Ophthalmology Research: An International Journal

7(2): 1-6, 2017; Article no.OR.35617
ISSN: 2321-7227

Acute Unilateral Intraocular Pressure (IOP) Elevation in the Emergency Room (ER): A Case Report

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Authors' contributions

This work was carried out in collaboration between all authors. Author AL designed the study and wrote the first draft of the manuscript. Authors DS and MM managed the literature searches. Authors CV, ML and FV carried out revision of the manuscript. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/OR/2017/35617
Editor(s):
(1) Jui-Teng Lin, Chairman & CEO of New Vision Inc, Taiwan and HE Medical University, China.
Reviewers:
(1) Valentin Huerva, Universitary Hospital Arnau de Vilanova, Lleida, Spain.
(2) Paaraj Dave, Dave Eye Clinic and Dr TV Patel Eye Institute Consultant and Head Gujarat, India.
Complete Peer review History: http://www.sciencedomain.org/review-history/20774

Case Study

Received 21st July 2017
Accepted 23rd August 2017
Published 1st September 2017

ABSTRACT

Aim: To present a clinical case of acute unilateral intraocular pressure (IOP) elevation in the emergency room (ER) that required a complex differential diagnosis, including acute angle closure, trauma, hypertensive uveitis, Posner-Schlossman Syndrome (PSS) or Glaucomatocyclic Crisis and Pigmentary Dispersion Syndrome (PDS).

Presentation of Case: A 15-year-old myopic female presented in the ER with two acute episodes of IOP elevation in the right eye - OD (40-50 mmHg), with mild discomfort and red eye and mild blurred vision in both episodes. Slit lamp evaluation of the OD revealed in the first episode mild pigment in the inferior portion of the endothelium and anterior chamber (AC), white and thin keratic precipitates in the second episode and tyndall in both episodes. Gonioscopy performed in the first episode revealed a thick pigmented trabecular meshwork and ultrasound biomicroscopy (UBM) revealed a mild anterior concavity of the peripheral iris. A laser iridotomy was performed in the first episode and an association of steroids and hypotensive medication was performed in both episodes.

Discussion: After a differential diagnosis analysis, a diagnosis of PDS was made in the first episode and a diagnosis of PSS was made in the second episode.

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Conclusion: This clinical case recalls the complexity of some clinical cases, questioning us about the presence of two different pathologies in the same eye, or whether it is the same pathology. Thus, it highlights the need of integration of clinical signs and symptoms to allow a correct differential diagnosis, treatment and follow-up.

Keywords: Acute unilateral IOP elevation; Posner-Schlossman syndrome; Pigmentary Dispersion syndrome; herpetic uveitis.

1. INTRODUCTION

Episodes of acute unilateral intraocular pressure (IOP) elevation often appear in the emergency room (ER) and a correct differential diagnosis is needed to allow the appropriate treatment and follow-up. Some examples are acute angle closure, trauma, hypertensive uveitis, Posner- Schlossman Syndrome (PSS) or Glaucomatocyclic Crisis and Pigmentary Dispersion Syndrome (PDS), which may be unilateral, but is most frequently bilateral [1,2].

A common type of hypertensive uveitis is the herpetic uveitis, caused most commonly by herpes simplex and herpes zoster virus infection. Clinically there is a sudden onset of severe ocular pain and photophobia. Slit lamp observation reveals ciliary hyperemia, keratitis (although not a mandatory find) with a variable anterior chamber (AC) reaction, iris atrophy (a characteristic finding, which may be sectorial or diffuse in more chronic cases) and acute IOP elevation. The treatment includes an association of topical steroids, antiviral medication (for prevention of viral replication during steroids treatment) and hypotensive medication (if the IOP elevation persists despite the reduction of the inflammation, it contributes to the reduction of IOP) [1-3].

The PSS, more common in young and middle-aged patients, with unknown and controverse etiology, which can namely include the herpes vírus infection (it has been implicated without prior herpetic keratitis) and the cytomegalovirus infection, CMV (its involvement in PSS is increasingly evident nowadays) [4]. It is characterized by recurrent episodes of acute IOP elevation and previous mild inflammation without precipitating factors and with spontaneous resolution. A key aspect of this syndrome is the contrast between mild signs and symptoms and an acutely elevated IOP. Clinically it is characterized by mild hyperemic or even white eye associated with an ocular discomfort and vision with halos. Slit lamp evaluation include the presence of small, round, white, thin keratic precipitates with a slight AC reaction in association with elevated IOP. Topical treatment is only given during the acute episodes, including an association of steroids and hypotensive medication [1-4,5].

The PDS is more common in myopic young male and it can be unilateral but is most frequently bilateral [6,7]. It results from the friction between the pigment epithelium of the iris and the zonule and the anterior lens capsule (associated with an anterior concavity of the peripheral iris), with resultant dispersion of pigment granules in the anterior segment of the eye. Precipitating factors of this mechanism are mydriasis and the practice of physical exercise. It is characterized by the association between mild signs and symptoms, namely ocular discomfort and blurred vision with halos, and an examination characterized by white eye, presence of pigment in the anterior segment and elevated IOP. It is characteristic (but not specific) the presence of a thick pigmented trabecular meshwork on gonioscopy and the presence of an anterior concavity of the peripheral iris in gonioscopy and in ultrasound biomicroscopy (UBM). Treatment with laser peripheral iridotomy when performed in the early stages seems to play an important role in the prevention of progression to glaucoma, given its involvement in the pathophysiological mechanism of this disease [1,2,6-10].

2. PRESENTATION OF CASE

A 15-year-old myopic female came to the ER complaining of blurred vision with halos, mild red eye and discomfort in OD, with sudden onset on that day. She referred having done physical exercise the day before. She hadn´t previous ocular trauma, neither signs and symptoms in the left eye (OS). She reported a similar episode a year ago with spontaneous resolution, without other relevant ophthalmologic history. She had no relevant systemic and family history, and hadn´t routine medication. Maximum corrected visual acuity was 10/10 bilaterally. Slit lamp

evaluation revealed in OD (Fig. 1) mild conjunctival hyperemia, transparent cornea without precipitates but with pigment in the lower portion of the endothelium, deep AC with pigmented tyndall ++, regular iris in a little reactive half-mydriasis, without posterior synechiae and with transparent crystalline. There was no alteration in the slit lamp examination of the OS. She had an IOP of 48-50 mmHg in OD and 19 mmHg in OS. Fundus examination was normal bilaterally. Gonioscopy revealed in OD a 4 grade open angle in the Shaffer classification in all quadrants, a thick pigmented trabecular meshwork, little iridian processes at 2-5 hours, and an anterior concavity in the peripheral iris. She was treated with topical Brimonidine 2mg/mL - Edol®, topical association of Brinzolamide and Timolol 10mg/mL+5mg/mL - Alcon®, Dexamethasone 1mg/mL - Théa® (4 topical administrations per day) and Acetazolamide 250mg (2 oral administrations per day), with an IOP decrease to 19 mmHg and improvement of the signs and symptoms. Then she performed an UBM (Fig. 2), that revealed a deep AC and iris with a slight anterior concavity in the periphery. Optical coherence tomography (OCT) of the optic disc highlighted the nerve fibers layer of the retina of the OD with inferior and inferior temporal borderline defects (Fig. 3). She performed a bilateral laser YAG peripheral iridotomy, after which she remained asymptomatic with IOP values of 18-19 mmHg without medication. She performed another UBM (post laser YAG), that revealed a slight flattening of the peripheral iris but there wasn´t an evident or significant change from the image of the first UBM.

After 5 months of absence of symptoms and alterations in the slit lamp examination, she came again to the ER with a new episode of acute IOP elevation of the OD (40 mmHg). Slit lamp evaluation revealed in OD (Fig. 4) mild conjunctival hyperemia, rounded and white inferior endothelial keratic precipitates, with tyndall +++ and without pigment. Fundus examination was normal and the OCT of the OD optic disc remained without relevant alterations. She presented clinical improvement after performing the same therapeutic approach of the first episode and started follow-up in the Ocular Inflammation consultation of the Ophthalmology Department and in the consultation of Autoimmune Diseases of the Pediatric Department. Complementary diagnostic exames that were performed for etiological investigation (chest x-ray, Mantoux test and blood tests including hemogram, coagulation, ionogram and infectious including CMV serologies, inflammatory and autoimmune parameters) didn't reveal any alteration.

Currently, the patient maintains follow-up in those consultations, remains asymptomatic and with IOP values of 16 mmHg bilaterally without medication and with the following refraction: OD -3.00D (visual acuity=9/10), OS -2.50D (visual acuity=10/10).

3. DISCUSSION

The discussion of this clinical case was divided in three parts, based on an initial global evaluation, followed by a specific evaluation of each episode and a final global evaluation of the clinical case.

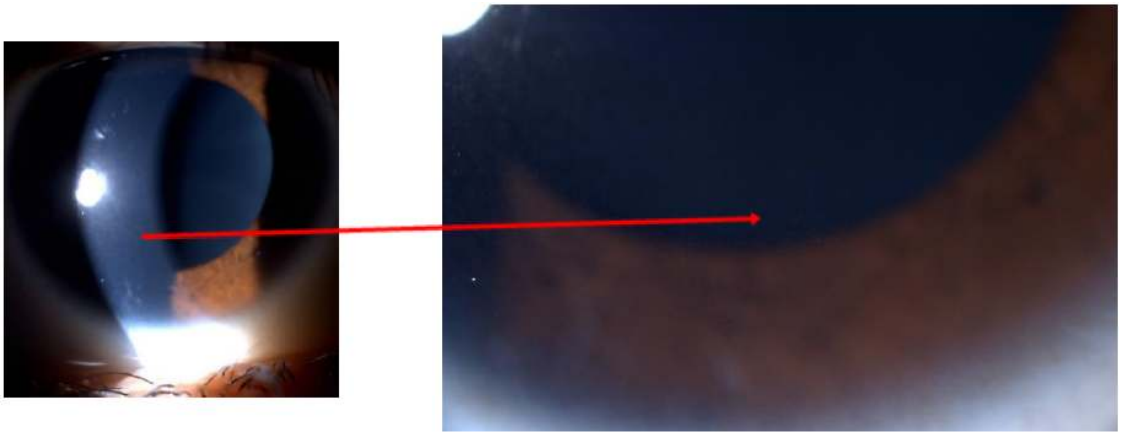

Fig. 1. Slit lamp examination of the OD: Mild pigment in the inferior portion of the endothelium and AC

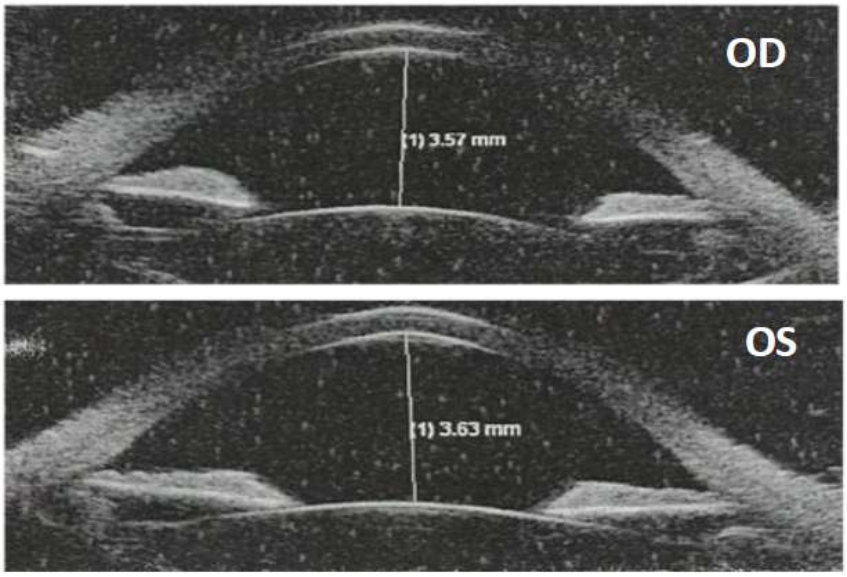

Fig. 2. UBM: Mild anterior concavity of the peripheral iris

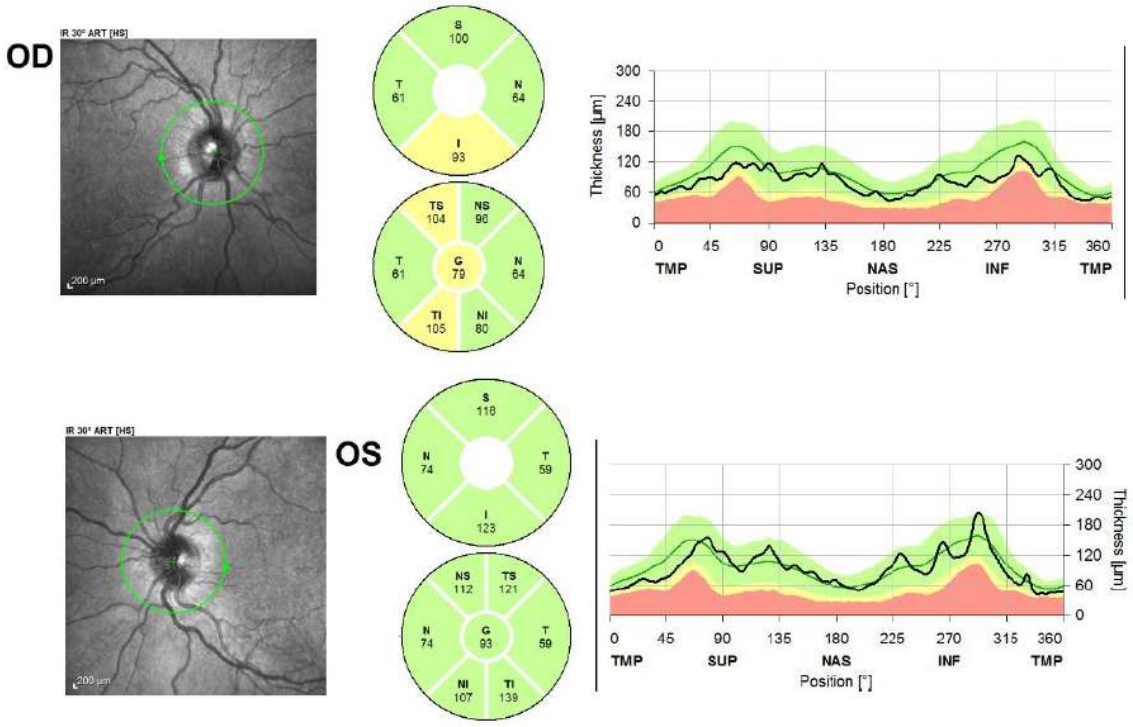

Fig. 3. OCT of the optic disc

3.1 Initial Global Analysis

The main diagnostic hypotheses considered were PSS, PDS and herpetic uveitis, considering the following factors in favor and in common to these hypotheses: The unilaterality of the clinical case (although PDS is bilateral more frequently), the recorrence of the episodes (the patient reported a similar episode a year ago, in addition to the two episodes presented and observed in the ER), the patient's signs and symptoms (halos, eye discomfort and mild red eye) and the

acute IOP elevation. There are also common factors between PSS and herpetic uveitis: the presence of AC reaction and the improvement of the signs and symptoms after steroids and hypotensive treatment. Regarding the referred factors, herpetic uveitis appeared to be the least likely hypothesis, since the symptoms presented by the patient were more slight than the most typical in this type of uveitis and given the patient to have improved in a short term without recourse to anti-viral medication (although not the main one, it is an important adjuvant and usually used in the treatment of herpes virus infection) [3]. Still in relation to the referred factors, the initial analysis of the other two hypotheses identifies the personal antecedent of myopia and the history of physical exercise as potential precipitating factors for the acute episodes associated to the PDS hypothese, whereas the presence of AC reaction and the discrepancy between mild signs and symptoms and sudden IOP elevation are very suggestive of PSS.

Fig. 4. Slip lamp examination of the OD: Keratic precipitates

3.2 Analysis of Each Acute Episode

In the first episode, the diagnostic hypothesis primarily excluded was the herpetic uveitis regarding the absence of the characteristic iris defects and keratitis (although not mandatory as referred before). Among the other two diagnostic hypotheses, the PDS appeared to be the most likely hypothesis regarding the typical (though not specific) presence of pigment at the slit lamp examination, the thick pigmented trabecular meshwork and the iridian processes in the gonioscopy, the characteristic anterior concavity of the peripheral iris in both gonioscopy and UBM, and also regarding the clinical improvement and the IOP normalization following laser peripheral iridotomy. However, it was remained in mind that the UMB revealed only a slight anterior concavity of the peripheral iris, and so not translating into the more typical and obvious image in PDS. On the other hand, the change in the UBM, even if mild, in associated with the clinical picture of the patient, made us keep the hypothesis of PDS. It was also considered the possible effect of the Dexamethasone drops in reducing the inflammation and so the possibility of masking the PSS picture. After the second episode, the PDS diagnosis was rethought and was compared again to the other two hypotheses. The hypothesis of herpetic uveitis was also excluded primarily, mainly by the mild signs and symptoms and the presence of factors more suggestive of the other diagnostic hypotheses. However, as referred before the herpes vírus infection has been implicated in the PSS without prior herpetic keratitis. The PSS appeared more likely in the second episode, regarding the absence of pigment at the slit lamp examination, the presence of keratic precipitates with the characteristics described before and the presence of AC reaction. The presence of patent iridotomies doesn´t suggest an acute episode of PDS, although this isn´t universal [5]. The absence of symptoms and medication performed between the acute episodes is another factor supporting the PSS hypothesis. The CMV involvement in PSS is increasingly evident nowadays and so an anterior chamber tap for CMV PCR could have been performed; however, the CMV serologies performed were negative.

3.3 Final Global Analysis

According to that, PDS was considered the most likely diagnosis in the first episode and PSS in the second episode.

4. CONCLUSION

In conclusion, this clinical case is a diagnostic challenge, questioning us about the presence of two different pathologies in the same eye, or whether it is the same pathology. It recalls the complexity of some clinical cases, highlighting the importance of performing a complete differential diagnosis to allow a correct treatment and to maintain an adequate follow-up of the patients.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
http://sciencedomain.org/review-history/20774