

Case reports

Lessons from practice

Tunnel vision and night blindness in a 52-year-old man

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MJA 2011; 195: 287–288
doi: 10.5694/mja11.10292

Clinical record

A 52-year-old man presented to the ophthalmology clinic with a 3-day history of tunnel vision and night blindness (nyctalopia). He reported recently needing to wear a headlamp to see adequately in dim lighting, notably while walking to work early in the morning.

The patient had a history of hypertension, hypercholesterol-aemia and osteoporosis. Twice in the previous 2 years, he had seen a neurologist for symptoms of lower limb paraesthesiae. At that time, peripheral nerve conduction studies gave normal results, and a clinical diagnosis of peripheral neuropathy secondary to vitamin B₁₂ deficiency was made. Supplementation with vitamin B₁₂ and folate was initiated. He also described a 3-hour episode of tunnel vision 2 years before presentation.

Examination revealed an otherwise well man with a body mass index of 27 kg/m² (ie, in the overweight range). His best corrected visual acuity was 6/18 in the right eye and 6/9 in the left eye.

Anterior segment examination revealed Bitôt spots, and no staining of the cornea with fluorescein (Figure). Pupillary examination revealed a relative afferent pupillary defect of the right eye, grade 1/4, indicating optic nerve dysfunction. Ocular motility and intraocular pressures were normal. Fundoscopy showed normal peripheral retinal pigment epithelium, and normal discs and maculae. Humphrey visual field 30-2 testing showed peripheral field constriction. Results of the Farnsworth–Munsell D-15 hue test for colour vision were normal. Dark-adapted electroretinography (ERG) revealed bilaterally decreased amplitudes, consistent with diminished rod function. The results of computed tomography and magnetic resonance imaging of the brain and orbits, using contrast, were normal.

Investigations revealed a markedly reduced vitamin A level (0.1 µmol/L [reference range, 1.4–4.0 µmol/L]). Results of a full blood count, electrolyte levels, and liver and thyroid function tests were normal. Vitamin B₁₂ and folate levels were high, consistent with supplementation. Normal results of a malabsorption screen, including levels of fat-soluble vitamins D, E and K, were obtained, and test results for parietal cell and intrinsic factor antibodies were negative. Test results for antiendomysial antibodies were also negative, as were those for IgA and IgG antigliadin antibodies, thus excluding coeliac disease.

A diagnosis of xerophthalmia (dryness of the conjunctiva and cornea) was made on the basis of the symptoms of nyctalopia

and tunnel vision, as well as the findings of bilateral Bitôt spots, hypovitaminosis A, and diminished rod function on ERG.

A dietary history revealed that the patient had suffered from self-diagnosed food intolerance for most of his life. Since childhood, his diet had consisted exclusively of potatoes, white bread (but he refused to eat butter and margarine) and cola. He described nausea and vomiting after eating any other foods.

The patient underwent multidisciplinary evaluation by a neurologist, gastroenterologist, psychiatrist, dietitian and psychologist. Dietary supplements were prescribed and cognitive behaviour therapy was initiated. The patient was treated with 100 000 IU of vitamin A daily, given orally for 3 days, followed by 50 000 IU for 14 days.

Humphrey visual field results and visual acuity improved within 3 days of commencing treatment. After a month, visual acuity had improved to 6/6 in both eyes, and the Bitôt spots had completely resolved. The relative afferent pupillary defect was no longer present, and Humphrey visual field and ERG results had returned to normal.



Bitôt spots on the temporal limbus of the left eye.

Vitamin A deficiency is a systemic illness which can increase an individual's risk of blindness, severe infections and mortality.¹ It is rare in developed countries like Australia.² Vitamin A deficiency and xerophthalmia in developed countries are reported in patients with malabsorption syndromes or liver disease, in those who have had major gastrointestinal surgery, in people with alcoholism, and in those with anorexia nervosa and other psychiatric disorders.^{3–6} This case is unique because our patient did not have any of these risk factors.

This case also highlights the importance of clinicians regularly taking a thorough dietary history, especially in the context of other indications of nutritional insufficiencies, such as osteoporosis and previous vitamin B₁₂ deficiency.

Vitamin A is a fat-soluble vitamin found as retinol in dairy products and as provitamin A carotenoids in some fruits and green leafy vegetables.^{1,7–9} The first clinical evidence of vitamin A deficiency often occurs in the visual system and produces xerophthalmia. The ocular changes of

Lessons from practice

- Vitamin A deficiency is rare in developed countries like Australia.
- Visual symptoms can often be the first manifestation of a systemic illness, such as vitamin A deficiency.
- A thorough nutritional screen, including a dietary history, is essential, especially in the context of any other nutritional deficiency.
- Early diagnosis and treatment of vitamin A deficiency can be curative, preserving vision and life. ◆

xerophthalmia generally occur in a predictable pattern, as described by the World Health Organization.¹

The first stage of xerophthalmia is nyctalopia, the result of defective regeneration of retinal rhodopsin.^{3,7-9} This responds rapidly to vitamin A therapy, and patients often report regaining their scotopic vision (vision under low-light conditions) within 24–48 hours after the initiation of treatment.^{1,3,7,9}

The second stage of xerophthalmia is conjunctival xerosis, or drying, in which loss of goblet cells and squamous metaplasia of the conjunctiva occur.^{1,3,7,8} Bitôt spots are bilateral triangular patches of keratinised epithelium at the temporal limbus and nasal limbus of the eye.^{1,3,7} Colonisation of these patches by saprophytic bacilli, including *Corynebacterium xerosis*, results in a foamy appearance.^{1,3,8} Conjunctival xerosis and Bitôt spots respond to vitamin A therapy in 1–5 days.^{1,3}

Corneal xerosis occurs primarily because of instability of the tear film as goblet cells are lost, with subsequent keratinising metaplasia of the ocular surface.^{1,8} Corneal xerosis usually responds to vitamin A therapy in 1–2 weeks.^{1,9} If left untreated, corneal drying can result in ulceration, with subsequent scarring and keratomalacia.^{2,3,7,8} Keratomalacia is a rapidly progressive and irreversible liquefactive necrosis of the cornea that can ultimately lead to perforation and spontaneous loss of intraocular contents.^{1,3,9}

Uncommonly, in vitamin A deficiency, the xerophthalmic fundus exhibits yellow and white dots peripherally, sometimes associated with a corresponding scotoma (area of diminished vision).^{1,7-10} These changes respond well to treatment, often returning to normal within 2–4 months.^{1,8,9}

The diagnosis of vitamin A deficiency is made by a directed history and clinical findings, and confirmed by the presence of a low serum vitamin A level, and an abnormal electroretinography result.^{3,9} In our case, the diagnosis was made accordingly, and all the pathological findings resolved with vitamin A therapy.

Vitamin A deficiency is usually treated by administering 200 000 IU of vitamin A orally on two successive days, followed by an additional dose 1–4 weeks later.¹ Administration of intramuscular vitamin A is reserved for patients with malabsorption, or those who are unable to tolerate medications orally.¹ Ocular vitamin A has not been shown to be beneficial because of the systemic nature of vitamin A deficiency.⁷

In summary, we present a unique case of vitamin A deficiency and xerophthalmia in an unlikely candidate for malnutrition. Hypovitaminosis A can have potentially devastating visual and systemic effects. To prevent this happening, it is important to maintain a high degree of suspicion, especially in the context of other nutritional deficiencies.

Acknowledgements: We thank Rahul Dubey, Michael C Wei, Philip J Townend, Tanya Karacanjic, Ravjit Singh and Ashish Agar for their valuable contributions to this case report.

Competing interests: No relevant disclosures.

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