CASE REPORT

GLOBE RETRACTION IN A PATIENT WITH NANOPHTHALMOS

Susanne Oner, Ahmet Fazıl Nohutcu, Hatice Nur Ortak
Ophthalmology Clinic I, Haydarpasa Numune Education and Research Hospital, Istanbul, Türkiye

ABSTRACT

We present a case of bilateral nanophthalmos with ipsilateral globe retraction and narrowing of the palpebral fissure on adduction of the left eye. A complete ophthalmological examination, including strabismic evaluation, gonioscopy, ultrasound and magnetic resonance imaging (MRI) was performed. A 14 year-old girl had bilateral nanophthalmos with pseudopapilloedemas and macular folds, anterior chamber angles of Shaffer grade II, and arcuate scotomas. She had orthotropia. There was ipsilateral globe retraction with narrowing of the palpebral fissure on adduction of the left eye, and no ocular motility restriction. Both abducens nerves were detectable on MRI. Her brother had bilateral nanophthalmos without ocular motility disorder. We suggest that the phenomenon of globe retraction in this case may be related to mechanical factors rather than innervational abnormalities.

Keywords: Nanophthalmos, Globe retraction, Duane's retraction syndrome

NANOFTALMUSLU BIR OLGUDA GLOB RETRAKSYONU

ÖZET

Sol gözün addüksiyonunda ipsilateral glob retraksiyonu ve palpebral fissür daralması olan bilateral nanoftalmuslu bir olguyu sunmak istiyoruz. Hastaya şaşılık muayenesi, gonyoskopi, ultrasonografi ve magnetik rezonans görüntülemesini (MRG) de içeren kapsamlı göz muayenesi yapıldı. On dört yaşındaki kız hastada psödopapilödem, maküla kıvrımları, Shaffer II derecesinde ön kamara açısı ve arkuat skotom ile birlikte nanoftalmus vardı. Olgu ortotropik ve göz hareketleri serbest idi. Sol gözün addüksiyonunda ipsilateral glob retraksiyonu ile birlikte palpebral fissür daralması görülmekteydi. MRG'de her iki abdusens siniri belirgindi. Kardeşinde de, göz hareketlerinde bozukluk olmadan, bilateral nanoftalmus vardı. Bu olgudaki glob retraksiyonunun innervasyonel sebeplerden çok mekanik faktörlere bağlı olduğunu düşünmekteyiz.

Anahtar Kelimeler: Nanoftalmus, Glob retraksiyonu, Duane'in retraksyon sendromu

INTRODUCTION

The characteristic features of nanophthalmos are narrow palpebral fissures in combination with a deep set globe in a small orbit, hyperopia, short axial length, reduced volume of the globe with normal lens volume, and normal or reduced corneal diameter[1]. Associated strabismus, in most cases nonaccomodative esotropia, and with lower incidence microesotropia and exotropia, can be observed[2]. Globe retraction and narrowing of the palpebral fissure on adduction are typical components of the Duane's retraction syndrome (DRS). Numerous systemic and ocular disorders have been reported in association with DRS[3]. We report a case of bilateral nanophthalmos with ipsilateral globe retraction and narrowing of the palpebral fissure on adduction of the left eye.

Corresponding author:
Susanne Öner, M.D.
I. Göz Kliniği, Haydarpaşa Numune Eğitim ve
Araştırma Hastanesi, *Haydarpaşa, İstanbul, Türkiye*
e-mail: susaoner@yahoo.com

Marmara Medical Journal 2005;18(3);129-130

CASE REPORT

A 14 year-old girl with normal prenatal and natal anamnesis, and no history of disease, had bilateral narrow palpebral fissures and deeply set globes (Fig. 1). Best corrected visual acuity was 0.4 (+ 13.0) OD, and 0.5 (+ 11.0) OS. Horizontal corneal diameters were 11.0 mm bilaterally, and the irises were thick with a convexity towards the cornea. Gonioscopy revealed bilateral anterior chamber angles of Shaffer grade II, peripheral anterior synechiae and prominent iris convexity. Intraocular pressures were 11 mm Hg in both eyes. Visual field testing showed bilateral arcuate scotomas, more extensive in the right eye. There were bilateral pseudopapilloedemas and macular folds. Orbital B-scan ultrasonography revealed small globe diameters, slightly small lens diameters, shallow anterior chambers, and thick scleras. MRI showed bilateral normal abducens nerves and symmetrical orbital anatomy. There was orthotropia in primary position and no restriction of ductions and versions. Diplopia, head turn or nystagmus were not observed. On adduction of the left eye, there was ipsilateral globe retraction and narrowing of the palpebral fissure (Fig. 1).

Nanophthalmos was present in 2 of 3 children of this family. One brother, an otherwise healthy 11 year-old boy without a history of abnormalities during pregnancy or birth, had bilateral nanophthalmos without any ocular motility disorder. Mother and father were secondary degree relatives.

DISCUSSION

Our case showed the typical features of nanophthalmos, and globe retraction with narrowing of the palpebral fissure of the left eye on right gaze. Globe retraction and narrowing of the palpebral fissure on adduction are typical components of DRS, and have been related to the paradoxical innervation of the lateral rectus muscle, leading to co-contraction of the horizontal rectus muscles[3]. In DSR magnetic resonance imaging can show absence of the abducens nerves[4]. Our case had no other features of DRS, and both abducens nerves were present. Gross et al.[5] report a case of DRS type II, with synchronous innervation of the ipsilateral horizontal rectus muscles without globe retraction. They propose that in DRS both innervational and mechanical factors are involved. In our case, we propose a mechanical factor for the globe retraction, though it remains interesting why despite symmetrical anatomy, globe retraction was observed in only one eye of the patient.

Fig. 1: The case had bilateral deep set globes with narrow palpebral fissures, and retraction of the left globe with narrowing of the palpebral fissure on right gaze.

REFERENCES

1. Singh OS, Sofinski SJ. Nanophthalmos. Guidelines for diagnosis and therapy. In: DM Albert, FA Jacobiec, eds. Principles and Practice of Ophthalmology. Philadephia: WB Saunders, 1994: 1528-1540.
2. Sener EC, Mocan MC, Sarac OI, Gedik S, Sanac AS. Management of strabismus in nanophthalmic patients. Ophthalmology 2003; 110: 1230-1236.
3. DeRespinis PA, Caputo AR, Wagner RS, Guo S. Duane's Retraction syndrome. Surv Ophthalmol 1993; 38: 257-288.
4. Ozkurt H, Basak M, Oral Y, Ozkurt Y. Magnetic resonance imaging in Duane's retraction syndrome. J Pediatr Ophthalmol Strabismus 2003; 40: 19-22.
5. Gross SA, Tien DR, Breinin GM. Aberrant innervational pattern in Duane's syndrome type II without globe retraction. Am J Ophthalmol 1994; 117: 348-351.