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Case Report

Sclerectomy Reverses Nanophthalmic Optic Neuropathy

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Keywords

Choroidal thickness · Nanophthalmos · Optic neuropathy · Peripapillary pachychoroid syndrome · Sclerectomy

Abstract

Introduction: Nanophthalmos is characterized by a short axial length, a thick choroid, and a thick sclera. Unilateral symptomatic disc swelling in nanophthalmos presents both a diagnostic and a therapeutic challenge. ***Case Presentation:*** A healthy 59-year-old man reported a two-week-long abrupt vision reduction in his right eye. 20/100 best spectacle (+17.25 diopter) corrected visual acuity, unilateral widespread disc enlargement, central scotoma, and a slight color vision disruption without an afferent pupillary defect were among the positive findings in the right eye. Workup for neuro-ophthalmology was negative. Numerous consultations did not suggest any form of treatment for the patient. Review of the optical coherence tomography (OCT) indicated a small, crowded optic nerve head and substantial diffuse choroidal thickening with dome-shaped temporal peripapillary area with choroidal expansion. In addition to circumferential anterior four-quadrant 95%-deep sclerectomy from recti insertion to the vortices, radial nasal posterior sclerotomy reaching the optic nerve sheath was performed on the patient. After the procedure, 2 weeks later, the patient's vision returned, and it persisted until the 6-month follow-up. By OCT, the two eyes were comparable as far as disc contour and nerve fiber layer thickness. ***Conclusion:*** This form of sclerectomy, which aims at decompressing the oncotic choroidal pressure, is an effective treatment for compressive optic neuropathy in the context of nanophthalmos. Could sclerectomy assist in treating other optic neuropathies associated with peripapillary pachychoroid?

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Introduction

In otherwise healthy people with a negative thorough workup, unilateral disc enlargement coupled with visual loss poses a diagnostic and therapeutic challenge. We need to identify new processes causing disc compaction and treat it appropriately after ruling out the usual causes of unilateral disc swelling (optic neuritis, nonarteritic anterior ischemic optic neuropathy (NA-ION), infectious or inflammatory neuropathies, and orbital compressive lesions). When extensive systemic and ocular workup fails to identify a particular etiology, unilateral optic disc swelling is described in the specific context of nanophthalmos.

Case Report

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. This 59-year-old slender, healthy Caucasian male schoolteacher reported painless visual loss with a 2-week central scotoma in his right eye. He had no prior medical history (sleep apnea, Raynaud's phenomenon, systemic hypotension or hypertension, smoking, alcohol consumption, thyroid or heart problem, and migraine) and was not using any medications. The refraction was +17.25 diopter in the right eye and +17.00 diopter in the left eye. Intraocular pressures were 18 mm Hg right eye and 12 mm Hg left eye. Best spectacle-corrected visual acuity was 20/100 and 20/40, respectively, in the right and left eyes. There was a partial loss of color vision in the right eye (Ishihara color plates were 8/10 in the right and 10/10 in the left eye), but no pupillary defect. Exam results showed an elevated disc with telangiectatic vessels in the right eye and a small optic disc in the left eye, as shown in Figure 1. The axial length of 16.28 mm right and 16.12 mm left (IOL Master 700; Carl Zeiss, Jena, Germany). In the right eye's visual fields, a significant central scotoma is visible in Figure 1. Optical coherence tomography (OCT) imaging of the right optic nerve head showed peripapillary edema and mild loss of the nerve fiber layer. A dome-shaped peripapillary retina was accompanied by a localized peripapillary retinoschisis without subretinal fluid. Central macular thickness measured 395 and 398 microns right and left eyes. Prominent choroidal folds bilaterally were reflected by the presence of undulations at the level of retinal pigment epithelium, Bruch's membrane, and choriocapillaris. Buried drusen and thickened choroid were also noted: subfoveal and temporal peripapillary choroidal thickness being, respectively, 607 μm and 594 μm right eye versus 534 μm and 566 μm left eye. The diameter of the left optic disc measured 1,008 μm in the left eye (PLEX® Elite 9000 swept-source OCT (Carl Zeiss AC, Jena, Germany). Thorough radiologic and blood examination to rule out inflammatory and neuro-ophthalmic causes were negative (including CBC, HbA1c, TSH, Vit B12, folate, CRP, ESR, MR brain with gadolinium, MR orbit with gadolinium).

Over the course of a month, the ocular status was unchanged. We provided the patient with a surgical option based on our preliminary diagnosis of nanophthalmic optic neuropathy caused by compression by a thick peripapillary pachychoroid restricting the blood flow in a crowded small optic nerve head with optic nerve drusen, leading to the end stage of NA-ION.

Surgical Procedure

The patient consented for surgery 45 days after the onset of his ocular symptoms. Under general anesthesia, lateral canthotomy was done to allow better exposure of the deep-set small orbit. Tenon capsule was very thick and vascular. A 360-degree peritomy was performed. The recti were exposed and bridled. The sclera behind the recti was severely thickened (2.5–3 mm) (Fig. 2). Sclerectomy (95%) [1] was completed at once by determining the depth of the incision as indicated by a violaceous hue at the scleral bed. We avoided

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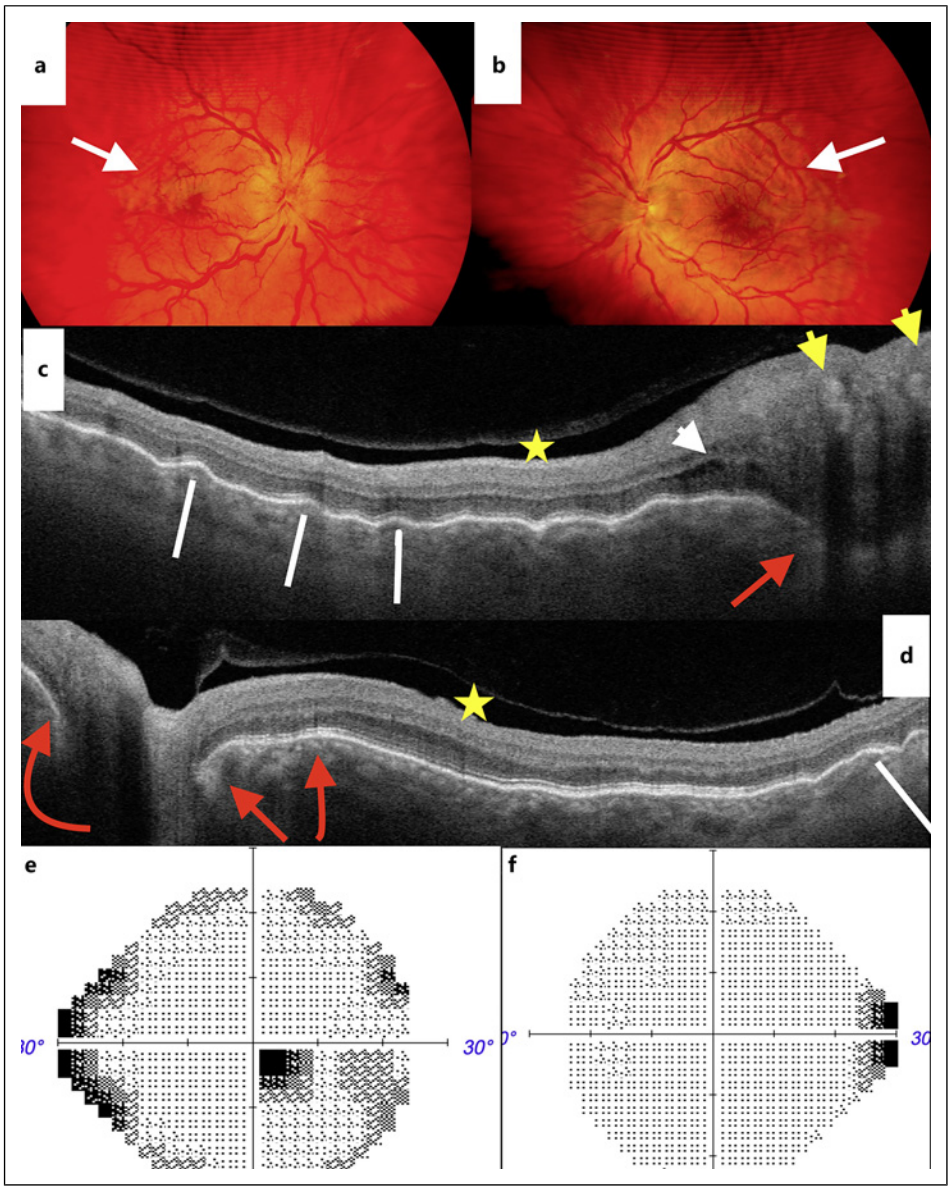

Fig. 1. a Wide angle fundus photograph of the right eye shows diffuse disc edema and choroidal folds (white arrow). **b** Wide angle fundus photograph of the left eye demonstrates a small optic disc and choroidal fold (white arrow). **c** A 12 mm horizontal OCT scan of the right macula reveals absence of foveal depression (yellow star), peripapillary retinoschisis (white arrowhead), disc edema, buried disc drusen (yellow arrowheads), undulated retinal pigment epithelium (white lines), diffusely thickened choroid, and dome-shaped peripapillary area (red arrow). **d** A 12 mm horizontal OCT scan of the left macula shows a small disc size (1,008 μm), absence of foveal depression (yellow star), diffusely thickened choroid and dome-shaped peripapillary area (red arrows) and subtle choroidal folds (white line). **e**, **f** Automated visual fields revealed a central scotoma in the right eye.

penetration into the suprachoroidal or subretinal spaces. A circumferential four-quadrant 95% depth sclerectomy from muscle insertion to the vortices [1] was completed. In addition, a nasal posterior radial sclerectomy reaching the optic nerve sheath is shown in Figure 2. Dexamethasone-tobramycin ointment was used twice daily for 1 week. Two weeks post-operatively, there was full recovery of vision (20/40 both eyes), color perception (10/10 color

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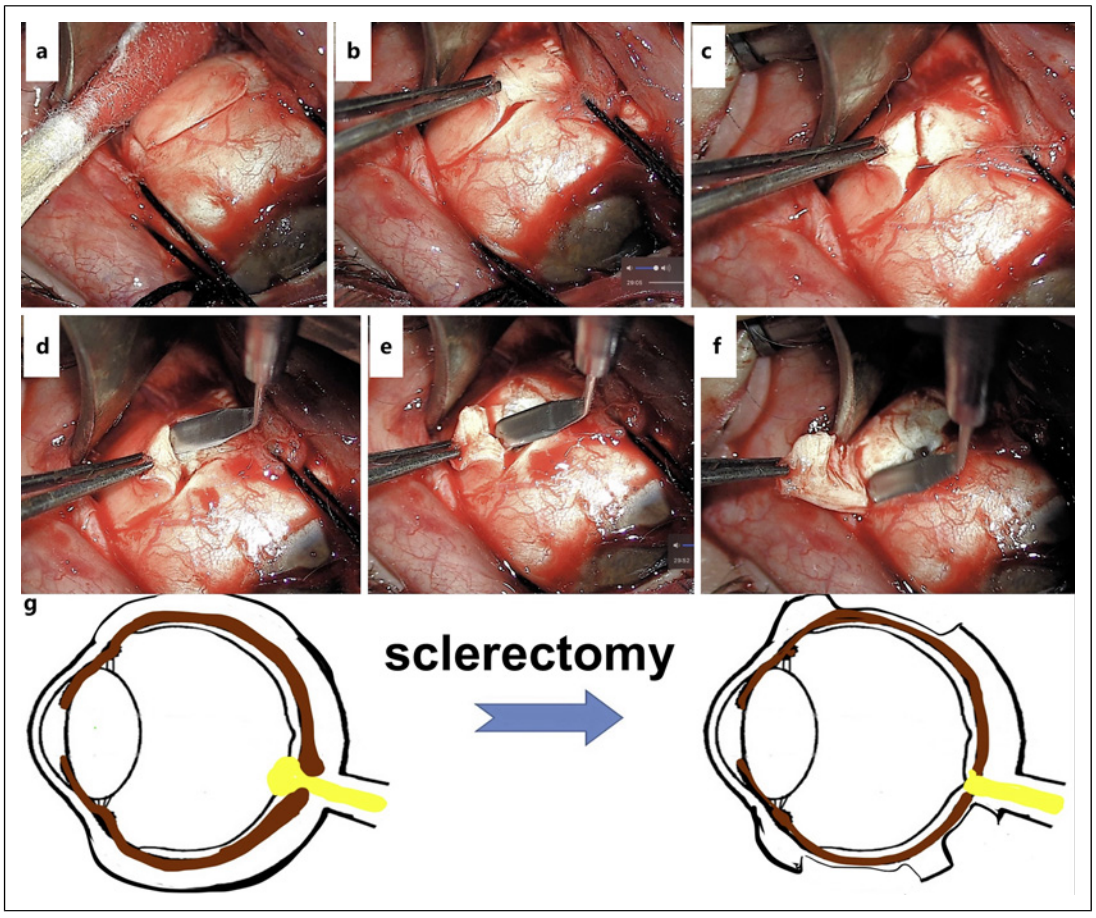

Fig. 2. a–f Operative sequences. Sclerectomy is initiated with a hockey blade behind the rectus muscle insertion in each quadrant by incising the thick sclera very deep till a violaceous hue appears. Then the flap is pulled up and the dissection is continued along the same plane using a bevelled crescent knife. **g** Following circumferential sclerectomy, radial nasal sclerectomy is also done reaching the optic nerve head sheath. This allowed resolution of the optic disc edema and a decreased peripapillary choroidal thickness as seen in Figure 3.

plates both eyes), and resolution of disc edema in the right eye (Fig. 3). No recurrence was noted 6 months postoperatively. The CARE Checklist has been completed by the authors for this case report and is attached as an online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000537829).

Discussion

Compressive optic neuropathy resulted in the present case from nanophthalmos-related peripapillary pachychoroid and responded to sclerectomy-based decompression surgery allowing relief of the increased choroidal pressure from congestion in a very small compartment and in a dwarf eye as seen in Figure 4. The choroidal vasculature surrounds the compartment through which the retinal ganglion cells pass; therefore, it is possible that localized swelling could result in a secondary compartment syndrome that causes compression [2, 3]. Recent meta-analysis evidence linked peripapillary choroidal thickening with NA-ION [3]. Using computer modeling, Feola et al. [2] examined the effect of choroidal

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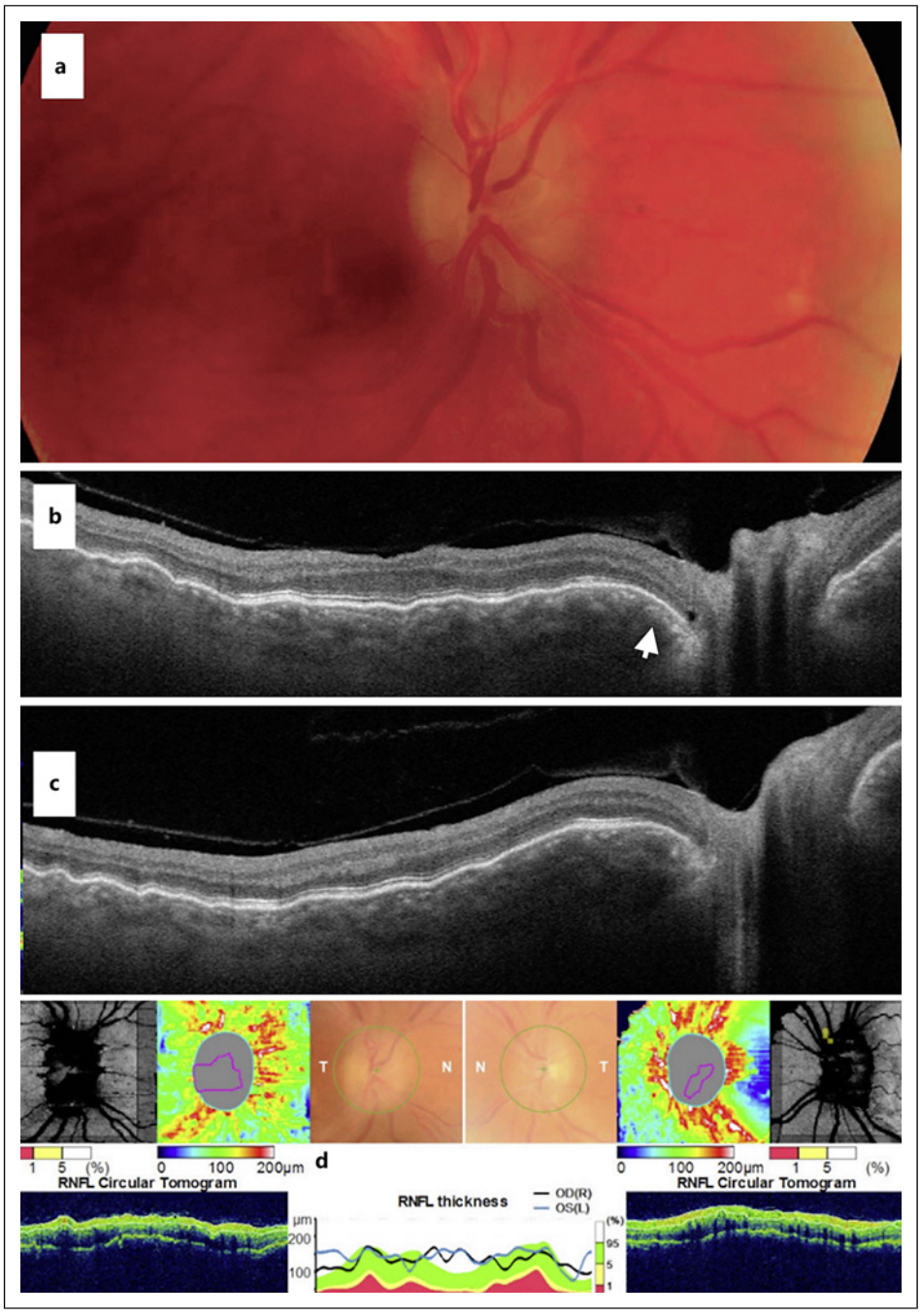

Fig. 3. **a** Complete resolution of the right disc edema is visible on fundus photography 20 days postoperatively. Baseline (**b**) and at 2-month post-surgery (**c**) horizontal OCT scan through the right disc revealed dome-shaped peripapillary area (white arrow). The peripapillary choroidal thickness decreased from 726 µm to 645 µm. **d** The two discs have comparable nerve fiber layer analysis denoting no permanent damage from the compressive optic neuropathy, aside from subtle superior temporal and inferior temporal thickness asymmetry.

expansion on tissue deformation at the optic nerve head as seen in Figure 4. According to that study, choroidal expansion may be a potential factor in the deformation of the optic nerve head tissue, which is similar to the neuro-ocular syndrome associated with spaceflight

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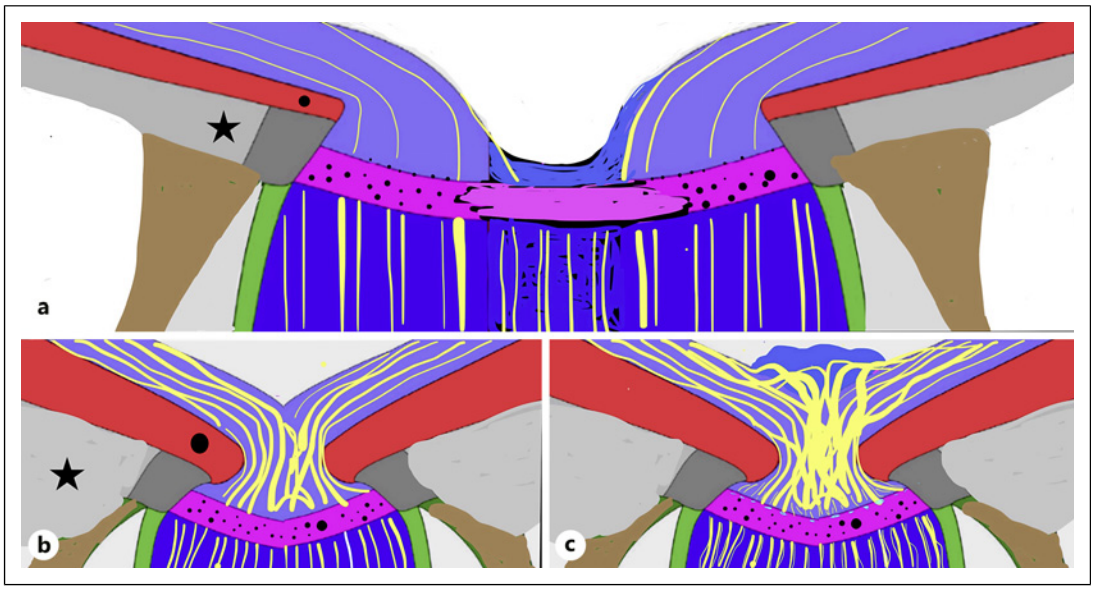

Fig. 4. a Schematic diagram of a normal eye with normal sclera (black star) and choroid (black dot). A peripapillary sector was adopted from Feola et al. [2]. **b** Schematic diagram of a nanophthalmos eye with small disc, thickened peripapillary choroid (black dot) and very thick sclera (Star). **c** Schematic diagram of a nanophthalmos eye with compression of the optic nerve.

(choroidal folds and disc edema) [4]. This is because choroidal expansion can lead to strains in the prelaminar neural tissue that are noticeably greater than those caused by increased intraocular or intracranial pressures. Some researchers have lumped many disorders with thickened choroid around the optic disc under the entity of peripapillary pachychoroid syndrome. Intervortex venous anastomoses are shown to be present in the macular region in central serous retinopathy and in the peripapillary region in the peripapillary pachychoroid syndrome, all sharing a common choroidal venous overflow [5].

A shift in the choroid tonicity is one potential cause of choroidal enlargement (cold exposure – vasospasm [6]; morbid obesity [7]). Alteration in vascular permeability, which can result in proteins moving into the extracellular matrix, is another potential cause of choroid thickening such as in uveitis and uveal effusion. The human choroid autoregulation is one more potential method [3]. It is well known that certain medications such as sildenafil can lead to increased choroidal perfusion and thickness [8].

Extensive literature search yielded 3 similar cases [9–11]: PubMed, Google scholar and Web of Science using MeSH terms “optic neuropathy” AND “nanophthalmos” or “uveal effusion”. The first case was a 48-year-old man with pseudo-phakic left uveal effusion accompanied by mild optic swelling [9] who responded within 1 month to scleral window surgery [9]. Axial length measured 15.76 mm in the left eye. Four-quadrant half thickness 4 × 4 mm sclerectomy was created with a central 1-mm full-thickness sclerotomy in inferior windows. Visual acuity improved from 20/400 to 20/50 with decrease in the disc edema at 1 month postoperatively. However, visual loss and disc edema recurred 3 months postoperatively. Four-quadrant 50% depth sclerectomy was enlarged to 6 × 10 mm. Three months following the second surgery, the choroidal effusions flattened, but visual acuity remained at the 20/150 level. Review of the fundus photograph, B-scan, and visual field revealed mild nasal disc swelling, the presence of disc drusen, and peripheral visual field constriction corresponding to the peripheral annular retinal detachment. Another case of nanophthalmos with severe optic disc edema was described by Sarraf and Schwartz [10]. That report

described a 51-year-old obese man with borderline diabetes and hypercholesteronemia, left severe 3-quadrant visual field depression, bilateral radial macular folds, bilateral disc drusen, and bilateral diffuse optic atrophy. Positive workup included a slightly elevated serum ANA and elevated CSF protein levels. The axial length was 20.13 mm right eye and 20.04 mm left eye. Hyperopia measured +7.25 diopter right eye and +6.75 diopter left eye. At the 4-year follow-up, the ocular exam remained unchanged. No OCT was available at that time. A related third case was reported by Rathinam et al. [11]. A 46-year-old healthy woman presented with sudden painless visual impairment in the left eye, with swollen left optic disc, small right optic disc and negative investigations for ischemic or inflammatory markers [11]. NA-AION in the context of a hypoplastic disc was confirmed by fluorescein angiography [11]. The degree of hyperopia was not mentioned.

The current case did have optic disc drusen. Nanophthalmos eyes have a high incidence of disc drusen [12]. Disc drusen have also been implicated in the pathogenesis of NA-AION in small discs by further compromising the blood supply, an entity described as "Optic Disc Drusen Associated Anterior Ischemic Optic Neuropathy" [13]. Anyway, disc drusen and NA-AION share a common etiology of a small size disc [14]. Dome-shaped macula is most noted in high myopia and uncommon in high hyperopia. One case of +5 diopter hyperopia was noted to have dome-shaped macula [15]. Dome-shaped peripapillary area in the current case falls within the spectrum of peripapillary pachychoroid syndrome.

Nanophthalmos is characterized by vascular thickened Tenon capsule, thickened sclera, thickened choroid, small optic disc, optic disc drusen, absent foveal avascular zone, and a high risk for uveal effusion, resulting in secondary retinal detachment or angle closure glaucoma. Choroidal congestion and impaired scleral permeability increase the osmotic pressure in the suprachoroidal space. Currently, the most long-lasting cure for uveal effusion is circumferential scleral window technique that produces rapid resolution of uveal effusion in nanophthalmic eyes [1].

Compressive optic neuropathy with dome-shaped peripapillary pachychoroid, narrow optic canal, and disc drusen in the context of severe thickening of the nanophthalmic sclera was successfully reversed by deep sclerectomy. Sclerectomy is one standard long-lasting procedure in nanophthalmos uveal effusion and is extended to secondary optic neuropathy from nanophthalmos. Future research may shed light on the role of sclerectomy in the therapy of peripapillary pachychoroid with resulting optic neuropathy or NA-AION.

Statement of Ethics

Ethical approval is not required for this study in accordance with national guidelines. Written informed consent was obtained from the patient for publication of the details of the medical case and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

A.M., S.U., and R.H. contributed to the design of the manuscript. A.M. and R.H. acquired the data. A.M. and H.S. interpreted the data. A.M., H.I.S., and S.U. drafted the manuscript, and R.H. substantially revised it. All authors read and approved the final manuscript.

Data Availability Statement

The data that support the findings of this study are not publicly available due to privacy reasons but are available from the corresponding author.

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