following occlusion of the left eye, she regained good vision in her amblyopic eye. Until June 2008, she retained a good vision of 0.9 OD and 1.0 OS. At this moment the left vision worsened to 0.2.

PHOTOGRAPH 2

On the left eye, she developed a fibroglial reaction secondary to a neovascular membrane in the macula, a relative rare complication of Best disease. OD didn't change. Fluorescein angiogram (photograph 3) confirm the presence of fibroglial reaction secondary to subretinal neovascularization. OCT demonstrated a thickened hyperreflective band and intraretinal fluid.



She was treated by intravitreal injection of anti-VEGF. The intraretinal fluid disappeared, but because of fibroglial reaction, the vision didn't improve.

CONCLUSION

Vitelliform macular dystrophy (Best's disease) is an autosomal dominant disease. Mutations in the VMD2 gene located on the long arm of chromosome 11 were found to be responsible for the disease.

The disease has a relative good visual prognosis. But subretinal neovascularization and fibroglial scar can occur as possible and rare complication.

This case illustrates also the importance of the detection and the treatment of the amblyopic eye.

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