ELSEVIER

Nutrition 21 (2005) 630–633

NUTRITION

www.elsevier.com/locate/nut

International ward rounds

Adult blindness secondary to vitamin A deficiency associated with an eating disorder

Antonio Augusto Velasco Cruz, M.D., Ph.D.[a], Flávia A. Attié-Castro, M.D.[a], Sandra L. Fernandes, M.D.[b], Jussara Fialho F. Cortes, M.D.[b], Paulo de Tarso P. Pierre-Filho, M.D.[a], Eduardo Melani Rocha, M.D., Ph.D.[a], and Júlio Sérgio Marchini, M.D., Ph.D.[b,*]

[a] *Department of Ophthalmology, Division of Nutriology, School of Medicine of Ribeirão Preto, University of São Paulo, Ribeirão Preto, São Paulo, Brazil*

[b] *Department of Clinical Medicine, Division of Nutriology, School of Medicine of Ribeirão Preto, University of São Paulo, Ribeirão Preto, São Paulo, Brazil*

Manuscript received November 1, 2004; accepted December 12, 2004.

Abstract

Objective: We examined an adult patient who lost one eye due to severe keratomalacia secondary to self-induced vitamin A deficiency.

Methods: This case report provides a clinical, ophthalmologic, and laboratory description in addition to a review of the medical literature.

Results: A 33-y-old woman with a 17-y history of an eating disorder presented with bilateral conjunctival xerosis, an infected corneal ulcer in the right eye and a large descemetocele in the left eye. Laboratory and clinical findings were consistent with vitamin A deficiency. Despite a tectonic penetrating keratoplasty, her left eye perforated and had to be eviscerated. In parallel, vitamin A replacement improved her clinical status and the ocular findings in her right eye.

Conclusions: The present report indicates that vitamin A deficiency secondary to eating disorders should be considered in the differential diagnosis of patients with severe dry eye and corneal ulceration.

Keywords: Eating disorder; Vitamin A deficiency; Xerophthalmia; Blindness

Introduction

Corneal xerosis and its progression to keratomalacia due to vitamin A deficiency are often seen in school-age children from certain parts of West Africa [1]. In Western countries, severe vitamin A deficiency due to dietary habits has been reported only sporadically [2–4], and most cases are secondary to malabsorptive syndromes, such as bypass surgery [5], short bowel [6] celiac disease [7], and pancreatitis [8,9].

To our knowledge, this is the first description of an adult patient who lost one eye due to keratomalacia secondary to an eating disorder.

Case report

A 33-y-old Brazilian woman was admitted with a complaint of discomfort, mucous discharge, and severe bilateral visual loss in both eyes. Her medical history revealed bizarre eating habits. For 17 y she refused to consume meat, fruits, or vegetables and ate only rice cooked in water, potatoes, and beans. Ocular antecedents revealed an episode of bilateral corneal ulcers 7 y previously. At that time vitamin A deficiency was diagnosed but the patient was lost to follow-up and continued with her dietary habits.

Physical examination demonstrated a cachectic, malnourished-appearing woman whose weight was 33.6 kg and height was 141 cm. Upon examination of her skin, the patient was found to have poor turgidity, follicular hyperkeratosis, scaling, and xerosis. Marked wasting of muscle mass was noted. Psychiatric evaluation attributed the mal-

* Corresponding author. Fax: 55-16-633-6695.
E-mail address: jsmarchini@fmrp.usp.br (J.S. Marchini).

0899-9007/05/$ – see front matter
doi:10.1016/j.nut.2004.12.003

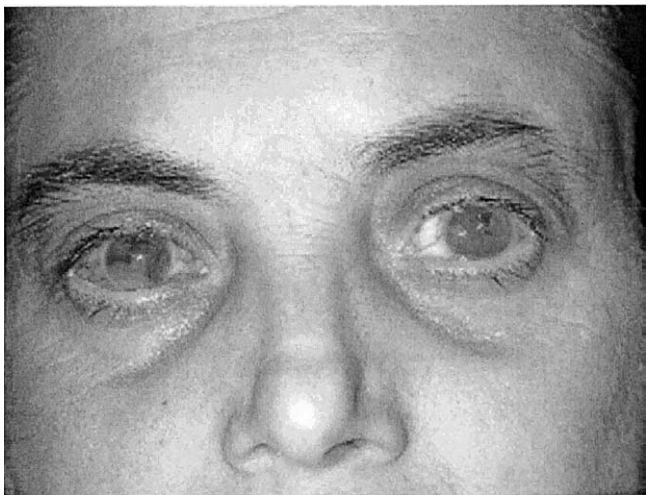

Fig. 1. Corneal opacity in the right eye and progressive corneal melting in the left eye.

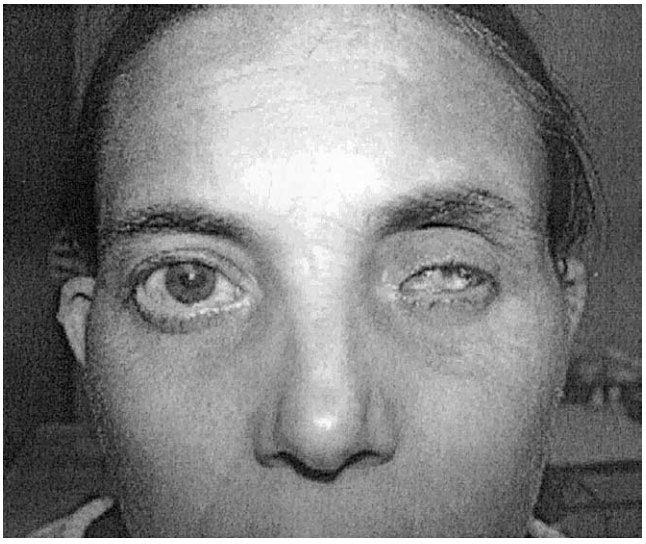

Fig. 2. Right eye after vitamin A supplementation and control of infection.

nourishment to an atypical eating disorder because anorexia nervosa and bulimia nervosa were rejected as diagnoses.

Ocular examination demonstrated that visual acuity was limited to hand motion for each eye. Marked conjunctival hyperemia with dry and keratinized epithelium was present bilaterally. Slit-lamp biomicroscopy showed a central corneal ulcer with a stromal infiltrate, an intact anterior chamber, and a hypopyon in the right eye. The left eye had a large descemetocele but intact anterior chamber (Fig. 1). Vascularization was present in both corneas and the lids were erythematous and edematous.

Microbial culture from corneal scrapings grew *Streptococcus viridans* and *Staphylococcus epidermidis*, both sensitive to cephalothin. Her serum vitamin A level was low (0.11 μM; normal range 0.88–6.98 μM), as was her plasma vitamin C level (0.1 μg/dL, normal value 0.3 μg/dL); her hemoglobin level was 9.9 g/dL, hematocrit was 30%, white blood cell count was 11 800 with 21% lymphocytes, and albumin level was 32 g/L (normal range 35–55 g/L).

A diagnosis of vitamin A deficiency with severe corneal involvement associated with protein-calorie malnutrition was made. The patient was initially treated with fortified cephalothin topically (100 mg/mL) and supplemented with vitamin A (200 000 U/d for 2 d and subsequently 200 000 U in the second week). A penetrating keratoplasty was performed in the left eye without success, and 2 mo after surgery, the eye had to be eviscerated after complications related to wound healing and re-epithelization delay that culminated with endophthalmitis and vision loss. The right cornea improved dramatically with control of the infection and complete healing of the ulcer (Fig. 2). Six months later, the patient had gained 6 kg. Because she was not able to make meaningful changes in her eating habits, she was prescribed multivitamin supplementation. On her final follow-up visit, visual acuity was 20/40 in the right eye, her weight was 40 kg, her serum level of vitamin A was 3 μg/dL, and her serum level of vitamin C was 0.3 μg/dL.

Discussion

Prolonged dietary vitamin A deficiency may cause a severe multiorgan, multisymptomatic, and life-threatening disease. Nightblindness is the earliest symptom of vitamin A deficiency. The condition may progress to conjunctival and corneal xerosis, corneal ulceration, and subsequent eye loss due to keratomalacia (corneal ulceration up to one-third the corneal thickness) [1,10].

Although the role of vitamin A in retinal photoreceptor function is well known, the physiopathologic mechanisms of hypovitaminosis A that lead to keratomalacia and disruption of eye globe integrity remain unclear [10]. The corneal manifestations of vitamin A deficiency may be secondary to epithelial defects, mucin deficiency and tear film instability [10]. Despite the exact mechanism remaining unknown, xerosis and keratinization of focal areas of the cornea have been experimentally shown to be associated with corneal ulcers and a rapidly developing keratomalacia followed by perforation in end-stage cases [10]. Eyelid alterations can be manifested as blepharitis or meibomian gland dysfunction [6].

Individuals who are deficient in vitamin A are commonly deficient in protein, and it is believed that transport of vitamin A into cells depends on adequate protein availability [11]. In addition, experimental studies have shown that an inflammatory response is more aggressive in vitamin A deficiency [12]. Therefore, the combination of vitamin A and protein deficiency and the associated infection may have been responsible for the severe inflammation presented in this case.

Table 1
Vitamin A deficiency in adults due to self-inflicted dietary restriction

Reference	Year	Age (y)	Duration of dietary restriction (y)	Sex	Clinical presentation			Outcome
					Vitamin A serum level (normal values)*	Visual Acuity	Eye findings	
Gombos et al. [13]	1970	58	Not stated	F	19.9 μg/dL (44 μg/dL)	OD: LP	OD: cornea opaque and vascularized, central perforation	OD: hand motion
						OS: LP	OS: corneal ulcer, opacity and neovascularization	OS: 20/40
Bors and Fells [14]	1971	25	5	M	13 IU/dL (150 IU/dL)	OD: LP	Bilateral mid-stromal vascularization, half-depth ulcer	OD: 20/80
						OS: 20/300		OD: 20/20
Smith et al. [15]	1975	27	5	F	10 mEq/dL	Not stated	Crusts on the lid margin, dry conjunctiva, corneas thickened and opaque	Death 3 d after admission
Oliver [2]	1986	39	7	M	0.56 μM (2.13–4.64 μM)	Not stated	OD: Marginal corneal perforation, iris prolapse, flat anterior chamber, hypopyon	OD: 20/60
							OS: desmetocele, formed anterior chamber, hypopion	OS: 20/40
Jaworowski et al. [16]	2002	24	2	F	0.7 μg/dL (25–200 μg/dL)	20/30 OU	Conjunctival xerosis, keratoconjunctivitis sicca with coarse punctate keratitis	Conjunctiva and corneas returned to normal
Present case		33	17	F	3 μg/dL (25–200 μg/dL)	Hand motion OU	OD: infected central corneal ulcer	OD: 20/40
							OS: large descemetocele; neovascularization OU	OS: evisceration

F, female; LP, light perception; M, male; OD, right eye; OS, left eye; OU, both eyes

* The term *vitamin A* is a generic description for retinoids with biologic activity of retinal. To convert micrograms per deciliter of vitamin A to the Système International unit, micromoles, multiply by 0.0349. To convert international units per deciliter of vitamin A to micrograms per deciliter, multiply by 0.3 [20].

Treatment with vitamin A supplementation results in the resolution of early ocular signs and the normalization of vitamin A level. Treatment usually consists of 200 000 IU of oral vitamin A in oil for 2 d, followed by an additional dose 1 to 2 wk later, but parenteral administration may be needed for cases related to non-absorptive syndromes [6,10]. Surgery may be necessary for corneal perforations, and in those cases established techniques remain useful to restore corneal integrity. Options include tissue adhesive for small perforations, patch grafts for medium-size defects, and penetrating keratoplasty for large defects [13].

A Medline search of the past 37 y disclosed only five previous reports of keratomalacia due to self-inflicted dietary restriction (Table 1) [2,14–17]. In all cases there was a severe deficiency of vitamin A resulting from a longstanding inadequate diet. Keratomalacia was present in four patients whose these dietary habits lasted at least 5 y. The variable ocular outcome may due to different metabolic and environmental backgrounds.

Corneal melting in vitamin A deficiency is a sign that the patient is severely malnourished and an indication that the condition may be fatal [16]. As presented in Table 1, the consequences for vision can also be serious. The final visual acuity of one eye was hand motion and we were unable to maintain the integrity of the left eye of our patient. Of the six patients listed in Table 1, only one man had a classic psychiatric condition (schizophrenia) [2]. For all remaining five patients (four women), bizarre dietary habits were associated with unclassified psychological factors (our patient) or related to pressure to lose weight [17] or to beliefs that their diets were healthier [14–16].

In the face of the current prevalence of cultural diet restrictions, ophthalmologists should be sensitive to detect mild and moderate ocular surface signs that may indicate vitamin A deficiency and consider eating disorders as an underlying cause of these signs. Early intervention is essential to minimize morbidity and prevent long-term sequelae from longstanding corneal and conjunctival xerosis [18].

It should be noted that unsupervised vitamin A replacement may lead to severe visual toxic effects such as papilledema. Clinical manifestations of vitamin A toxicity commonly appear after months of overdosage and are

accompanied by headaches, somnolence, dryness and desquamation of the skin, hepatomegaly and hypomenorrhea [19]. Patients with vitamin A deficiency must be carefully observed during replacement therapy.

References

[1] Sommer A. Vitamin A deficiency and xerophthalmia. Arch Ophthalmol 1990;108:343–4.

[2] Olver J. Keratomalacia on a 'health diet'. Br J Ophthalmol 1986;70: 257–360.

[3] Ramsay AS, Sabrosa N, Pavesio CE. Bitot's spots and vitamin A deficiency in a child from the UK. Br J Ophthalmol 2001;85:372.

[4] Rodrigues MI, Dolman, CH. Blindness in an American boy caused by unrecognized vitamin A deficiency. Arch Ophthalmol 2004;122: 1228–9.

[5] Brown GC, Felton SM, Benson WE. Reversible night blindness associated with intestinal bypass surgery. Am J Ophthalmol 1980; 89:776–9.

[6] Cella W, Urbano AP, Vinhandelli WS, Donatti M, Rocha EM. Xerophthalmia secondary to short bowel syndrome. J Pediatr Ophthalmol Strabismus 2002;39:125–7.

[7] Alwitry A. Vitamin A deficiency in coeliac disease. Br J Ophthalmol 2000;84:1079–80.

[8] Suan EP, Bedrossian Jr, RC Eagle Jr, EH Laibson PR. Corneal perforation in patients with vitamin A deficiency in the United States. Arch Ophthalmol 1990;108:350–3.

[9] Nicolai U, Rochels R. Bilateral severe keratomalacia after acute pancreatitis. Cornea 1993;12:171–3.

[10] Sommer A. Xerophthalmia and vitamin A status. Prog Retin Eye Res 1998;17:9–31.

[11] Sommer A. Nutritional factors in corneal xerophthalmia and keratomalacia. Arch Ophthalmol 1982;100:399–403.

[12] Wiedermann U, Chen XJ, Enerback L, Hanson LA, Kahu H, Dahlgren UI. Vitamin A deficiency increases inflammatory responses. Scand J Immunol 1996;44:578–84.

[13] Donzis PB, Mondino BJ. Management of noninfectious corneal ulcers. Surv Ophthalmol 1987;32:94–110.

[14] Gombos GM, Hornblass A, Vendeland J. Ocular manifestations of vitamin A deficiency. Ann Ophthalmol 1970;2:680–4.

[15] Bors F, Fells P. Reversal of the complications of self-induced vitamin A deficiency. Br J Ophthalmol 1971;55:210–4.

[16] Smith RS, Farrell T, Bailey T. Keratomalacia. Surv Ophthalmol 1975;20:213–9.

[17] Jaworowsky S, Drabkin E, Rozenman Y. Xerophthalmia and undiagnosed eating disorder. Psychosomatics 2002;43:506–7.

[18] Walsh BT, Devlin MJ. Eating disorders: progress and problems. Science 1998;280:1387–90.

[19] Grant WM. Description of drugs, chemicals, plants and venoms, and their effects on the eyes. In: Grant WM, editor. Toxicology of the eye. 2nd ed. Springfield: Charles C. Thomas; 1974, p. 1084–5.

[20] Olson JA. Vitamin A, retinoids and carotenoids. In: Shils ME, Olson JA, Shike M. editors. Modern nutrition in the health and disease. Volume 1. Philadelphia: Lea & Febiger; 1994, p. 287–307.