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Lens Dislocation and Phacoantigenic Glaucoma Secondary to Ocular Trauma in a 54-Year-Old Male: A Case Report and Literary Review

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ABSTRACT

Phacoantigenic glaucoma (previously known as phacoanaphylactic glaucoma) is a rare condition in which a patient presents a sensitization to their own ocular lens proteins after surgery or trauma that produces an inflammatory response. The clinical picture is variable, but the majority of patients present a moderate reaction in the anterior chamber. It is recommended the initiation of topical corticosteroids and aqueous suppressors with the goal of reducing the inflammatory response and the intraocular pressure. Surgical removal of the residual material may be necessary once the inflammation is under control as presented in the following case. The description of a clinical case and a review of the topic are carried out considering the little literature available on this pathology, due to reasons such as infrequency and scarce publication of this type of medical case. This report describes a clinical case and reviews the limited literature on this pathology, aiming to provide a summary of available information and improve early intervention and prognosis.

Keywords: Lens subluxation, glaucoma, cataract, vitrectomy.

Introduction

Lens dislocation is a relatively infrequent event [1], this can be partial (subluxation) or complete (dislocation)[1] depending on its etiology. It can be classified congenital (ectopia lentis); associated with alterations in connective tissue like Marfan´s syndrome or it can be acquired, which can be iatrogenic; for example, during the extraction of cataracts or ocular trauma. Ocular trauma, being the most common acquired cause of all cases of lens dislocation [4].

The main clinical indications of lens dislocation are a reduction in visual acuity and diplopia in the affected eye in most cases. The dislocated lens can be in the posterior chamber; however, it can also become lodged in the anterior chamber[1]. Ultrasound is a highly useful diagnostic tool in emergency services, providing a quick and economical means for precise and timely diagnosis, especially when fundoscopy is not feasible.[1].

On the other hand, phacoantigenic glaucoma, previously known as phacoanaphylactic glaucoma, is the rarest form of lens induced glaucoma and generally presents a grand diagnostic challenge. It is caused by an Arthus-type immune response, mediated by immunoglobulin G (IgG) and a complement system against lens proteins. [5,6]. The predominant clinical manifestations are palpebral edema, chemosis, conjunctival injection, corneal edema, anterior and posterior synechia, loss of vision and pain [5,6].

Definitive diagnosis is confirmed by the presence of polymorphonuclear leukocytes and lens protein in samples of vitreous humor or aqueous cultures and Gram stains.[6]. Initial management focuses on controlling intraocular pressure (IOP) and inflammation with topical corticosteroids and antiglaucoma medications. However, in most cases, surgical intervention is necessary to remove lens material. [5,6].

Next, we present a case of blunt trauma with intraocular lens displacement into the posterior chamber, associated with phacoantigenic glaucoma in a 54-year-old male. This case is atypical, as the cause is trauma rather than cataract extraction surgery, which is the more common cause. The patient's condition, clinical diagnosis based on provided data, and favorable evolution after immediate surgical intervention are discussed.

Clinical Case

We introduce a 54-year-old male patient from Montelibano, Cordoba, Colombia, with a past medical history of cataracts for the past four years. He presented for urgent care after experiencing blunt trauma from a tree branch to the right eyeball three months ago, resulting in subsequent blurred vision. However, he did not seek medical attention until the current date, exhibiting intense pain and loss of vision in the affected eye. The patient was assessed by a specialist in ophthalmology, who noted pterygium in both eyes, clear corneas, transparent formed anterior chambers (AC), and reactive pupils with mydriasis. The left eye (OS) was normotonic with 0.3 excavation. Right eye (OD) with afaquia, while the right eye (RE) exhibited aphakia and slightly elevated intraocular pressure (IOP) due to visible ocular trauma complicated by lens dislocation. The patient was prescribed dorzolamide and timolol, 1 drop every 12 hours, and required an assessment by a retinal specialist, who confirmed aphakia, lens dislocation, posterior pole, posterior vitreous detachment (PVD), dislocated cataracts, and phacoantigenic glaucoma. A surgical procedure consisting of posterior vitrectomy, phacofragmentation, and cataract extraction by phacoemulsification with intraocular lens implantation was performed. Patient tolerated procedure without any complications.

Discussion

This condition was first described as phacoanaphylactic endophthalmitis by Straub in 1919 and later by Verhoeff and Lemoine in 1922. They described a severe uveitis that developed in 1 to 14 days after the leak of lens material to the anterior chamber[7]. At that time, the term 'anaphylactic' was used due to the sudden appearance of an inflammatory process. Whereby during many years, it was believed that phacoanaphylactic endophthalmitis is a response to the rejection of foreign matter[7]. Johann Sebastian Bach (1685-1750) was one of the greatest composers of all time. Besides being a brilliant organist, he composed more than 1,100 compositions in almost all musical genres. He was recognized as a hard worker and a devout Christian. Persuaded by friends, he underwent cataract surgery performed by a British ophthalmologist. After the surgery, Bach became completely blind and could no longer play the organ, compose, or direct choirs and orchestras. He suffered immense pain in his eyes and body and died four months after the surgery. [8,9].

In conclusion, a secondary untreatable glaucoma might have caused Bach's blindness and symptoms. This case is one of the most relevant in the history of ophthalmology. Although there are questions about the true nature of Bach's ocular disease and the details of the operation, it is unclear whether the surgery or postoperative treatment led to his death.[9].

In the literature, the incidence of phacoantigenic glaucoma after traumatic ocular lesions ranges from 0.1% to 3% and approximately 0.01% after intraocular surgery. Among iatrogenic causes, vitreoretinal surgery has the highest rate of this condition, presumably due to alterations in the blood-retinal barrier and damage to retinal and choroidal tissues, which are susceptible to traction, atrophy, and chronic inflammation. In 90% of the patients, the disease developed in the upcoming year since the event that triggered it, and it is associated with the risk of potential bilateral blindness. Typical symptoms include bilateral visual impairment with photophobia, dull pain, and photopsia.[10].

It has been proposed that the disease develops when lens proteins are exposed to the immune system due to the disruption of the lens capsule. A series of careful studies that provided contradictory evidence and some hypotheses suggest that it is not a rejection mediated by cells of strange substances of a tissue[7]. Phacoantigenic glaucoma is an inflammatory reaction directed against lenticular antigens, with elevated intraocular pressure (IOP) due to secondary obstruction by inflammatory

cells. It is important to note that 'phacoanaphylactic' is considered an incorrect term since this condition is not an allergic reaction. The mechanism that causes the reaction seems like a reaction of the immune complex type Arthus mediated by IgG and its complementary system [7,9]. Patients develop a response where they are sensitized to their own lens antigens, which are usually sequestered within the lens capsule. After ocular surgery or trauma to the lens capsule, these antigens become exposed to the circulation and may be recognized as foreign by the immune system, triggering an inflammatory response[11,12].

A less common mechanism is the formation of synechiae that cause pupillary block. It is unclear why some patients develop this reaction while others do not. It has been proposed that this condition is induced by a T-cell-mediated response to lens degradation, typically maintained by small amounts of circulating lens proteins. Bacterial lipopolysaccharide is mitogenic to B lymphocytes and can inhibit necessary T-cell tolerance. If B lymphocytes recognize the antigens, they may be stimulated by lipopolysaccharides introduced by wound contamination, causing the disease without prior sensitization [12].

In certain cases, infection at the lens site is a different focus of infection, eliciting a granulomatous response. The response to the lens may be caused by infectious agents like Propionibacterium acnes, which can cause granulomatous inflammation in intraocular implants and produce a condition similar to phacoantigenic glaucoma, termed pseudo-phacoantigenic glaucoma. The interval between trauma and the onset of inflammation ranges from 24 hours to 14 days. In the presented case, the patient sought urgent care three months after the traumatic event, which is atypical. However, the interval can vary from months to years after traumatic capsular rupture or surgical procedures.[7].

The clinical signs include reduction of vision, palpebral edema, chemosis, conjunctival injection, corneal edema, intense response of the anterior chamber, and posterior synechiae[13]. The presentation and signs observed in our patient are consistent with the diagnosis. The inflammation usually is uniocular and affects the eye that has been traumatized. You can use ultrasound to detect inflammation centered around the lens or inside the vitreous. The diagnostic can be confirmed through the study of the anterior chamber or aspirated vitreous that contains fragmented material of the lens[7,14]. Initial treatment is immediate systematic therapy with high doses of steroids. The course of the disease usually relapses to progressively chronic. Immunomodulators like ciclosporin A, azathioprine, cyclophosphamide, and mycophenolate mofetil and biological products are utilized each more and contribute a significant better prognostic of the disease [7,15].

The treatment focuses on controlling inflammation and performing surgical intervention as soon as possible. In our patient, after the surgical removal of the lens, the inflammation was quickly controlled, preventing further disease progression. Due to the similarity of the patient's symptoms to infectious endophthalmitis and the prolonged course of the disease, diagnosing endophthalmitis and phacoantigenic glaucoma can be challenging. Clinically, for the identified patients, a vitrectomy, complete removal of the residual lens, and anti-inflammatory therapy should be performed to further control the inflammation. There are many challenges in managing phacoantigenic glaucoma, and little evidence is published about this condition. Surgical treatment and timing should be carefully considered. A holistic approach was adopted, and a detailed discussion was held with the patient about the various surgical options, which received a positive response.

Conclusion

Lens dislocation and phacoantigenic glaucoma are relatively rare conditions, with the latter being the rarest form of lens-induced glaucoma. It occurs in less than 1% of cataract surgeries. Managing this rare and complicated condition is challenging, especially with few documented cases and limited literature on the subject. Despite the various methods available for assessment, this pathology presents a significant diagnostic challenge. Therefore, it is crucial to recognize and provide timely treatment to reduce potential consequences.

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