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APLASTIC ANEMIA: INSIGHT INTO PATOPHYSIOLOGY AND MULTI-MODAL IMAGES

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Aplastic anemia: Insight into patophysiology and multi-modal images

PURPOSE

To report a case of aplastic anemia with retinal changes in a young patient and discuss aspects of multimodal evaluation in aplastic anemia.

METHODS

A complete ophalmic examination and complementary evaluation with fundus widefileld, optical coherence tomography (OCT) of the macula and 3 x 3 mm scans of optical coherence tomography angiography (OCT-A).

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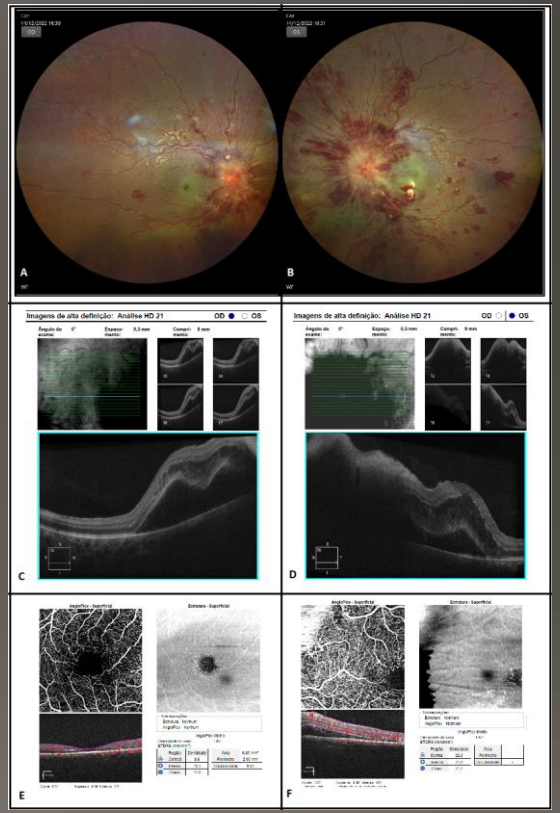
CASE REPORT

We report a case of a 20-year-old female patient with a diagnosis of aplastic anemia under investigation that complaint of sudden visual impairment in both eyes. In the examination, corrected visual was hand movement in both eyes, a phacic patient without changes in anterior biomicroscopy and, in the fundoscopy, she presented retinal hemorrhages and vascular tortuosity in all the four quadrants and swollen disc suggesting of central retinal vein occlusion in both eyes (Figures 1A and 1B). Intraocular pressure was 16 mmhg in both eyes.

The multimodal evaluation at the first moment showed accumulation of intraretinal fluid in the outter plexiform layer and the presence of considerable subretinal fluid (Figures 1C and 1D). OCT-A of the superficial plexus showed a subtly increased and irregular foveal avascular zone (FAZ) (Figures 1E and 1F). Fluorescein angiography was not performed due to the clinical condition of the patient.



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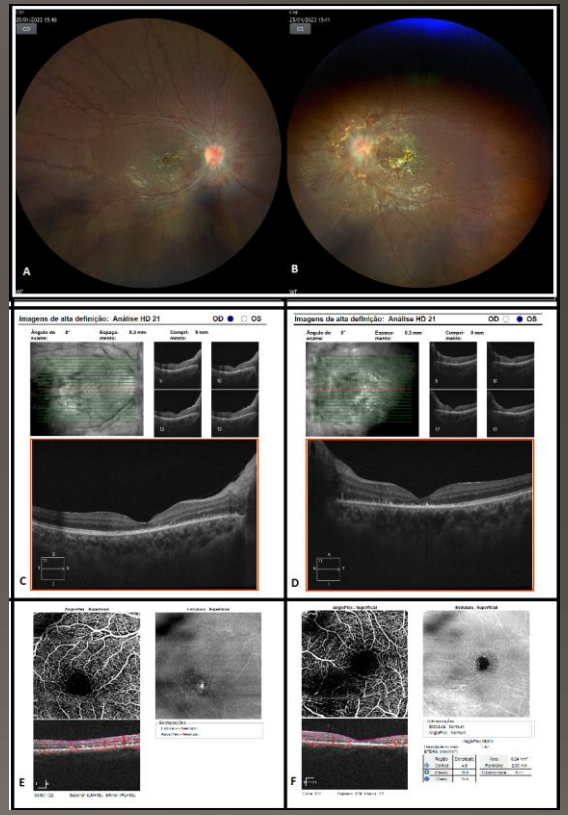
One moth after, following the treatment of the aplastic anemia and improvement of the hematologic parameters, the patient reported improvement in visual acuity to 20/30 in right eye and 20/80 in left eye. Multimodal images showed a significant improvement in fundus findings in the right eye and a partial improvement in the left eye (Figure 2A and 2B). OCT showed improvement of the intraretinal and subretinal fluid, with disruption of the ellipsoid zone and irregular hyperreflectivities at the level of external retina in both eyes (Figure 2C and 2D) . OCT-A showed a more regular FAZ (Figure 2E and 2F).



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DISCUSSION

Aplastic anemia is a hematologic disorder caraterized by bone marrow failure, leading to pancitopenia and its clinical manifestations. This process is belived to be due to imune-mediated destruction of hematopoietic stem cells, with some trigger factores including drug side effects, viral infection, autoimmunity and radiochemoteraphy.

Ocular manifestations of aplastic anemia include spontaneous hemorrhages (like retinal, vitreous ou subhyaloid hemorrhages) and retinal vein occlusion-like events. In aplastic anemia, the drop in red blood cell count leads to retinal hypoxia, which promotes vasodilation and increased retinal vascular transit. This causes greater vascular turbulence, leading to endothelial injury and hemorrhagic diathesis, aggravated by thrombocytopenia.[1] The presence of a thick choroid is thought to be another predisposing factor for the venous occlusion may create a "compartment syndrome" of the optic nerve by compression.[2]

With improvement of hematological parameters, a gradual improvement of the fundoscopic finding is expected.

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