Scholars Journal of Applied Medical Sciences
Abbreviated Key Title: Sch J App Med Sci
ISSN 2347-954X (Print) | ISSN 2320-6691 (Online)
Journal homepage: https://saspublishers.com/sjams/

OPEN ACCESS

Ophtalmology B

Spontaneous Anterior Dislocation of the Lens in Homocystinuria

Karim Amhoud*, Karmoune S, El Marzouqi B, Belaydi W, Lalaoui A, Abdallah E, Boulanouar A, Berraho A

Department of Ophtalmology B, Ibn Sina Hospital, University Mohamed V, Rabat, Morocco

DOI: 10.36347/sjams.2020.v08i10.005 | **Received:** 22.09.2020 | **Accepted:** 30.09.2020 | **Published:** 06.10.2020

***Corresponding author:** Karim Amhoud

Abstract **Case Report**

Purpose: To report an unusual case of a spontaneously dislocated crystalline lens into the anterior chamber associated with Homocystinuria**.** ***Methods:*** A 44-year-old woman with no occular trauma history presented with a spontaneously dislocated crystalline lens into the anterior chamber accompanied by corneal touch and increased intraocular pressure. The crystalline lens was extracted by phakoemulsification followed by a retro-pupillary iris claw. ***Results:*** Surgery was successful with improved visual acuity stabilized eye pressure and clarification of the cornea. ***Conclusion:*** Homocystinuria is a rare etiology of spontaneous anterior dislocation of the lens that requires multidisciplinary management.

Keywords: Dislocation - Homocystinuria - Lens.

INTRODUCTION

Homocystinuria is an inherited metabolic disease causing an accumulation of homocysteine and methionine at a toxic level in the body. Its incidence varies across regions [1]. Ranging from 1/1800 to 1/900,000. Patients show a wide spectrum of severity and age at presentation [2], the diagnosis is often delayed. Ectopia lentis is the outstanding ophthalmic feature of homocystinuria [3].

CASE REPORT

We report the case of an 44 years old femal patient, who had an emergency consultation for a blurred vision and painful right eye for 24 hours.

The patient had a history of venous thrombosis of the lower limbs 6 years ago treated with anticoagulants, with no ocular trauma history.

An uncorrected visual acuity of hand motions was found at the right eye. The slit-lamp microscopy examination revealed spontaneous crystalline lens dislocation into the anterior chamber with corneal touch, stromal edema and Descemet fold at the right Intraocular pressure was 58 mmHg at the right eye and 14 mmHg at the left.

The examination of the left eye showed uncorrected visual acuity at 2/10, superior crystalline ectopia and normal posterior segment. A general physical examination revealed no other abnormalities.

Medical treatment has been started including Intravenous Mannitol, oral acetazolamide and local hypotonics to reduce IOP, we used Mydriasis with patient supine position to redislocate the lens posteriorly.

After the eye pressure has been normalized and the cornea cleared, we proceeded to surgery to remove crystalline lens.

Iconography

Fig-1: Slit lamp photo of the right eye showing conjunctival hyperemia, with total anterior- lens dislocation with endothelial decompensation

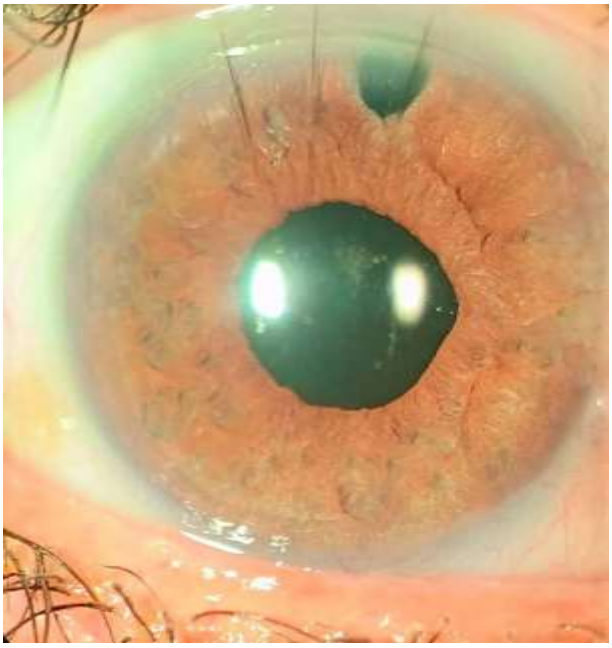

Fig-2: Post-operative image showing a posterior chamber implant attached to the iris and a peripheral iridotomy

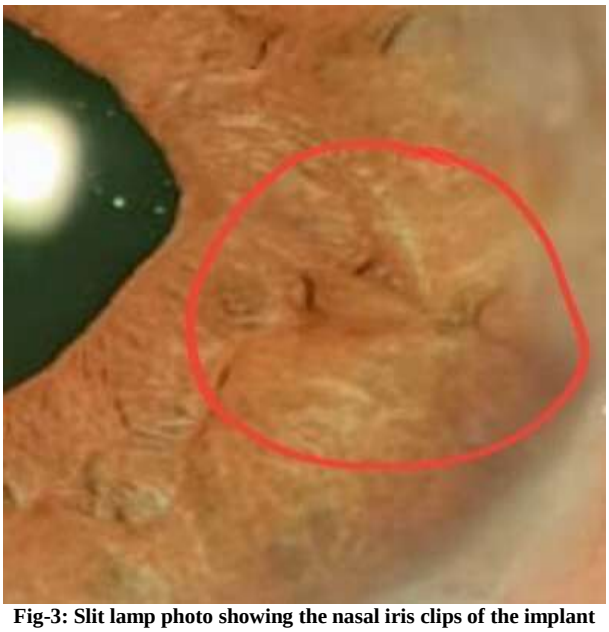

Fig-3: Slit lamp photo showing the nasal iris clips of the implant in the posterior chamber

DISCUSSION

There are many causes for anterior crystalline lens dislocation: trauma, inherited disorders, including, Marfan syndrome, homocystinuria, spherophakia, retinitis pigmentosa, pseudoexfoliation syndrome, as well as spontaneous cases [4,5]. Homocystinuria is an autosomal recessive disorder of methionine metabolism first described in 1962 [6].

Due to a deficiency in beta-synthase cystathione which it pathophysiology is not fully understood. As well as the accumulation of Homocysteine, the defect leads to increased concentrations of S-adenosylhomocysteine, enhanced remethylation to methionine, and depletion of cystathionine and cysteine [2].

Lens zonules normally have a high cysteine content, and its deficiency may affect normal zonular development, thereby predisposing to myopia and lens dislocation. Homocysteine inhibits cross-linkage in collagen and elastic tissue and predisposes to zonule degeneration [7].

Some patients have a severe childhood-onset multisystem disease, whilst others are asymptomatic into adulthood. Ocular (crystalline luxation, myopia, glaucoma), skeletal (osteoporosis, arachnodactylia), vascular (venous thrombosis, heart attack of the young subject, Cerebrovascular stroke), neuropsychiatric (mental retardation, personality disorders) and cutaneous abnormalities are the usual symptoms [2].

Thrombotic vascular occlusions constitute the main cause of morbi-mortality of these patients and increase the risk of anesthesia. The risk of thromboembolism also increases after surgical procedures [3]. Ectopia lentis is the most consistent clinical finding in CBS deficiency [8].

Other reported opthalmic complications of homocystinuria include retinal detachment, microphthalmos, optic atrophy, peripheral cystoid retinal degeneration, retinal arterial occlusions, band keratopathy, and secondary glaucoma [3].

Plasma total homocysteine should be the frontline test for diagnosis of CBS deficiency. Plasma free homocystine imeasurement is not recommended because of its low sensitivity and reproducibility [2].

Our patient has a high level of Plasma total homocysteine 112 µmol/l (N<15 µmol/l) and Plasma methionine 80 µmol/l (N<40 µmol/l). She followed a low-methionine died and was refered to an internist doctor for the dosage of vitamins and supplement treatments.

The early initiation of methionine restriction will reduce the incidence of the ectopia lentis. This dietary restriction and use of vitamins B6 and folic acid should be the first line of treatment of this disease [3].

Lens luxation into the anterior chamber is rare compared with luxation into the vitreous body. However, anterior dislocation of the crystalline lens can cause severe complications such as corneal edema and acute glaucoma. Therefore, unlike with a lens dislocated into the vitreous, an anteriorly dislocated crystalline lens should always be removed Jaffe *et al*. [9] suggest an intracapsular extraction through a limbal incision, while Peyman *et al*. [10] recommend vitrectomy with scleral incision. Choi *et al*. [11] and Seong *et al*. [12] suggest phakoemulsification with anterior vitrectomy.

In our case, Surgical treatment by a limbic incision under topical anaesthesia, a phakoemulsification was performed by using viscoelastic material in order to protect endothelial cells during surgical invasion, then we proceeded to a retro-pupillary iris claw to correct aphakia, peripheral iridectomy and anterior vitrectomy.

However, due to frequently associated high intraocular pressure, surgical treatment shows higher risk of expulsive hemorrhage and endothelial cell loss [13].

CONCLUSION

Spontaneous anterior dislocation of the lens is a rare antity, it may be secondary to homocystinuria, a metabolic disease requiring multidisciplinary management.

It is an ophthalmological emergency that needs rapid treatment to avoid severe complications such as corneal endothelium damage and acute glaucoma.

REFERENCES

1. Gascon P and Spontaneous bilateral dislocation of the crystallinelens in the case of a patient with homocystinuria, withanterior chamber OCT showing endothelial damage.
2. Andrew A. M. Morris and al.Guidelines for the diagnosis and management of cystathionine beta-synthase deficiency J Inherit Metab Dis. 2017; 40:49-74
3. Harrison. Management of Ophthalmic Complications of Homocystinuria Ophthalmology. 1998;105:1886–1890
4. Jarett WH. Dislocation of the lens. A study of 166 hospitalized casent. Arch Ophthalmol. 1967; 78(3):289–296
5. Kwon YA, Bae SH, Sohn YH. Bilateral spontaneous anterior lens dislocation in a retinitis pigmentosa patient. Korean J Ophthalmol.2007; 21:124-126
6. Gerritsen T. Vaughn JG, Waisman HA. The identification of homocystine in the urine. Biochem Biophys Res Common. 1962:9: 493-6.
7. Burke JP and al. Ocular complications in homocystinuria -early and late treated British Journal of Ophthalmology. 1989, 73, 427-431
8. Mudd. Vascular disease and homocysteinemetabolism. N Engl J Med.1985; 313:751-753.
9. Jaffe NS, Jaffe MS. Cataract Surgery and its Complications. 6th ed. St. Louis, MO: Mosby-Year Book; 1997.
10. Peyman GA, Raichand GA, Goldberg MF, Ritacca D. Management of subluxated and dislocated lenses with the vitrophage. Br J Ophtalmol. 1979; 63:771-8.
11. Choi DY, Kim YG, Song BJ. Surgical management of crystaline lens dislocation into the anterior chamber with corneal touch and secondary glaucoma. J Cataract Refract Surg. 2004; 30:718-21.
12. Seong M, Kim MJ, Tchah H. Argon laser iridotomy as a possible cause of anterior dislocation of a crystaline lens. J Cataract Refract Surg. 2009; 35:190-2.
13. M. Garza-Leon Medical treatment of crystalline lens dislocation into the anterior chamber in a patient with Marfan syndrome Int Ophthalmol. 2012; 32:585–587.