



# Diğer Yönlerden Normal Olan Bir Gözde Çift Konjenital Ön Skleral Stafilom

## *Dual Congenital Anterior Scleral Staphylomas in an Otherwise Normal Eye*

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### **Summary**

In most cases, congenital anterior staphyloma presents with a disfigured eye and very poor vision. Only one pediatric case in the literature was reported as a mild form with a solitary lesion in an otherwise normal eye. We herein report an adult case with two neighboring congenital anterior scleral staphylomas confirmed with ultrasound biomicroscopic imaging, with no associated ocular abnormalities. (*Turk J Ophthalmol* 2013; 43: 211-2)

**Key Words:** Anterior staphyloma, congenital, ultrasound biomicroscopy

### **Özet**

Çoğu olguda konjenital ön stafilom şekli bozulmuş bir göz veya çok düşük görme ile kendini gösterir. Literatürde sadece bir pediatrik olguda, diğer taraftan normal olan bir gözde tek lezyon ile hafif şekli bildirilmiştir. Biz bu yazıda eşlik eden oküler anormallığı bulunmayan, ultrasonik biyomikroskopi görüntülemesi ile doğrulanmış iki komşu konjenital ön stafilomlu erişkin bir olguyu bildiriyoruz. (*Turk J Ophthalmol* 2013; 43: 211-2)

**Anahtar Kelimeler:** Konjenital, ön stafilom, ultrason biyomikroskopi

### **Introduction**

Staphyloma is a defect in the sclera or cornea through which uveal tissue and other intraocular structures bulge. It can be either congenital or acquired. Congenital staphyloma can occur anteriorly in the corneal region or posteriorly in the optic nerve region.<sup>1</sup> Acquired form can occur anteriorly in many conditions such as rheumatoid scleritis, after trans-scleral contact diode laser cyclophotocoagulation, neurofibromatosis, after lensectomy in homocystinuria, and posteriorly in conditions such as high myopia.<sup>1-5</sup>

In this case report, we demonstrate two tiny congenital anterior scleral staphylomas adjacent to each other in an otherwise normal eye of a young adult.

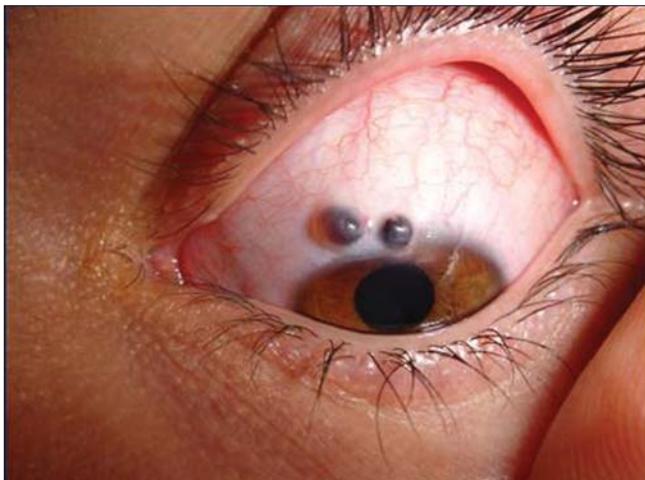
### **Case Report**

A 22-year-old young man presented with a complaint of two black spots on his left eye. According to him, these lesions have been there since his childhood. He was the product of a normal

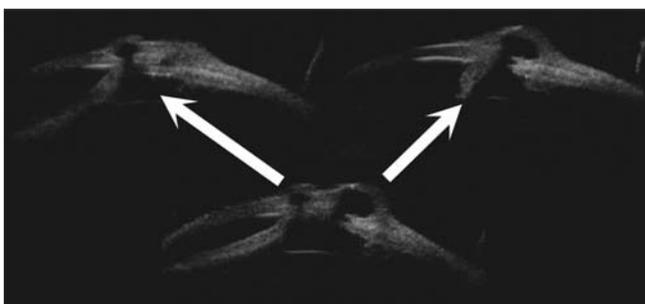
pregnancy and delivery, and had grown-up normally. On slit lamp examination, two tiny neighboring black lesions were located within the limbal area of the left eye (Figure 1). One of the lesions measuring 2x2 mm was located 2 mm superior to the corneoscleral limbus, and the other one measuring 1.5x1.5 mm was located on the limbus. The lesions were elevated 1 mm from the scleral surface and covered by thin conjunctiva. Transillumination showed slight transmission of light through the lesions. Fundus examination was normal. His visual acuity was 20/20 in both eyes, and intraocular pressure was normal. No systemic and concurrent ocular abnormality was noted. These clinical findings were suggestive of localized anterior scleral staphyloma and were confirmed by ultrasound biomicroscopy (Figure 2). Six months later, follow-up examination demonstrated no change in the lesions.

### **Discussion**

Mostly, we encounter congenital anterior staphylomas with other ocular abnormalities causing severe functional disturbance



**Figure 1.** Clinical appearance of anterior scleral staphylomas



**Figure 2.** Ultrasound biomicroscopic appearance of staphylomas; scleral defect in the small (long arrow) and the big (short arrow) staphylomas

in a disfigured eye, and reconstruction of a satisfactory vision stands as a big challenge for caring ophthalmologists.<sup>6</sup> To our knowledge, isolated congenital form of anterior staphyloma is quite rare. The pathogenesis still remains unclear, but it is certain that there is a scleral defect, allowing uveal tissue to protrude through the opening.<sup>3</sup> The data obtained from histopathologic and electron microscopic studies suggest that this weakness on the wall occurs during embryogenesis.<sup>7</sup> Furthermore, it is postulated that lack of mesodermal differentiation or intrauterine keratitis (either blood-borne or trans-amniotic) may be the causative factors.<sup>8,9</sup> From this point of view, potency of causative agent and the time of insult may be the factors that determine the extension of ocular involvement. Currently, there is no standard classification system for congenital anterior staphyloma regarding severity of the disease. In our case, except for these two black spots, ophthalmologic examination was normal. However,

we had no explanation for the possible insulting factor and mild form of the pathology. Literature review revealed only one similar case, but staphyloma was single, and located 6 mm away from the limbus.<sup>3</sup> Though the diagnosis is clear in most cases, congenital anterior scleral staphyloma must be differentiated from lesions such as conjunctival nevus or melanoma, foreign body, and extraocular extension of ciliary body melanoma.<sup>1</sup> It can be differentiated from such lesions simply by transillumination. But ultrasound biomicroscopy presents detailed data to help diagnosing in suspected cases. In our case, ultrasound biomicroscopic findings confirmed the anterior scleral staphyloma. Though follow-up was not adequate enough to reach a conclusion regarding progression, we did not observe any change in the morphology and recommended avoiding any ocular trauma and wearing protective eyeglasses.

In conclusion, anterior scleral staphyloma can be congenital or acquired and also can be more than one. Ultrasound biomicroscopy and transmission of light with transillumination can help to differentiate anterior scleral staphyloma from pigmented conjunctival tumors and extraocular extension of ciliary body melanoma. Protective eyeglasses should be recommended for the patients with scleral staphyloma.

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