A 16-Year-Old Female with Blurry Vision

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Introduction:

A 16-year-old female with no prior medical history presented with blurry vision in her left eye. Initially, the decreased vision was noted during a failed vision test while attempting to obtain a driver's license. Visual acuity was 20/20 in the right eye and 20/40 in the left eye. Fundus examination of the left eye revealed intraretinal hemorrhages and exudation with associated edema centrally. Fluorescein angiography of the left eye demonstrated avascular retina peripherally and central leakage as a result of microaneurysms. The patient underwent a work-up for systemic conditions and hematologic studies yielded a normal CBC, PT, aPTT, and INR. Although coagulopathy studies were negative for lupus anticoagulant, anti-cardiolipin antibody, and showed normal levels of homocystine, Protein S, and Protein C, it did reveal a heterozygous Factor V Leiden mutation.

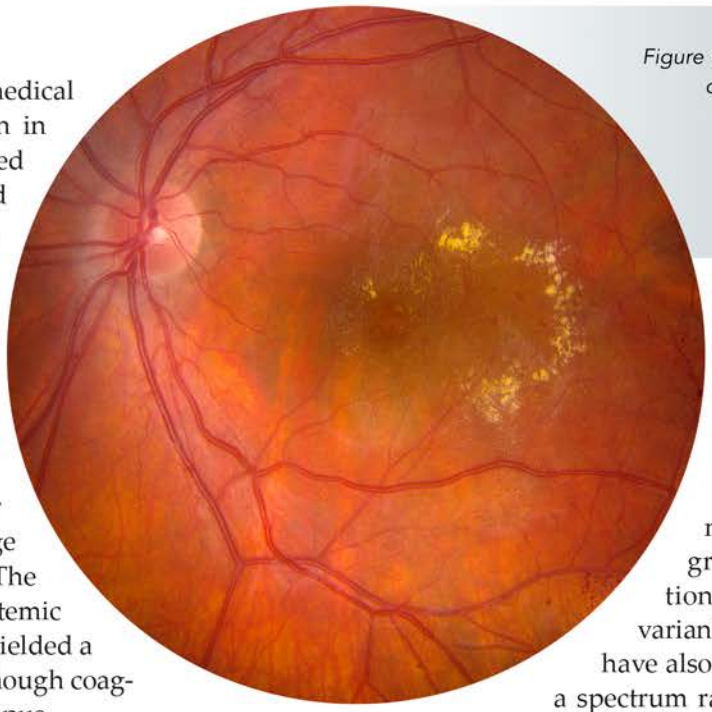

Figure 1. Fundus photograph of the left eye showing central exudation and intraretinal hemorrhages in the temporal macula.

Diagnosis:

The patient was diagnosed with Coats' disease (also known as retinal telangiectasis), which is characterized by abnormal telangiectasias and aneurysms of retinal vessels leading to retinal exudation and potential serous retinal detachment[1]. This condition generally has a unilateral presentation and affects males more than females with no ethnic or geographic associations[2]. Adult-onset variants of Coats' disease have also been reported with a spectrum ranging from milder forms limited to macular abnormalities to findings identical to that in children[3].

Early stages of Coats' disease involve telangiectasis predominantly in the temporal macula and mid-periphery. Vascular abnormalities such as vessel enlargement and radially-oriented aneurysmal dilations surrounding areas of capillary dropout can also be found. Retinal edema and accumulation of lipid exudates in the macula are common causes for vision loss. In later stages,

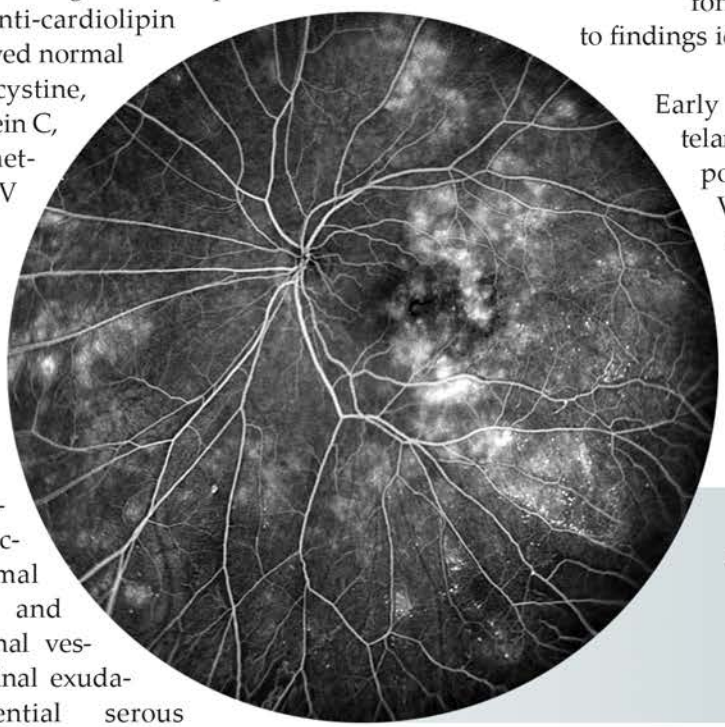

Figure 2. Fluorescein angiography of the left eye highlighting numerous microaneurysms and associated leakage within the macula.

migration and proliferation of retinal pigment epithelial cells in the subretinal space can lead to fibrosis and retinal detachment. Alternatively, serous retinal detachments can occur with progressive exudation[2,4,5]. Fluorescein angiography highlights areas of peripheral nonperfusion, microaneurysms, and telangiectatic capillaries in the temporal macula[6].

Historically, the treatment of choice is laser photocoagulation and/or cryotherapy in early to middle stages of Coats' disease. In late stages, surgical intervention for retinal detachment repair or enucleation for a blind, painful eye may be necessary[7]. In recent literature, anti-vascular endothelial growth factor (VEGF), namely intravitreal bevacizumab, has been reported as an effective adjunctive therapy either alone or combined with other treatment modalities for subretinal fluid and exudation in Coats' disease[8-11]. In their series of patients treated with intravitreal bevacizumab combined with laser and/or cryotherapy, Ramasubramanian et al. demonstrated efficacy of combined treatment with resolution of retinopathy, exudation and subretinal fluid. However, given the formation of vitreoretinal fibrosis in 50% of patients, with a subset subsequently developing tractional retinal detachment, the authors concluded that some caution is advised in employing intravitreal bevacizumab for Coat's disease management.

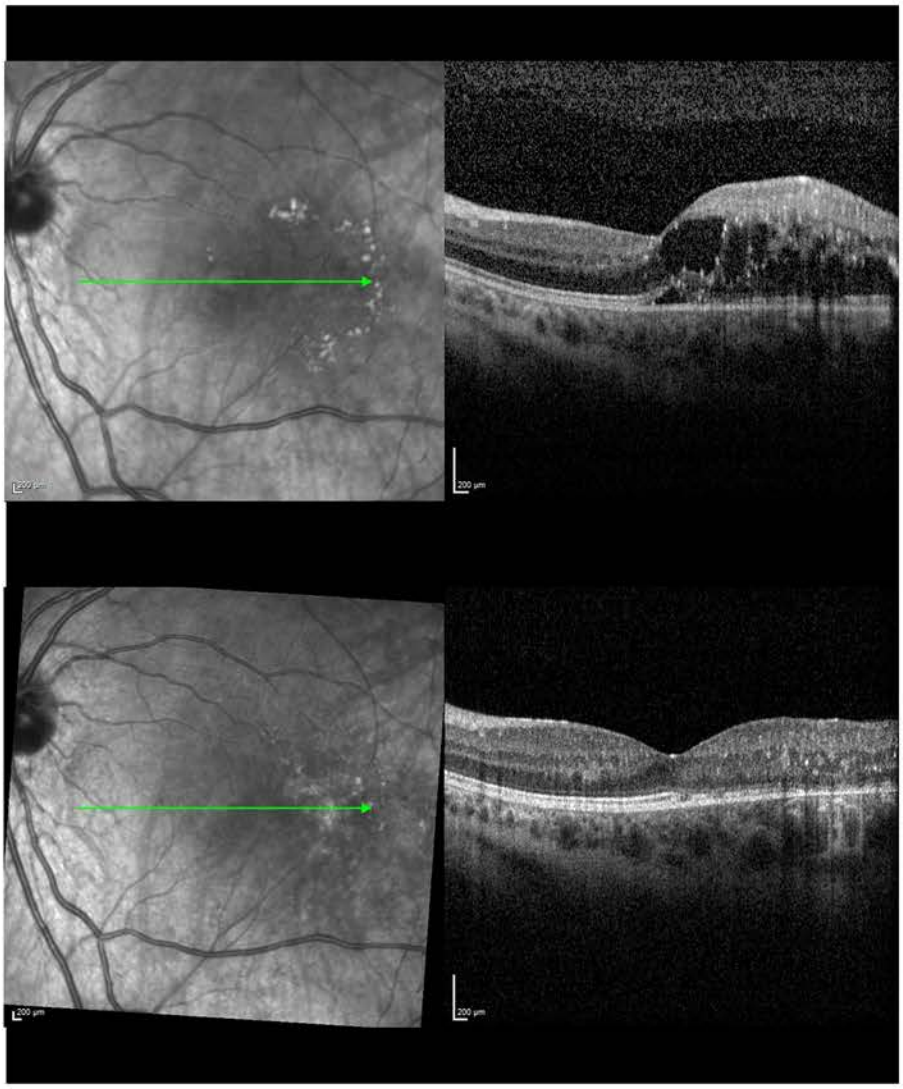

Figure 3. (Top) Initial OCT of the macular with intraretinal edema and subretinal fluid. (Bottom) Resolution of macular edema after combined treatment with peripheral laser photocoagulation and serial intravitreal anti-VEGF injections.

Conclusion:

Our patient was initially treated with peripheral laser photocoagulation and intravitreal bevacizumab in the left eye. Focal laser photocoagulation was added later in an attempt to treat the refractory edema. The anti-VEGF therapy was switched to intravitreal aflibercept with resolution of the edema after multiple injections and repeat focal laser photocoagulation. The patient's best corrected visual acuity currently remains stable at 20/25 in the left eye.

With regards to the finding of a Factor V Leiden allele has been found in a significant proportion of patients with retinal vascular occlusions such as central retinal vein occlusions and branch retinal vein occlusions[12]. In young women, detection of this mutation has implications for appropriate counseling on the risks of thrombosis associated with oral contraceptive use and reproductive planning.

References:

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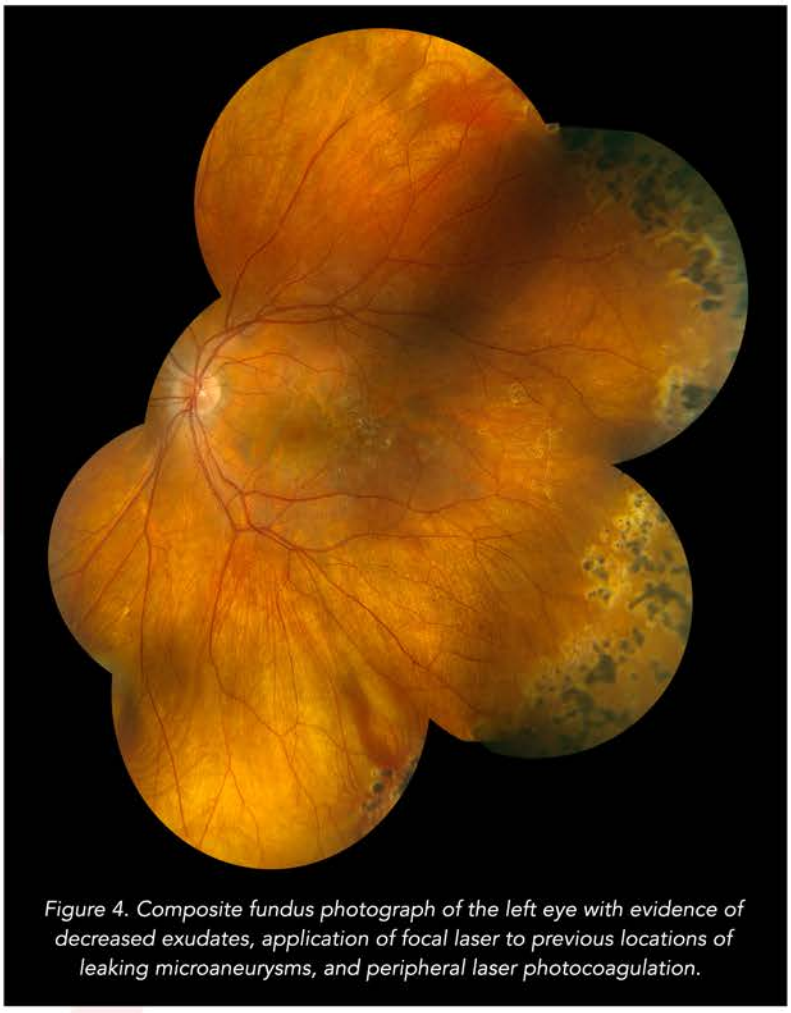

Figure 4. Composite fundus photograph of the left eye with evidence of decreased exudates, application of focal laser to previous locations of leaking microaneurysms, and peripheral laser photocoagulation.

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