

Surgical Management of Anterior Lenticonus in a Patient with Alport's Syndrome

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Background: To discuss ophthalmic features and management of a patient with Alport's syndrome and anterior lenticonus.

Materials and Methods: A 22-year-old female patient with Alport's syndrome who, in another center, had undergone laser in situ keratomileusis (LASIK) because of high myopia was determined to have bilateral anterior lenticonus. We treated the other eye by phacoemulsification and intraocular lens (IOL) implantation.

Results: Lens extraction and IOL implantation gave satisfactory results. The eye undergone LASIK was rehabilitated by a contact lens.

Conclusion: As anterior and/or posterior lenticonus is easily overlooked, careful evaluation of patients with high refractive errors is essential before any decision for refractive surgery. Treatment of lenticonus should be by lens extraction and IOL implantation.

Key Words: Alport's syndrome, high myopia, lenticonus

Alport Sendromlu Bir Hastada Ön Lentikonusun Cerrahi Tedavisi

Amaç: Alport sendromlu bir hastada oftalmik bulguları ve ön lentikonusun tedavisini tartışmak.

Materyal ve Metod: Alport sendromlu 22 yaşındaki bayan hastada oftalmik bulgu olarak bilateral ön lentikonus mevcuttu. Sağ gözüne başka bir klinikte yüksek miyopi nedeniyle laser in situ keratomileusis (LASIK) uygulanmış olan hastanın diğer gözüne kliniğimizde fakoemülsifikasyon ve katlanabilir intraoküler lens implantasyonu yapıldı.

Bulgular: Fakoemülsifikasyon ile lens ekstraksiyonu ve intraoküler lens implantasyonu ile iyi bir görsel sonuca ulaşılmıştır. Daha önce LASIK uygulanmış diğer göz yumuşak kontakt lens ile rehabilite edilmiştir.

Sonuç: Ön ve arka lentikonus kolaylıkla gözden kaçabileceğinden refraktif cerrahi uygulanacak olgularda ameliyat öncesinde hastanın çok dikkatli muayene edilmesi şarttır. Lentikonusun tedavisi lens ekstraksiyonu ve intraoküler lens implantasyonu olmalıdır.

Anahtar Sözcükler: Alport sendromu, yüksek miyopi, lentikonus

Introduction

Alport's syndrome is a hereditary progressive disease that affects kidneys and auditory and visual systems. In 1927, Alport was the first to recognize the link between progressive nephropathy and sensorineural deafness (1). Renal findings may be unremarkable, such as asymptomatic microscopic hematuria seen in females, but may indicate a serious and life threatening condition, like terminal renal failure, usually seen in male patients. Hearing problem is more frequent in men than women. Ophthalmological findings are also frequently seen and occur in increased frequency in adulthood compared to childhood (2). Most typical and frequent ophthalmologic pathology is anterior lenticonus and more than 90% of the cases are associated with Alport's syndrome (3,4). It produces high refractive errors and optic image degradation leading to visual disability uncorrectable by glasses or contact lenses. In this case report, surgical treatment of a patient with bilateral anterior lenticonus is discussed.

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Case Report

A 22-year-old female patient came to our clinic with the complaint of gradual painless decrease in visual acuity over a 4-year period. She had been diagnosed with Alport's syndrome in a nephrology clinic and she had undergone renal transplantation 7 years before. She was using a hearing aid because of bilateral hearing loss. Ocular evaluation revealed a best corrected visual acuity of 20/100 in the right eye with $-12.0(-2.5 \times 80)$ correction and 20/125 in the left eye with $-26.0(-2.0 \times 160)$ correction. Slit lamp biomicroscopic examination showed presence of a LASIK flap (Figure 1) in the right eye and no corneal pathology in the left eye. Bilateral anterior lenticonus was present (Figures 2 and 3). Direct ophthalmoscopy aided in the visualization of an oil droplet in each lens (Figure 4). Both lenses were clear, with no evidence of cataract. Dilated fundus examination revealed no macular or retinal abnormality.

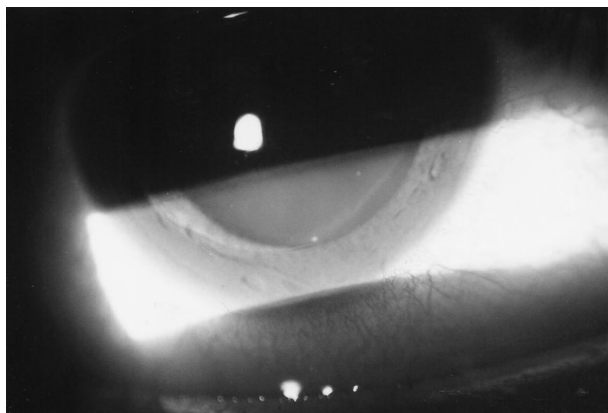


Figure 1. LASIK flap in the right eye.

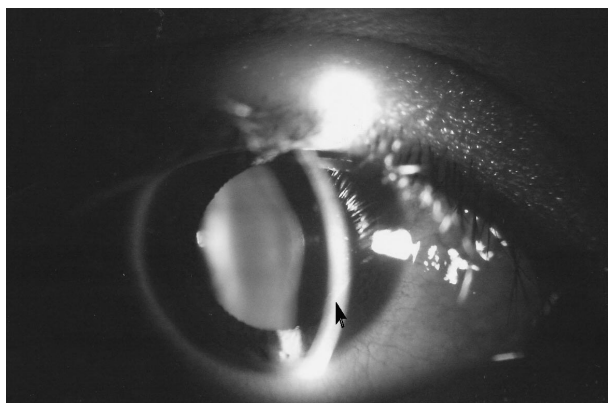


Figure 2. Anterior lenticonus in the right eye (arrow indicates margin of the LASIK flap).

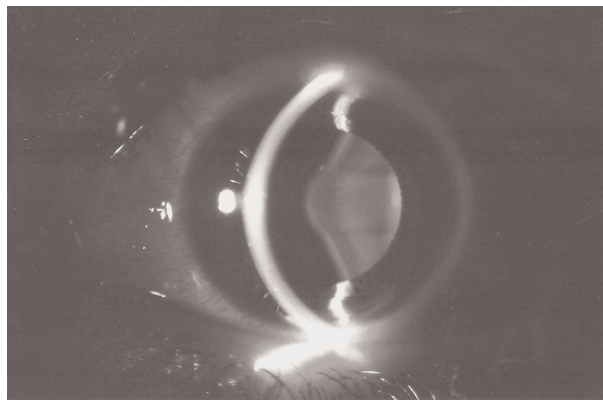


Figure 3. Anterior lenticonus in the left eye.

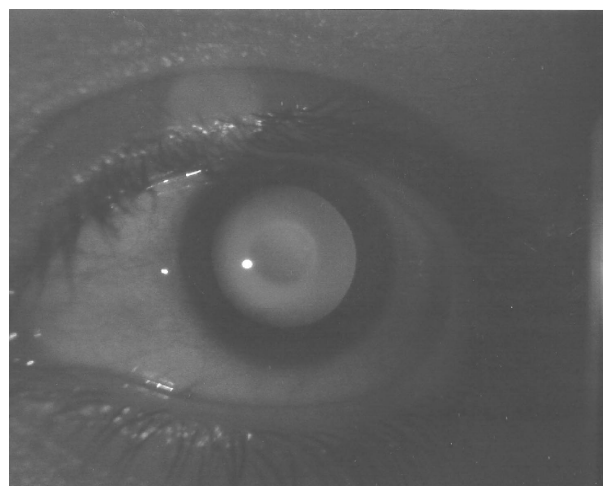


Figure 4. Oil droplet appearance of the left eye.

Keratometric values of the right eye were 40.25/39.12 dioptre (D) and the axial length was 22.10 mm. Keratometric values of the left eye were 44.50/46.75 (D) and the axial length was 22.12 mm. Biometric measurement of the intraocular lens (IOL) power was 22.5 diopters for the left eye.

B mode ultrasonography using 20 MHz probe (Figures 5 and 6) clearly demonstrated bilateral anterior lenticonus. With these findings, it was clear that the reason for her refractive error was anterior lenticonus.

We performed phacoemulsification and intraocular lens (IOL) implantation in the left eye. Anterior capsulorhexis was started from mid-peripheral region to avoid radial tears. During capsulorhexis no difficulty was encountered. After careful hydrodelineation, nucleus material was removed with the phaco probe mostly with irrigation aspiration mode and minimal ultrasonic power.

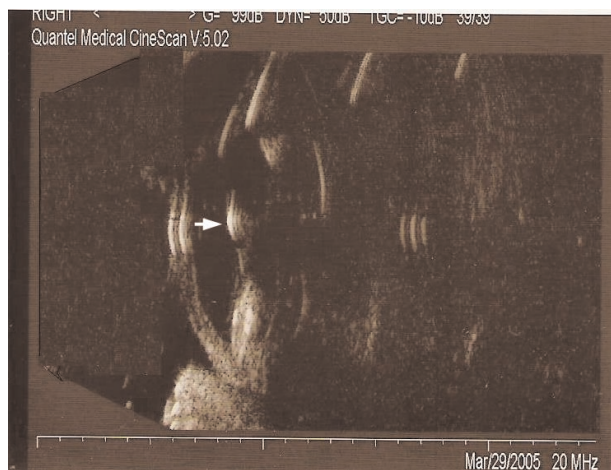


Figure 5. Ultrasonographic appearance of central lens area demonstrating anterior lenticonus in the right eye (arrow).

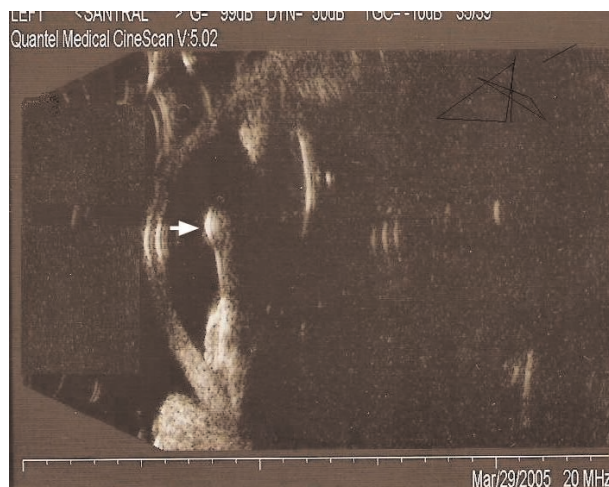


Figure 6. Ultrasonographic appearance of central lens area demonstrating anterior lenticonus in the left eye (arrow).

After cortical clean up, a 22.5 D foldable IOL was inserted in the capsular bag. Uncorrected visual acuity was 20/32 and best corrected visual acuity was 20/25 with +0.50 D spheric correction 1 week postoperatively. Since the patient did not want any operation to the right eye, this eye was fitted with a soft contact lens. Her best corrected visual acuity with the contact lens was not better compared to spectacle correction.

Discussion

Ocular abnormalities can be seen in 30% to 35% of patients with Alport's syndrome. Ocular abnormalities are rare in childhood and in females and increase with age (2,5,6). Frequent ocular findings are anterior lenticonus, posterior lenticonus, juvenile arcus, posterior polymorphous dystrophy, and macular and mid-peripheral retinal white-yellow flecks (4-6). Systemic and ocular pathologies are the results of a genetic abnormality in type 4 collagen production. In most cases, there is a mutation in COL4A5 gene in X-chromosome resulting in basal membrane pathologies (5).

Anterior lenticonus was considered as a part of Alport's syndrome by Nielsen (3). Ninety percent of patients with anterior lenticonus are expected to have Alport's syndrome so the presence of anterior lenticonus

helps to confirm the diagnosis of this syndrome. Anterior surface of lens protrudes anteriorly and histologic structure of the anterior lens capsule changes. The lens may be otherwise clean, or may show opacities. Electron microscopic studies of anterior lens capsule revealed thin and irregular basal membrane (4). Ultrastructural and immunohistochemical examination of anterior capsule material showed that lens capsule was thinner than normal, resulting in basal membrane pathologies (7).

Most of the patients with Alport's syndrome come to ophthalmology clinics with progressive deterioration of visual acuity because of high myopia due to lenticonus. Removal of lens and intraocular lens implantation is the choice of surgical therapy in these patients (8-10). In our case capsulorhexis, phacoemulsification, and intraocular lens implantation was completed successfully and visual acuity and quality increased significantly.

Anterior lenticonus can easily be overlooked. In these cases phacoemulsification and intraocular lens implantation should be the choice of surgical therapy for the refractive errors. With this case, we would like to emphasize that careful refractive and systemic evaluation of patients who are candidates for refractive surgery is of great importance and helps the surgeon choose the correct surgical intervention.

References

1. Alport AC. Hereditary familial congenital haemorrhagic nephritis. *Br Med J* 1927; 1: 504-6.
2. Gubler M, Levy M, Broyer M, Naizot C, Gonzales G, Perrin D et al. Alport's syndrome: A report of 58 cases and a review of the literature. *Am J Med* 1981; 70: 493-505.

3. Nielsen CE. Lenticonus anterior and Alport's syndrome. *Acta Ophthalmol (Copenh)* 1978; 56: 518-530.
4. Junk AK, Stefani FH, Ludwig K. Bilateral anterior lenticonus. *Arch Ophthalmol* 2000; 118: 895-897.
5. Pajari H, Setälä K, Heiskari N, Kääriäinen H, Rosenlöf K, Koskimies O. Ocular findings in 34 patients with Alport syndrome: correlation of the findings to mutations in COL4A5 gene. *Acta Ophthalmol Scand* 1999; 77: 214-217.
6. Jacobs M, Beng BJ, Kriss A, Jeffrey B, Kriss A, Taylor D et al. Ophthalmologic assessment of young patients with Alport syndrome. *Ophthalmology* 1992; 99: 1039-1044.
7. Ohkubo S, Takeda H, Higashide T, Ito M, Sakurai M, Shirao Y et al. Immunohistochemical and molecular genetic evidence for type IV collagen $\alpha 5$ chain abnormality in the anterior lenticonus associated with alport syndrome. *Arch Ophthalmol*. 2003; 121: 846-850.
8. Grondalski SJ, Bennett GR. Alport's syndrome: Review and case report. *Optometry and Vision Science*. 1989; 66: 396-398.
9. Mavrikakis I, Zeilmaker C, Wearne MJ. Surgical management of anterior lenticonus in Alport's syndrome. *Eye* 2000; 16: 798-800.
10. Sukhija J, Saini JS, Jain AK. Phacoemulsification and intraocular lens implantation in an Alport's syndrome patient with bilateral anterior and posterior lenticonus. *J Cataract Refract Surg* 2003; 29: 1834-1836.