

A Patient with Cutis Verticis Gyrata Having Thick Cornea and High Cup to Disk Ratio

Kalın Kornea ve Yüksek Cup Disk Oranına Sahip Kutis Vertisis Giratalı Bir Olgu

Remzi KARADAĞ¹, Ayşe Serap KARADAĞ², Gülçin Güler ŞİMŞEK³

Case Report

Olgu Sunumu

ABSTRACT

A 33-year-old male patient was referred to the dermatology clinic for a complaint of furrowing of the scalp that had begun 7 years previously and which had slowly progressed. A diagnosis of cutis verticis gyrata (CVG) was made, and the patient was referred to our clinic for assessment of possible associated ocular conditions. High central corneal thickness (CCT) and high cup to disk ratio were detected in a patient with primary essential CVG who had normal intraocular pressure (IOP). In patients with CVG, high CCT may lead to artifactually high measurements of IOP, resulting in unnecessary treatments.

Key Words: Cutis verticis gyrata; central corneal thickness; cup to disk ratio.

ÖZ

Otuz üç yaşındaki erkek hasta 7 yıldan beri var olan ve giderek artan kafa derisinin kıvrımlı görünüm alması şikayetiyle dermatoloji polikliniğine başvurdu. Hastaya kutis verticis girata (KVG) tanısı konularak eşlik edebilecek göz bulgularının değerlendirilmesi amacıyla kliniğimize gönderildi. Primer esansiyel KVG'li hastada yüksek santral korneal kalınlık (SKK), yüksek cup disk oranı ve normal sınırlarda göz içi basıncı (GİB) tespit edildi. KVG'li hastalarda yüksek olabilen SKK yanlılıkla GİB'nin yüksek ölçülmesine neden olabilir ve kişiler gereksiz tedavi edilebilir.

Anahtar Kelimeler: Kutis verticis girata; santral korneal kalınlık; cup disk oranı.

Glo-Kat 2010;5:60-62

INTRODUCTION

Cutis verticis gyrata (CVG) is a condition in which hypertrophy and folding of the skin of the scalp result in a cerebriform or gyrate appearance.¹ CVG is uncommon, with an estimated incidence of 1 in 100.000 among the male population and 0.026 in 100.000 among the female population.^{2,3}

CVG is classified into primary (idiopathic) and secondary forms. Primary CVG is further divided into essential and nonessential types. Essential primary CVG is not associated with other conditions; however, the nonessential form can be associated with mental retardation, epilepsy, and various ophthalmic abnormalities.² Secondary CVG may result from local inflammation or neoplasia, or

from a systemic disease that affects areas of hairy skin. Several diseases have been reported in association with secondary CVG including inflammatory dermatoses, acanthosis nigricans, nevi, dermatofibromas, hamartomas, trauma, and systemic diseases (e.g. syphilis, tuberous sclerosis, acromegaly, leukemia, neurofibromatosis, amyloidosis and myxedema). In addition, several syndromes have been reported in association with CVG.⁴

Although the condition is well addressed in the dermatology literature, it is rarely described in the ophthalmology literature. Here we report a patient with CVG in whom central corneal thickness (CCT) and cup to disk ratio are high in the presence of normal intraocular pressure (IOP).

Geliş Tarihi : 31/08/2009

Kabul Tarihi : 13/10/2009

Received : August 31, 2009

Accepted : November 13, 2009

- 1- Fatih Üniversitesi Tıp Fakültesi Hastanesi, Göz Hastalıkları Kliniği, Ankara, Yrd. Doç. Dr.
- 2- Ankara Keçiören Eğitim ve Araştırma Hastanesi, Dermatoloji Kliniği, Ankara, Uz. Dr.
- 3- Ankara Keçiören Eğitim ve Araştırma Hastanesi, Patoloji Kliniği, Ankara, Uz. Dr.

- 1- M.D. Asistant Professor, Fatih University School of Medicine, Department of Ophthalmology Ankara/TURKEY
KARADAĞ R., drkaradag@yahoo.com
- 2- Ankara Keçiören Training and Research Hospital Dermatology Clinic, Ankara / TURKEY
KARADAĞ A.S.
- 3- Ankara Keçiören Training and Research Hospital Pathology Clinic, Ankara / TURKEY
ŞİMŞEK G.G.,

Correspondence: M.D. Asistant Professor, Remzi KARADAĞ
Fatih University Medical School, Department of Ophthalmology, Alparslan Türkeş
Caddesi No: 57 Emek Ankara/TURKEY

CASE REPORT

A 33-year-old male patient was referred to the dermatology clinic for a complaint of furrowing of the scalp that had begun 7 years previously and which had slowly progressed. In the inspectional examination there were many folds located diffusely on the vertex, occipital and parietal areas of the scalp that could not be obliterated by pressure (Figure 1). A diagnosis of CVG was made, and the patient was referred to our clinic for assessment of possible associated ocular conditions.

The patient had no neurological symptoms and was of normal intelligence. All laboratory tests, including serum levels of growth hormone, thyroid hormone, immunoglobulins and blood sugar, gave normal results. Standard X-rays revealed no periostosis in the long bones. Magnetic resonance imaging revealed no intracranial pathology.



Figure 1: Skin findings of the patient.

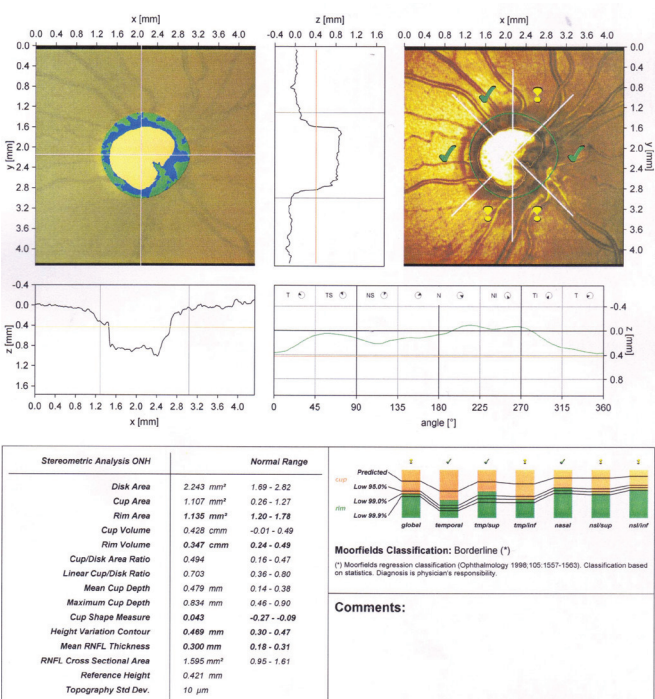


Figure 2: HRT II print out of right eye.

In the patient's ophthalmological examination, visual acuity was 20/20 in both eyes. IOP was 19 mmHg in the right eye and 18 mmHg in the left eye. CCT was 598 μm in the right eye and 594 μm in the left. Slit lamp examination of the anterior segment revealed normal findings in both eyes. In the fundus examination, a cup to disk ratio of 0.7 was measured for the right eye (Figure 2) and 0.8 was measured for the left eye (Figure 3). The patient's visual field exam was within normal limits. In six months of follow up, no changes were encountered in the patient's ocular findings or visual field test. Because the patient had no obvious conditions associated with CVG, we classified the disorder as the primary essential type.

DISCUSSION

Cutis verticis gyrata is classified into primary and secondary forms, with the secondary type arising from an identifiable cause. The primary type is further divided into essential and nonessential forms, according to whether it is seen as an isolated condition (essential) or in association with other conditions (nonessential), particularly neurologic or ophthalmologic conditions.² The essential form is extremely rare. In most cases, the onset of this type of CVG occurs around or after puberty, and in 90% of patients onset occurs after the age of 30. Secondary CVG often presents later in life and may be due to a variety of underlying disorders.⁵

Some ocular disorders may be associated with the primary nonessential form of CVG. In the literature, less than 200 cases of CVG have been reported, and ocular findings have been described in only a few of these. Megarbane et al.⁶ reported two patients who had CVG associated with progressive retinitis pigmentosa and cat-

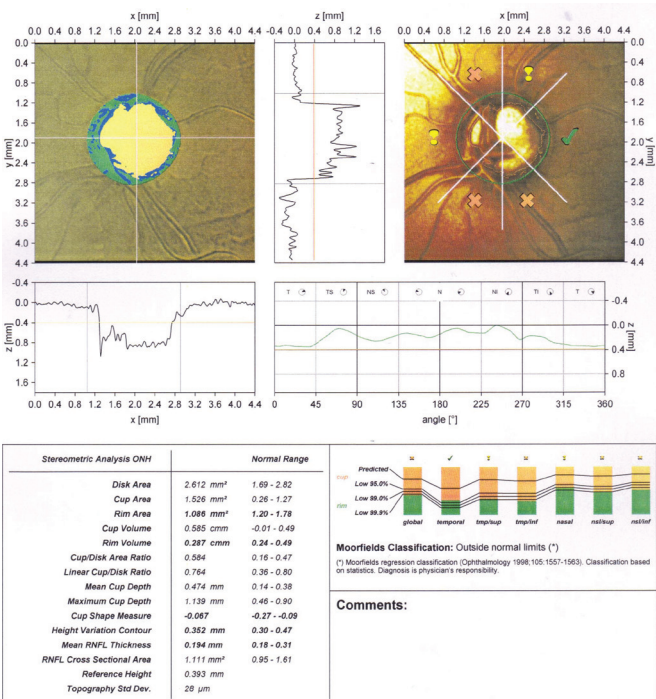


Figure 3: HRT II print out of left eye.

aract. Bilen et al.⁷ reported a patient with CVG who had Graves' disease and exophthalmos. In addition, CVG associated with corneal leukoma in patients with the acromegaloid phenotype has been reported.^{8,9}

In our patient, ocular findings included high CCT in both eyes and high cup to disk ratio in both eyes. However, IOP was normal in both eyes. Visual field test results were also normal and did not change during follow up, so the high cup to disk ratio was interpreted as non-pathologic.

In conclusion, in patients with CVG, high CCT may lead to artifactually high measurements of IOP, and in the presence of a high cup to disk ratio, this can lead to a mistaken diagnosis of glaucoma and cause to begin of an unnecessary treatment. Furthermore importance of the CCT was emphasized in this text by virtue of a rare case.

REFERENCES/KAYNAKLAR

1. Misirlioglu A, Karaca M, Akoz T.: Primary cutis verticis gyrata and scalp reduction in one stage with multiple pinwheel flaps (revisited). *Dermatol Surg.* 2008;34:935-938.
2. Chang GY.: Cutis verticis gyrata, underrecognized neurocutaneous syndrome. *Neurology.* 1996;47:573-575.
3. Snyder MC, Johnson PJ, Hollins RR.: Congenital primary cutis verticis gyrata. *Plast Reconstr Surg.* 2002;110:818-821.
4. Larsen F, Birchall N.: Cutis verticis gyrata: three cases with different aetiologies that demonstrate the classification system. *Australas J Dermatol.* 2007;48:91-94.
5. Al-Bedaia M, Al-Khenaizan AS.: Acromegaly presenting as cutis verticis gyrata. *Int J Dermatol.* 2008;47:164.
6. Mégarbané A, Waked N, Chouery E, et al.: Microcephaly, cutis verticis gyrata of the scalp, retinitis pigmentosa, cataracts, sensorineural deafness, and mental retardation in two brothers. *Am J Med Genet.* 2001;98:244-249.
7. Bilen H, Atasoy M, Akcay G, et al.: Elephantiasic pretibial myxedema and cutis verticis gyrata caused by Graves' disease. *Thyroid.* 2006;16:815-816.
8. Rosenthal JW, Kloepper HW.: An acromegaloid, cutis verticis gyrata, corneal leukoma syndrome. A new medical entity. *Arch Ophthalmol.* 1962;68:722-726.
9. Nakamura M.: Acromegaloid phenotype-cutis verticis gyrata-corneal leukoma. *Ryokibetsu Shokogun Shirizu.* 2001;33:135.