Case Report

Decompensated Superior Oblique Palsy Secondary to Bilateral Nutritional Optic Neuropathy Following Bariatric Surgery

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Keywords

Superior oblique palsy · Nutritional optic neuropathy · Strabismus

Abstract

Optic neuropathy can occur secondary to nutritional deficiencies in patients who have undergone bariatric surgery. We present a unique case of a 39-year-old man, claiming to be generally healthy, who presented with intermittent vertical diplopia and bilateral decreased vision in each eye. Visual acuity was 6/18 in the right eye and 6/12 in the left eye. Ishihara testing was defective for both eyes. Automated visual fields showed a severe generalized reduction in sensitivity in both eyes. The patient had a left head tilt and a right intermittent hypertropia of 30 prism diopters in primary position. CT of the orbits revealed a right superior oblique of small caliber. On further questioning, the patient admitted to a history of bariatric surgery 7 years prior to presentation with failure to take any nutritional supplements. Blood work demonstrated deficiencies in folate, thiamine, and copper. Within 6 months of initiating nutritional supplements, the vision in each eye was markedly improved and the diplopia resolved. There was an associated normalization of thiamine and copper, but folate levels remained low. We believe that the nutritional deficiency caused a bilateral optic neuropathy and the resulting vision loss precipitated a manifestation of a congenital superior oblique palsy that had previously just been a phoria. The case emphasizes the importance of considering occult sensory etiologies of acquired strabismus.

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Introduction

Bariatric surgery is associated with a high prevalence of nutrient deficiencies, with patients being predisposed to many vitamin and mineral deficiencies [1]. Numerous studies have demonstrated that nutritional deficiencies following bariatric surgery can result in decreased vision due to optic neuropathy [2, 3]. We present a unique case of a decompensated congenital superior oblique palsy (SOP) secondary to presumed bilateral optic neuropathy. To our knowledge, this is the first reported case of nutritional optic neuropathy manifesting as a decompensated strabismus.

Case Presentation

A 39-year-old man presented to our neuro-ophthalmology clinic complaining of intermittent vertical diplopia, occurring at various times of the day, with increased frequency and duration over a year. Initially, he claimed to be generally healthy and indicated that he did not take any medications. Best-corrected visual acuity was 6/18 in the right eye and 6/12 in the left eye. Ishihara testing was defective for both eyes – right eye saw control plate but 0/8 test plates and left eye saw control plate but 2/8 test plates. Pupils were equal with no relative afferent pupillary defect. Intraocular pressure was 16 mm Hg in both eyes. No abnormalities were found in the eyelids or anterior and posterior segments of the eyes, including both optic nerve heads. The patient had a left head tilt, a right intermittent hypertropia of 30 prism diopters in primary position (shown in Fig. 1), and a positive Parks-Bielschowsky 3 step test. Optical coherence tomography demonstrated normal macula and retinal nerve fiber layers for each eye. Automated visual fields showed marked generalized depressions for each eye, with superimposed superior partial arcuate defects and inferior nasal steps for each eye (shown in Fig. 2).

Further investigations were carried out. Thyroid function tests, antithyroid peroxidase antibody, and thyroglobulin antibody assays all came back normal. A CT scan of the head and orbits with contrast demonstrated a thin right superior oblique muscle (shown in Fig. 3), raising the suspicion of a long-standing right SOP.

On repeat questioning, the patient admitted undergoing Roux-en-Y gastric-bypass (RYGB) surgery 7 years prior to presentation without any subsequent nutritional supplementation. Laboratory investigations revealed deficiencies of folate (1.80 ng/mL, normal 3.10–20.50),

Fig. 1. Ocular movements in different gazes, showing right hypertropia in primary position (5). The hypertropia increases in left gaze (8).

Fig. 2. 24-2 SITA-fast automated perimetry at time of presentation reveals bilateral generalized reduction in sensitivity (total deviation plots) with superimposed bilateral superior arcuate defects and inferior nasal steps.

Fig. 3. CT scan of the head and orbits with contrast, coronal plane, demonstrating a thin right superior oblique muscle.

vitamin B1 (19 µg/L, normal 28–85), and copper (67 µg/dL, normal 70–140). There were normal levels of vitamin B12, homocysteine, methylmalonic acid, and zinc. Electrophysiology revealed no evidence of vitamin A retinopathy.

Six months following initiation of nutritional replacement therapy, which included multivitamins with various components and nutritional replacements, the patient reported a resolution of his diplopia. On exam, visual acuity in each eye was 6/7.5. Color vision testing was still impaired for each eye. Automated visual field testing was essentially normal for the right eye and showed only an inferior nasal step for the left eye. There was a right hyperphoria of 25 prism diopters in primary position and right inferior oblique overaction. The optic nerves had a normal appearance. An updated OCT study showed extremely mild signs of thinning in the RNFL bilaterally.

On exam 2 months later, the visual acuity had improved to 6/6 in each eye. Right eye color vision had moderately improved, and left eye color vision had nearly fully recovered. While the magnitude of vertical strabismus was essentially the same, it manifested as a phoria without diplopia rather than an intermittent tropia. Repeat laboratory investigations revealed normal levels of vitamin B1 (44 µg/L) and copper (120 µg/mL). The folate level remained low (2.00 ng/mL).

Discussion

Obesity has grown to epidemic proportions, with more than 78 million adult Americans suffering from obesity in 2010 [3]. Bariatric surgery is an effective weight loss option for morbidly obese patients, with approximately 228,000 operations performed annually in the USA [4]. Bariatric surgery affects weight loss in a malabsorptive, restrictive, or combined mechanism. Macro- and micronutrient deficiencies are becoming more common in patients that have undergone bariatric surgery, a problem previously considered one of the underdeveloped world. The degree of nutritional deficiency associated with bariatric surgery is linked to the type of surgery performed. At 5 years post-surgery, nutritional deficiencies were reported in 28, 70, and 87% of adjustable gastric banding, sleeve gastrectomy, and RYGB patients, respectively [5]. Frequency additionally depends on the patient's nutritional status before surgery, and the patient's compliance to post-surgery vitamin supplementation. The RYGB is one of the most commonly performed bariatric procedures and carries one of the greatest risks for post-surgery nutritional deficits [1]. The high prevalence of nutrient deficiencies after bariatric surgery makes lifelong nutritional monitoring and supplementation essential [6].

Bariatric surgery can, in mid to long term, induce ophthalmic complications that can affect almost every component of the optic system [7]. There are several reports of neurological syndromes occurring after bariatric surgery linked to micronutrient and vitamin deficiencies including Wernicke's encephalopathy, polyneuropathy, acute polyradiculoneuropathy, posterolateral myelopathy, optic neuropathy, and myopathy [8]. A disruption in mitochondrial oxidative phosphorylation and factors leading to ganglion cell toxicity has been proposed as the mechanisms by which these deficiencies cause optic neuropathies [9].

Congenital SOP is the most common cause of hypertropia followed by thyroid eye disease [10]. Patients present with an ipsilateral hypertropia and a head tilt to the contralateral side, as was the case in our patient. Our patient also had a thin right superior oblique muscle on CT scan, suggesting a long-standing SOP. Due to the urgency of the situation, the patient was immediately referred for CT scan. Once we saw the thinning of the right superior oblique muscle on CT scan, it provided us with a specific diagnosis of long-standing SOP, therefore, we did not examine other features, such as measuring fusional amplitudes that are typically performed when assessing for a long-standing SOP. In the case of our patient, we believe that bariatric surgery-associated optic neuropathy triggered a manifestation of a congenital SOP due to decompensated fusional compensation triggered by decreased sensory visual input.

The body's reserves of different vitamins and minerals take different times to become depleted before the onset of symptoms. Vitamin B1/Thiamine takes a few weeks to months, vitamin B12 takes more than a year, and copper reserves take 3 years on average and up to over 20 years [11]. Our patient's symptoms of diplopia appeared 6 years after surgery, suggesting that it was the copper deficiency that provoked the optic neuropathy. There are a number of reports implicating copper deficiency as a cause of nutritional optic neuropathy [12, 13].

We suspect that the reason for the lack of a detectable relative afferent pupillary defect was that the optic neuropathy affected the right and left eye symmetrically, as seen in the mean deviations of the initial visual field test. Furthermore, nutritional optic neuropathy may have overlapping mechanisms with hereditary mitochondrial optic neuropathy, where afferent pupillary defects have been reported to be less detectable due to a suspected sparing of the intrinsically photosensitive retinal ganglion cells [14].

The normal retinal nerve fiber layer thickness of each eye at the time of presentation is consistent with the known literature on optic neuropathies. Kupersmith et al. [15] showed

that thinning of the RNFL could not typically be detected before 3 months after disease onset. Therefore, the RNFL thickness will typically be normal in the first months after the onset of vision loss. Ganglion cell layer analysis does show earlier deterioration [15], but the technology was not available at the time of the patient's presentation. Although treatment was started 6 months after presentation, this was probably too late to stop damage that had already occurred, as was shown by the OCT RNFL and as the follow-up visual fields showed.

A possible limitation of this report is the fact that no brain MRI was performed. Therefore, we cannot altogether rule out the possibility that the onset of the diplopia was due to a focal brain lesion. We believe that this possibility is extremely unlikely because (1) the thin right superior oblique accounted for the strabismus, (2) the vitamin deficiency provided a mechanism for the decreased vision and decompensated binocular fusion, and (3) initiation of nutritional supplementation improved the vision and alleviated the diplopia.

It is also important to emphasize that on presentation the patient omitted a major medical history detail, which was revealed only on repeat questioning. As so it is important to be aware of the significance of accurate medical history and on being skeptical when there is a gap between the history and examination findings. To our knowledge, this is the first case report of nutritional optic neuropathy manifesting as a decompensated strabismus.

Conclusion

To our knowledge, this is the first case report of nutritional optic neuropathy manifesting as a decompensated congenital SOP. Our case not only highlights the importance of a detailed medical history but also contributes to the limited literature on nutritional optic neuropathies following bariatric surgery.

Statement of Ethics

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest Statement

The authors report no conflicts of interest.

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Author Contributions

Eleanor Nche and Ravid Ben-Avi: manuscript writing and editing. Ari Shemesh: patient management, data collection, and manuscript editing. Joshua M. Kruger: patient management and data collection, manuscript writing and editing. All coauthors read and approved the final version of the manuscript.

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