



PO 11

AN UNUSUAL PRESENTATION OF A BILATERAL TYPE 1 IDIOPATHIC MACULAR TELANGIECTASIA: CASE REPORT AND MULTIMODAL IMAGING

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Introduction and Purpose: Idiopathic macular telangiectasia type 1 (MacTel 1) is characterized by a macular telangiectasia with visible aneurysms classically affecting men unilaterally. The telangiectasis usually involves a two-disc diameter area temporal to the fovea. Macular edema and lipid deposition are characteristic features. Bilateral presentation in a female patient is uncommon. We report a case of a female patient with asymmetric bilateral MacTel1.

Material and Methods: Case report and description of findings in multimodal imaging.

Results and Discussion: A 92-year-old female presented to our hospital with complaints of gradual vision loss that was more pronounced in the right eye. Her past medical history was positive for asthma and dyslipidaemia. Best-corrected visual acuity (BCVA) was 20/100 and 20/32 in the right eye and left eye, respectively. Slit-lamp examination of the anterior segment was unremarkable bilaterally, except for the intraocular lens inside the capsular *bag*. Ophthalmoscopy of the right eye showed small haemorrhages and hard exudates located inferior to the fovea and loss of normal foveal reflex with inferior macular oedema. The left eye showed slight pigmentary changes. Early- and late-phase fluorescein angiography of the right eye demonstrated marked macular ischemia with perfoveal telangiectasia and microaneurysms mainly in inferior and temporal area of the macula with late staining and leakage. The left eye showed the same alterations in superior and nasal area of the macula but less severe. Spectral-domain optical coherence tomography revealed intraretinal fluid and some microaneurysms within the inner retinal layers. Optical coherence tomography angiography (OCTA) revealed parafoveal capillary telangiectasia and capillary dropout as well as decreased vascular density in both superficial and deep capillary plexus, more evident in the right eye and deep capillary plexus. Based on the multimodal imaging and patient's medical history the diagnosis of atypical bilateral MacTel1 was made. The patient was unresponsive to anti-VEGF treatment, in contrast to triamcinolone intravitreal injection. Therefore, she was proposed to place a steroid intravitreal implant. Thus, the visits of an elderly patient to the hospital were significantly reduced.

Conclusion: Bilateral presentation in an old female patient is very rare in MacTel1, as well as the location of the telangiectasis in the nasal area and associated macular ischemia. Other possible disorders that can cause similar presentation must be excluded. The absence of perfusion delay on fluorescein angiography and the fact that they do not respect a single vascular territory exclude paramacular branch occlusion. OCTA seems to be efficient to identify vascular anomalies typical of MacTel1 patients, mostly in this rare and asymmetrical presentations.