



Unusual rapid resolution of postsclerectomy exudative retinal detachment with topical NSAIDs therapy in a case of nanophthalmos

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Abstract

Nanophthalmos is a developmental ocular malformation that has been associated with high risks of uveal effusion syndrome and exudative retinal detachment (ERD). A variety of surgical procedures and systemic/topical steroids have been described as effective for treatment of ERD. However, the possibility of side effects should be considered. Here, we describe a patient with nanophthalmos who was treated for recurrent ERD during long-term follow-up, and we discuss non-surgical treatment options that are available in such cases. A 43-year-old woman with bilateral nanophthalmos exhibited ERD in her right eye for one month. After partial thickness sclerectomy with central sclerostomy, the retina was completely reattached. However, ERD recurred 3 years later. The patient refused surgery; therefore, we employed conservative treatment of topical nonsteroidal anti-inflammatory drugs (NSAIDs) and observation. One month later, the retina was completely reattached. To the best of our knowledge, no previous report has described resolution of recurrent ERD in a patient with nanophthalmos using NSAIDs during long-term follow-up after successful surgical treatment. Our success using this approach suggests that it could be used as alternative treatment for ERD in patients with nanophthalmos before application of further treatments.

Keywords

Nanophthalmos, exudative retinal detachment, uveal effusion syndrome, sclerectomy, nonsteroidal anti-inflammatory drugs, sclerostomy, conservative treatment, retina, sclera

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Introduction

Nanophthalmos is a developmental ocular malformation in which the eye is smaller than normal¹ and eye volume is reduced without other major ocular or systemic anomalies.² Eyes with nanophthalmos have a thick and impermeable sclera, which has been associated with a high risk of uveal effusion syndrome (UES) and exudative retinal detachment (ERD), regardless of intraocular surgery.^{3,4}

Ultrastructural and histochemical studies have demonstrated that with increasing age, scleral collagen fiber abnormalities occur and the production of glycosaminoglycans is altered in scleral cells in patients with nanophthalmos.⁵ These processes tend to obstruct the vortex veins and reduce the transscleral drainage of protein-rich fluids from the suprachoroidal space, thus leading to UES, as well as both choroidal and retinal detachment.⁵

A variety of surgical procedures have been described as effective for treatment of ERD in patients with nanophthalmos, including sclerotomy or sclerectomy with sclerostomy.^{6,7} However, ERD may recur due to proliferation of scleral tissue. Here, we describe a patient with nanophthalmos who was treated for idiopathic recurrent ERD during long-term follow-up after surgery, and we discuss non-surgical treatment options that are available in such cases.

Case report

This study was approved by the Ethics Committee of The Second Xiangya Hospital of Central South University, and the patient provided consent to publish this case report.

A 43-year-old woman with bilateral nanophthalmos was referred to our clinic because of decreasing visual acuity (VA) and shadow in her right eye that had persisted for one month. Family history

revealed that, among their relatives, only the patient and her brother had nanophthalmos.

Clinical examination of the patient showed that VA was limited to counting fingers (CF)/10 cm in the right eye. Both eyes were deeply set in the orbits, with narrow palpebral fissures and corneal diameters of 11.2–11.4 mm. Examination with IOL Master 500 (Carl Zeiss Co., Ltd., Shanghai, China) revealed an axial length of 15.34 mm in the right eye and 15.15 mm in the left eye. Fundus examination revealed the presence of smooth, bullous retinal detachments (Figure 1); tortuous retinal vessels and fine macular folds were also observed. A diagnosis of nanophthalmos with ERD was made, and the patient was treated with 80 mg prednisolone once-daily for 1 week; however, the ERD persisted. Therefore, we performed one-half thickness sclerectomy with central sclerostomy in each quadrant of the right eye (Figure 2), as described in a prior report.⁶ At the 1.5-month follow-up, the retina was completely reattached (Figure 3), and VA in the right eye had improved to 1/50. The patient's VA remained stable, and there was no evidence of UES during a three-year follow-up period.

Subsequently, the patient experienced reduced VA for 10 days, and returned to our clinic for treatment. ERD was observed, and VA in the right eye was again limited to CF (Figure 4). Because of the risks of surgery, the patient refused additional procedures; considering that steroid therapy was ineffective, we selected minimal treatment of topical nonsteroidal anti-inflammatory drugs (NSAIDs) and observation. Surprisingly, fundus photography examination showed that the retina had partially reattached within 7 days. Thus, we chose to observe the patient's progress without additional treatment. At the 1-month follow-up, the retina had completely reattached and VA had recovered to 1/30;

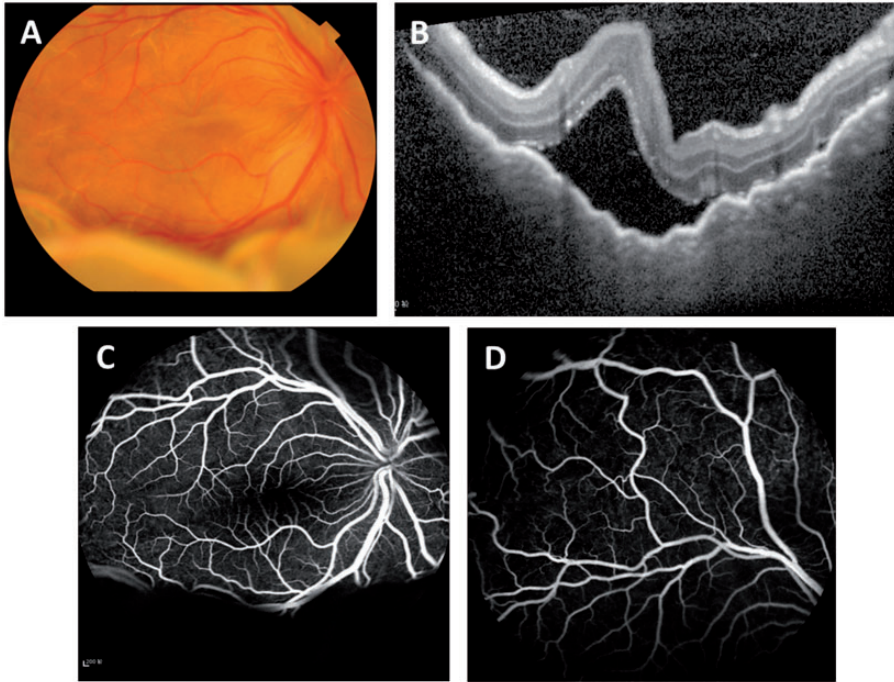


Figure 1. Findings of initial examinations in the patient's right eye. (a) Fundus photograph of right eye showed the retina had become detached and that smooth, elevated bullous without retinal breaks were present, fine macular folds were also found. (b) Optical coherence tomography revealed subretinal fluid at macula. (c,d) Fluorescein angiography revealed loss of fluorescence in the inferior pole and tortuous retinal vessels, as well as the appearance of focal hyperfluorescence in the periphery.

notably, VA remained stable throughout a 3-month follow-up period (Figure 4).

Discussion

Nanophthalmos⁸ is classically viewed as a rare condition characterized by a small eye that exhibits arrested development without other ocular malformations. The etiology of nanophthalmos involves developmental arrest of the globe after closure of the embryonic fissure. This event results in a variety of clinical features, including abnormally thickened sclera, short axial length (<20 mm in adults),⁹ small corneal diameter, and crowded anterior chamber.

The present report describes ERD, which is a complication frequently observed

in patients with nanophthalmos; ERD is thought to result from an abnormality in the sclera that impedes transscleral intraocular fluid outflow and compresses the vortex vein, thereby causing fluid accumulation in the choroid and leading to ciliochoroidal detachment.⁵ Previous studies⁴ have suggested that in patients who exhibit nanophthalmos with ERD, inelastic scleral fibers might replace normal elastic fibers; however, the fundamental mechanism underlying ERD in patients with nanophthalmos remains unclear and warrants further research.

Previous studies have demonstrated that some treatments can effectively treat ERD in patients with nanophthalmos; these treatments include corticosteroid therapy,

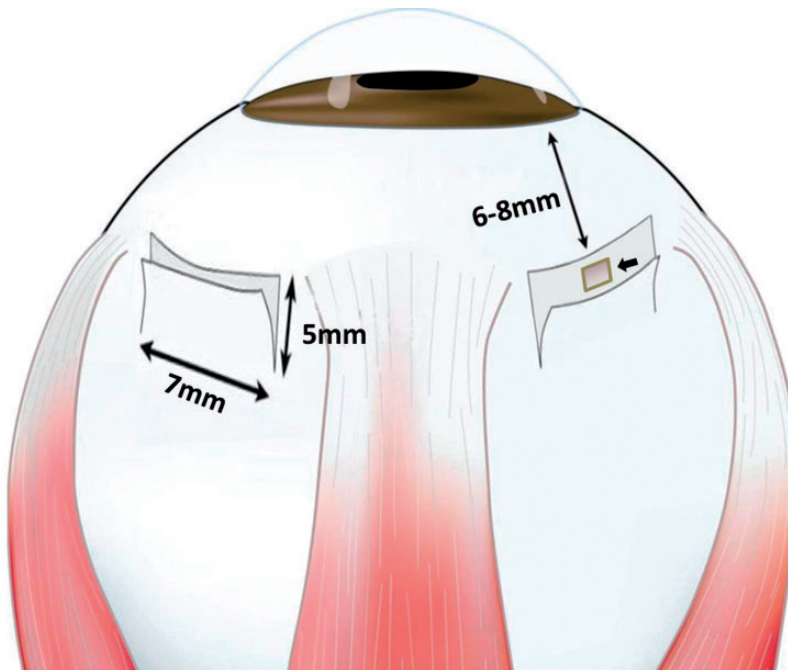


Figure 2. Schematic of sclerotomy procedure. A square bracket-shaped (5 × 7-mm) one-half thickness scleral incision was made in the quadrant between the rectus muscles, with a central sclerostomy of 1 × 2-mm area (black arrow) in each square. Scleral flaps were removed after sclerostomy.

NSAID therapy, and surgical procedures.^{6,7,10,11} The mechanism by which ERD is resolved by corticosteroids is a matter of speculation. Ufuk et al.¹⁰ suggested that treatment with high doses of corticosteroid reduces inflammation, and that this may be related to the ability of corticosteroids to stabilize the blood–ocular barrier, thereby improving regression. Those authors described a patient with nanophthalmos¹⁰ in whom ERD was unusually rapidly resolved by treatment with topical corticosteroid alone. However, in our patient, during the initial instance of ERD, the patient did not respond to steroids, and we therefore employed a surgical approach. Scleral resection with sclerotomy, as well as sclerectomy with sclerostomy^{6,7} are reportedly effective treatments because some patients do not respond to medication or experience side effects, such as increased

intraocular pressure during long-term usage. However, the hypoplastic/fragile nature of the vortex veins should be carefully considered because surgical procedures could lead to intraoperative amputation or rupture of these veins; moreover, because scleral tissues proliferate, ERD could recur during long-term follow-up.¹²

The surgical procedure used in our patient successfully provided symptomatic relief, but ERD recurred 3 years later. The importance of scleral permeability to water and colloids in maintaining hydrostatic and osmotic gradients for the choroid has long been recognized.¹³ As a result of disturbance of the normal movement of fluid from the subretinal space to the suprachoroidal space via the vortex veins, as well as disruption of the mechanism by which the retinal pigment epithelium pumps water but excludes colloids, the subretinal fluid

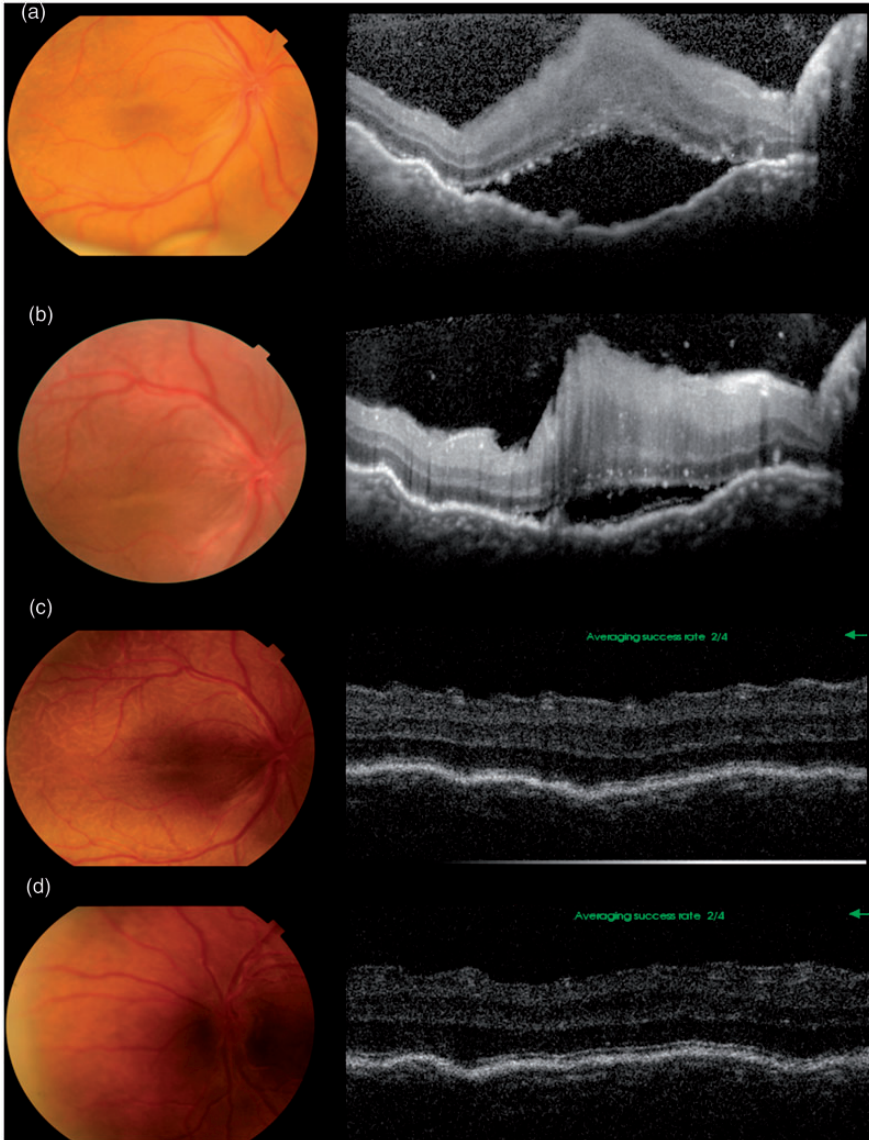


Figure 3. Findings of follow-up fundus photography and optical coherence tomography examinations in the patient's right eye after surgery for initial treatment of exudative retinal detachment. The retina had completely reattached within 1.5 months, and remained stable throughout the follow-up period. (a) One day postoperatively, subretinal fluid remains at the macula. (b) Two weeks postoperatively, the retinal fluid was partially absorbed. (c) 1.5 months postoperatively, the subretinal fluid was completely absorbed, and the retina was reattached. (d) 1.5 years postoperatively, the retina remained stable.

becomes enriched in protein.¹³ Recurrence of ERD after scleral resection has been reported, and histological examination showed that both scleral and fibrous scar

tissues obtained from previous sclerectomy sites were able to regenerate;⁵ in this situation, a second surgery is indicated. Although several studies^{1,3,9} have

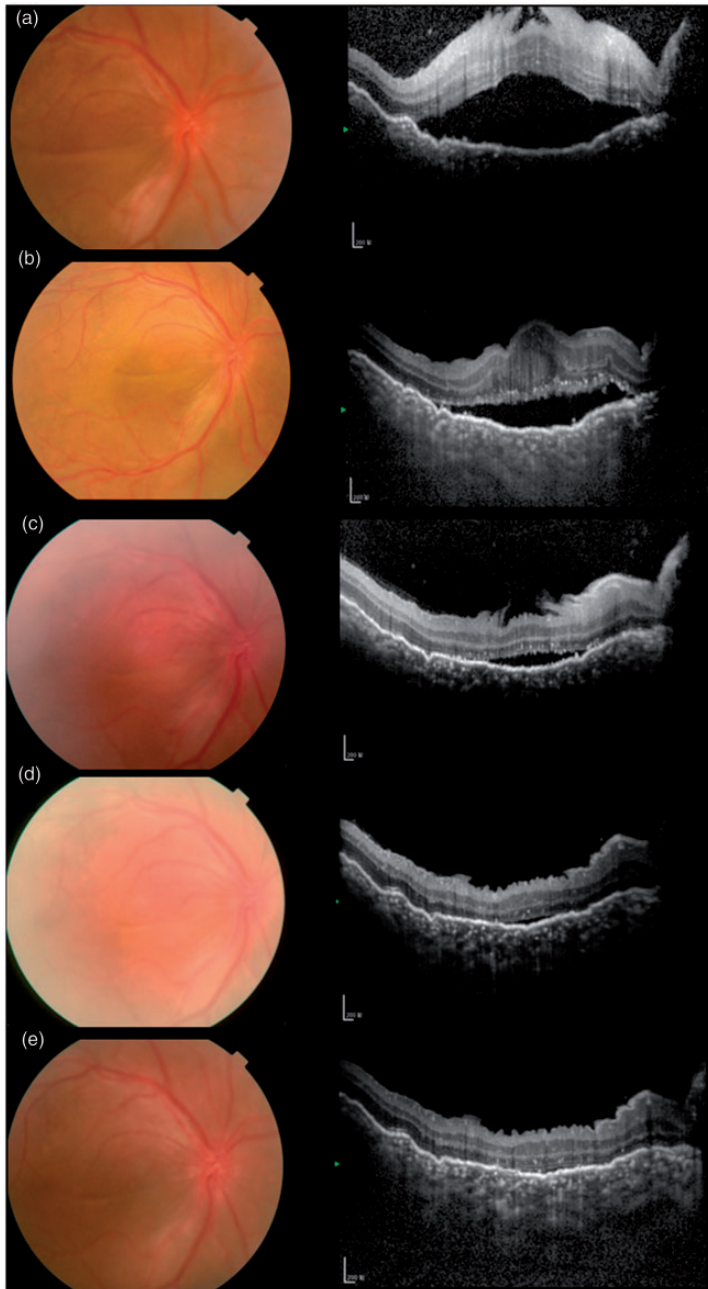


Figure 4. Findings of fundus photography and optical coherence tomography examinations in the patient's right eye after treatment of recurrent exudative retinal detachment with non-steroidal anti-inflammatory drugs. The retina had completely reattached within one month, and remained stable at the 3-month follow-up. (a) Day of admission. Left: Fundus photography showed superior, inferior, and nasal retinal detachment without retinal breaks; fine macular folds were also present. Right: Optical coherence tomography revealed subretinal fluid in the macula. (b) At the 7-day follow-up, the subretinal fluid was partially absorbed. (c) At the 14-day follow-up, the retinal fluid was partially absorbed. (d) At the 1-month follow-up, the subretinal fluid was completely absorbed, and the retina was reattached. (e) At the 3-month follow-up, the retina remained stable.

demonstrated that certain gene mutations can cause nanophthalmos, no gene mutation was identified in our patient.

When a patient with nanophthalmos presents with indications for surgery, extensive risk analysis must be performed. We chose observation with short-term topical NSAID therapy, instead of other classical approaches, as treatment for the second instance of detachment; subsequently, the retina completely reattached and VA improved. The mechanism by which the ERD resolved is unclear, but several cases have demonstrated that the anti-inflammatory effect of NSAIDs could protect against choroidal inflammation and effusion, causing the protein-rich subretinal fluid to be reabsorbed.¹¹ Furthermore, prostaglandin administration could reduce scleral collagen levels by increasing scleral metalloproteinase levels, thus stimulating the pump mechanism of the retinal pigment epithelium and facilitating reabsorption of the fluid.^{11,14} Psychological factors might also have contributed to recovery because, during treatment for the second instance of ERD, the patient was calmer and more relaxed than during treatment for the first instance of ERD. It has been shown that long-term anxiety can cause imbalances in certain chemicals, such as 5-hydroxytryptamine, norepinephrine, and dopamine, leading to disturbances in body functions, such as local edema and stress-induced inflammatory exudation.¹⁵ Arias Palomero et al.¹⁶ described a patient who developed acute UES after treatment with escitalopram, which is a highly selective 5-hydroxytryptamine reuptake inhibitor; the drug was prescribed to treat depression, but its use led to subretinal fluid accumulation. Hence, 5-hydroxytryptamine might play an important role in the course of UES and efforts should be taken to ensure it remains balanced; moreover, maintenance of a positive outlook might relieve anxiety-induced functional disturbances in

some patients. However, the mechanisms underlying these effects should be further investigated.

Conclusions

This report showed that conservative treatment comprising observation and short-term topical NSAID administration might be appropriate for patients with nanophthalmos who exhibit ERD. To the best of our knowledge, no previous report has described resolution of recurrent ERD in a patient with nanophthalmos during long-term follow-up after successful surgical treatment. Thus, observation with topical NSAIDs over several weeks could serve as an alternative treatment for ERD in patients with nanophthalmos before application of surgical treatment or systemic/topical corticosteroid therapy.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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