Choroidal Neovascular Membrane Secondary to Adult Vitelliform Macular Dystrophy

Abstract

This case outlines the clinical findings associated with adult vitelliform foveomacular dystrophy and the rare development of a choroidal neovascular membrane (CNVM). It will cover the common differential diagnoses as well as treatment options.

Case History

- 74 year old while male
- Presented the clinic with complaints of a dark black spot in the center of his right eye for the past two weeks. He reports his vision is distorted and blurry.
- The patient’s ocular history is positive for adult vitelliform macular dystrophy and pigment dispersion syndrome.
- The patient’s medical conditions include hypertension, hyperlipidemia, coronary artery disease, adrenal cortical adenoma, and eczema.
- Current medications include aspirin, clopidogrel bisulfate, nitroglycerin, and propranolol.

Pertinent Findings

- Corrected visual acuity in the right eye was counting fingers at 18 inches and 20/40 in the left eye. Pupils were equal, round, and reactive to light. No afferent pupillary defect was present.
- Slit lamp examination showed 1+ Krukenburg spindle and 2+ nuclear sclerotic cataracts in each eye. Intraocular pressures measured 16 mmHg OU.
- Fundus examination revealed average optic nerve head cupping at 0.30 OD and 0.35 OS. There were bilateral central yellow lesions in the macula. The right eye had several intra-retinal hemorrhages with apparent edema.
- Ocular coherence tomography (OCT) images showed intra-retinal and sub-retinal fluid in the right eye surrounding the lesion.

Differential Diagnoses

Differential diagnoses for this case include adult vitelliform foveomacular dystrophy, Best’s disease, and age related macular degeneration.

- Patients with adult vitelliform macular dystrophy may be asymptomatic, or present with complaints of mild decreased vision. The condition typically presents between ages 30 and 50. Classic signs include bilateral large, central yellow colored lesion in the center of the macula. Electrophysiological testing shows a normal electroretinography (ERG) and may or may not have a normal electro-oculography (EOG). Patients with vitelliform
typically do not develop vision problems until around age 60, when the retina starts to atrophy.

- Best’s disease is considered the juvenile form of adult vitelliform and lesions are typically present at birth or develop in early childhood. Best’s disease is more visually impairing, with many patients reaching legal blindness by age 50. Best’s has a normal ERG; however the EOG is abnormal, often before vision is compromised.
- Macular degeneration can have a similar age of onset as adult vitelliform, but is more common to present after age 60. Drusen associated with macular degeneration are usually in groups of several lesions and are smaller in size compared to a vitelliform lesion. The lesions tend to increase in both size and number with age.

**Diagnosis and Discussion**

The OCT images confirmed the presence of a choroidal neovascular membrane in the right eye. CNVM causes subretinal fluid accumulation and/or retinal pigment epithelium detachments. Based on the patient’s history of adult vitelliform macular dystrophy, this was determined to be the cause of the net. Only about 20% of patients with vitelliform dystrophy develop a CNVM.

Adult vitelliform macular dystrophy is considered a type of pattern dystrophy. It has an autosomal dominant inheritance pattern. The yellow lesions are composed of lipofuscin. About 10% of affected individuals have multiple lesions. There may be a darker pigmented border surrounding the yellow lesions. Although the disease is bilateral, patients are often only symptomatic in one eye. Visual acuity typically maintains 20/40 or better in the majority of patients.

**Treatment and Management**

There are no effective treatment options for vitelliform lesions. However, if a neovascular membrane develops, treatment may be initiated. Treatment of the net is considered controversial with vitelliform dystrophy because they oftentimes resolve spontaneously without significant impact on acuity.

The patient in this case was referred to a local retinal specialist for treatment of the choroidal neovascular membrane. The patient received Avastin injections in the right eye. Other treatment options for a CNVM include laser if located outside of the fovea. Photodynamic therapy may also be considered for a sub-foveal lesion.

**Conclusion**

While the development of a choroidal neovascular membrane is rare with vitelliform, it is vision threatening. Early detection and treatment increases the visual prognosis in these patients. Home amsler grid monitoring can be useful tool for early detection.
References