



## A 48-Year-Old Female with Nyctalopia

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### Introduction:

A 48 year-old female referred for evaluation night blindness and blurry vision for one week. Her history is notable for multiple gastric bypasses, intestinal revisions, and a cholecystectomy. She has been taking 10,000 units of Vitamin A orally and has had a history of vitamin deficiencies.

### Exam:

Best corrected visual acuity was 20/20- in the right eye and 20/25 in the left eye. Intraocular pressures were within normal limits. No afferent pupillary defect was appreciated. Visual fields were full to confrontation and extraocular motility was full in both eyes. Anterior segment exam was notable for normal findings.

Dilated fundus examination revealed bilateral diffuse yellow retinal flecks (Figure A) along with yellow pigmentary changes of the fovea (Figure B). OCT imaging highlights the presence of outer nuclear layer irregularities (Figure C). Auto-fluorescence reveals focal areas of hyper-autofluorescence but overall normal RPE autofluorescence (Figure D).

### Discussion:

Our patient was sent to obtain urgent lab work in order

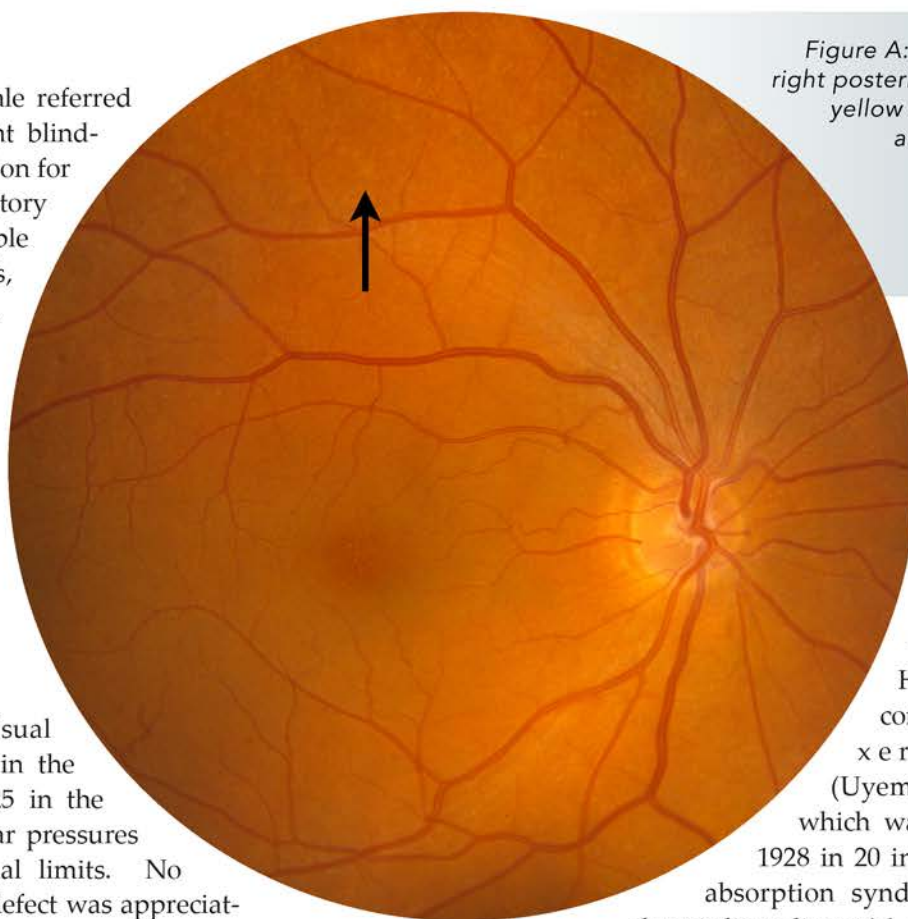


Figure A: Color photo of the right posterior pole. Notice the yellow retinal flecks (arrow) amongst the superior temporal vascular arcade. Identical findings were seen in the left eye.

to assess her vitamin A status. Her labwork revealed a vitamin A level of 5 (normal 38-98). Her gastroenterologist was contacted and follow arranged in the next few days. Her presentation is consistent with fundus xerophthalmicus (Uyemura Syndrome) which was first described in 1928 in 20 individuals with malabsorption syndromes (diet, celiac, chronic liver disease).<sup>1</sup> Disturbances of vitamin A metabolism can have profound effect on the eye, and vitamin A deficiency continues to be a leading cause of blindness in underdeveloped countries and those with a high prevalence of measles.<sup>2</sup> Within developed countries, vitamin A deficiency is much rarer but still seen in adults who underwent gastric bypass, duodenal resection, or cholecystectomy.<sup>3</sup> Patients with chronic vitamin A deficiency can present with a variety of ocular manifestations to include night blindness, corneal or conjunctival xerosis, severe dry eye, corneal ulcerations, or yellow-white retinal spots.<sup>2</sup> Corneal xerosis, dry eye, and ulceration are the result of lack of epithelial differentiation; and it is well known that vitamin A metabolism is crucial for the formation of retinal



within the chemical processes of the photoreceptor. Nyctalopia is a direct manifestation of its deficiency as the rod photoreceptors lack enough retinal to send transmit scotopic stimulation to neural signals. This can be investigated through dark adaptometry, rod scotometry, or electroretinography.

Interestingly, there are two similar retinal conditions that may mimic this diagnosis. Retinitis punctata albescens is characterized by white spots in the fundus, restriction of visual fields, attenuation of retinal vessels, and progressive deterioration of vision.<sup>1</sup> Fundus albi punctatus is associated with childhood night blindness, white fundus spots (macular sparing), normal retinal vessels, and stable visual acuity if given adequate lighting.<sup>1</sup> Both can be differentiated from vitamin A deficiency by relative lack of anterior findings and appropriate systemic levels of vitamin A.

This case highlights the formation of diffuse yellow-white spots in the retina with accompanying night blindness and mild blurring of vision in a patient whose

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E). Although symptoms  
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dismiss if the patient  
presents with normal  
visual acuity and fields,  
a quick review of the  
patient's medical record  
can reveal important sys-  
temic contributions and identify preventable causes of  
blindness. This patient was taking the appropriate vita-  
min A supplementation but do to her lack of bile pro-  
duction (cholecystectomy) and lack of available duode-  
num for absorption (bypass of duodenum), she still  
remained vitamin A deficient. Assuming that because  
the patient was taking oral vitamin A supplementation,  
she could not be deficient, would have incorrectly man-  
aged this patient. Additional oral supplementation or  
intramuscular vitamin A injections may be required to

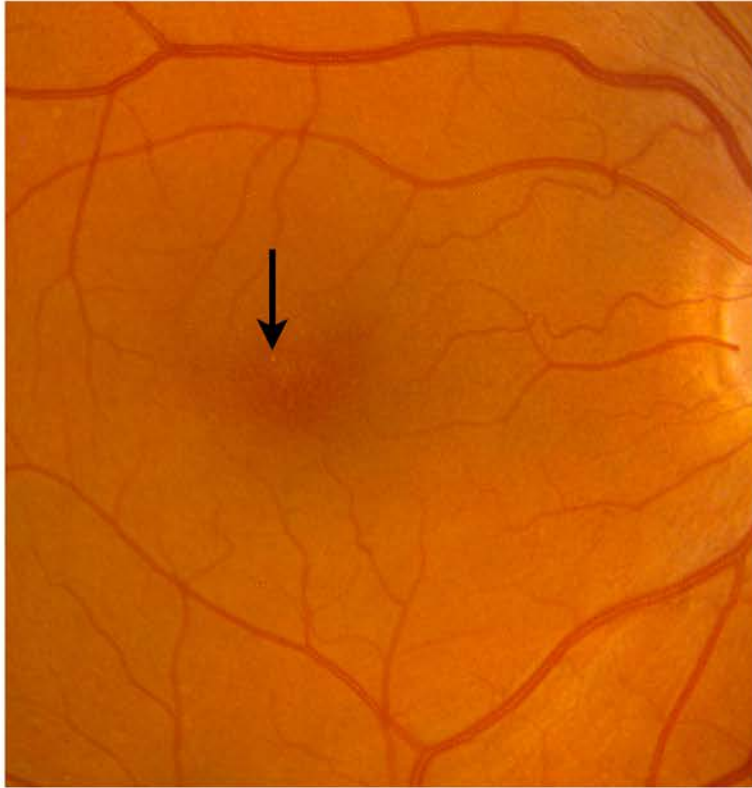


Figure B: Color photo of the right macula. Notice the yellow punctate discolorations (arrow) of the fovea. Identical findings were seen in the left eye.

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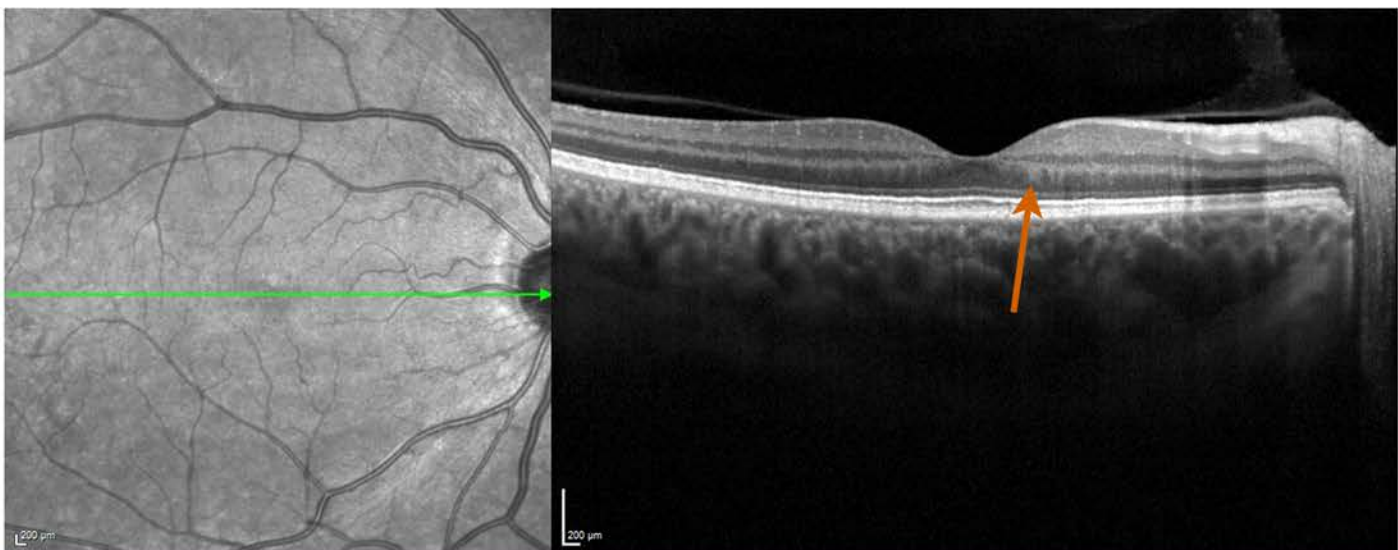


Figure C: OCT of the right macula. Notice the irregularities of the outer nuclear layer (arrow) in the parafoveal region. The ellipsoid zone, RPE, and choroid appear grossly normal. Identical findings were seen in the left eye.

improve her vitamin A levels. In most cases, proper supplementation and normalization of vitamin A levels will slowly reverse xerosis, dry eye, nyctalopia, and retinal changes. Supportive care to include lubrication therapy is necessary until normal eye anatomy returns.

## References:

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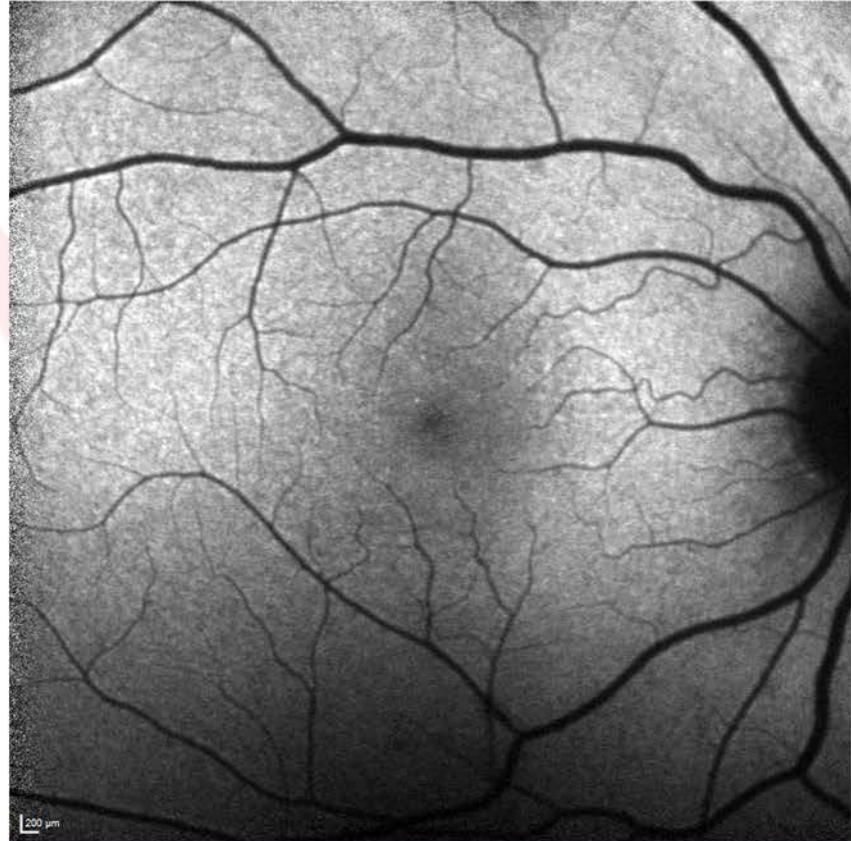


Figure D: Autofluorescence of the right macula. Notice a few focal points of hyper-autofluorescence but overall normal auto-fluorescent response of the RPE.

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