Rare disease

Nyctalopia: the sequelae of hypovitaminosis A

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Summary

We present a case of night blindness due to vitamin A deficiency in a gentleman who had previously undergone a pancreaticoduodenostomy 10 years ago. The deficiency was attributed self-cessation of his pancreatic supplements without being aware of the possible consequences.

The diagnosis and management of this patient is presented here, along with a discussion of similar cases in the literature.

BACKGROUND

Vitamin A is an essential fat soluble vitamin needed for the function of various body systems. It is a vital component for vision where it is required for the synthesis of visual pigments in rods and cones. In addition, vitamin A also plays a role in photo signal transduction, as well as in the maintenance of epithelial surfaces on the cornea and conjunctiva. Vitamin A is also required in building immunity, gene transcription and maintaining skin health.

Well-documented ophthalmological complications of vitamin A deficiency include night blindness and xerophthalmia.^{1 2} This can progress to permanent blindness if left untreated. In the other body systems, hypovitaminosis A can cause the upper respiratory and urinary tract epithelium to be replaced by keratinising squamous cells. This will predispose patients to the development of pulmonary and urinary infections, as well as the formation of urinary stones. Immune impairment is a well-known consequence that leads to higher mortalities from common infections such as measles and bacterial gastroenteritis in developing countries.

The causes of vitamin A deficiency include malnutrition, malabsorptive conditions and diseases that impair vitamin A metabolism. Vitamin A deficiency due to malnutrition is usually seen in patients from developing countries, whereas the latter can manifest among the elderly and the chronically sick populations of developed nations.

In this case report, we highlight a patient who developed night blindness 10 years after a pancreaticoduodenostomy. The cause of this was the self-cessation of pancreatic enzyme supplementation a year before the development of symptoms. There is literature regarding vitamin A deficiency after a pancreaticoduodenostomy, especially where there is no recurrence of carcinoma. In addition, vitamin A deficiency is a rare entity in developed countries, and thus, many clinicians might not be familiar with its sequelae. We hope that this case report can highlight the importance of maintaining adequate vitamin A levels after similar surgeries so that potentially devastating ocular complications can be reduced.

CASE PRESENTATION

A 61-year-old man with a history of pancreaticoduodenostomy (Whipple's procedure) for papillary mucous neoplasm, presented with a 2-month history of night blindness with associated skin dryness, weight loss and anaemia.

He is a regular attendee at the hospital's diabetic eye clinic, and his symptoms were first noticed during one of his annual reviews. He complained of poor vision in dim conditions over the past 2 months. Specifically, he reported having to turn on his lights in the house during day light hours and giving up driving at night. He also described widespread skin dryness throughout his body.

In 1998 he was diagnosed with a pre-malignant pancreatic lesion and subsequently underwent a Whipple's procedure. Postoperative complications of cholechojejunal stricture and enterocutaneous fistula were treated surgically.

He was commenced on pancreatic enzymes supplementation following his surgery. However, the patient decided to cease the enzymes because he felt he was developing nausea from it.

His other medical history included diabetes, depression and an episode of severe pneumonia requiring intensive care admission.

On examination he was cachectic with diffuse skin dryness. Ophthalmological examination revealed visual acuity of 6/9 and 6/6 (both aided), right and left, respectively. He had mild cataracts bilaterally, mild diabetic maculopathy and normal visual fields. Testing of colour vision and pupillary responses were normal.

Other systems were normal.

INVESTIGATIONS

Haematological results of note were the diminished levels of the liposoluble vitamins. His vitamin A level was <0.3 µmol/l, vitamin D 18 nmol/l and vitamin E <5 µmol/l. Other haematological results were unremarkable.

Electroretinography (ERG) showed absent rod responses bilaterally. The maximal rod-cone ERG is bilaterally grossly abnormal with substantially reduced a- and b-wave

Figure 1 ERG showing absent rod function bilaterally.

Figure 2 ERG showing normalisation of rod function after treatment. The patient is now symptom free on his pancreatic supplements.

amplitudes. The ERG results confirmed the absence of rod function (figure 1).

The Arden ratios were also substantially reduced for each eye (1.22 (R) 1.32 (L)).

TREATMENT

A diagnosis of vitamin A deficiency secondary to malabsorption was made. The patient was restarted on his pancreatic enzymes and prescribed oral vitamin A. He was monitored with regular ophthalmology and nutritional follow-up to ensure compliance was maintained.

OUTCOME AND FOLLOW-UP

The patient's symptoms improved within 2 months of treatment with vitamin A and pancreatic enzyme supplementation. Vitamin A assay showed normalisation of his level at 0.9 $\mu\text{mol/l}$. The electroretinograph was repeated, showing a significant improvement with the Arden ratios being within normal ranges for each eye (3.03 (R) 2.69 (L)).

There was a significant improvement in amplitude for the rod ERG and maximal rod-cone ERG (figure 2).

DISCUSSION

Vitamin A deficiency is a common cause of blindness in developing countries. Around 2.8 million preschool children are affected with vitamin A deficiency in over 60 countries and subclinical vitamin A deficiency is considered a problem for at least 251 million people.³ However, it is rare in developed countries and is usually only encountered in patients with malabsorptive states or liver disease.

Being a liposoluble nutrient, vitamin A is dependent on the dietary lipids for transport. The reduction of dietary lipids into its more simple forms for transport is carried out by the pancreatic enzyme lipase. As a result, any pancreatic impairment can limit vitamin A absorption and transport.

In a pancreaticoduodenectomy, the head of the pancreas, a portion of the bile duct, the gallbladder, the duodenum and sometimes part of the stomach is removed. The surgery can result in reduced enzymatic production and, hence, necessitates dietary supplementation in patients.

Here, our patient presented with nyctalopia, which is one of the earliest symptoms of vitamin A deficiency. He had stopped taking his pancreatic supplements about a year before he started developing the symptoms. He did not have other ocular complications seen in prolonged vitamin A deficiency, which includes conjunctival and corneal xerosis, Bitot's spots, severe punctate keratitis, corneal scarring and ultimately keratomalacia in severe cases.

The other symptoms that he presented with, that is, the weight loss and the skin dryness, can also be attributed to vitamin A deficiency. It is also possible that he might have had subclinical vitamin A deficiency when he was in ICU for severe pneumonia.

Here, the diagnosis was made promptly and allowed treatment to be instituted before his symptoms progressed. However, it can easily be missed, especially in developed countries where vitamin A deficiency is rare.

Vitamin A deficiency has been previously documented in patients suffering from chronic pancreatic insufficiency (alcoholic pancreatitis, cystic fibrosis) and in intestinal bypass surgery.^{2 4-6} These studies have had success with both oral and intramuscular treatment of vitamin A. However, it is evident that the normalisation of vitamin A levels is achieved quicker with intramuscular treatment.^{3 5}

Our case emphasises that patients who have had a pancreaticoduodenostomy procedure need to be monitored closely from a nutritional point of view. Before undergoing any pancreatic surgery, the possibility of blindness from lack of vitamin A intake must be discussed with the patient. In particular, there is a need to stress the importance of compliance with nutritional supplements after surgery, even many years down the track.

Learning points

- ▶ Vitamin A deficiency can occur in patients who have had pancreaticoduodenostomy, even many years after the surgery.
- ▶ Night blindness is an early sign of vitamin A deficiency – recognition of this and instituting early treatment can prevent devastating late complications.
- ▶ Vitamin A deficiency can be treated effectively with either oral and intramuscular treatment.
- ▶ Patients who undergo a pancreaticoduodenectomy or other similar procedures need to be counselled about the importance of maintaining their nutrition.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Sommer A.** Xerophthalmia, keratomalacia and nutritional blindness. *Int Ophthalmol* 1990;**14**:195–9.
2. **Lee WB,** Hamilton SM, Harris JP, *et al.* Ocular complications of hypovitaminosis a after bariatric surgery. *Ophthalmology* 2005;**112**:1031–4.
3. **Stephenson LS,** Latham MC, Ottesen EA. Global malnutrition. *Parasitology* 2000;**121** Suppl:S5–S22.
4. **Lopez-Rodriguez N,** Faus F, Sierra J, *et al.* Night blindness and xerophthalmia after surgery for morbid obesity. *Arch Soc Esp Oftamol* 2007;**82**:133–6.
5. **Spits Y,** De Laey JJ, Leroy BP. Rapid recovery of night blindness due to obesity surgery after vitamin A repletion therapy. *Br J Ophthalmol* 2004;**88**:583–5.
6. **Appaswamy S,** De Silva D, Tyagi A, *et al.* Reversible night blindness in a patient with neuroendocrine tumour of pancreas. *Eye (Lond)* 2005;**19**:720–2.

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