



## Spontaneous posterior dislocation of the cataractous lens in a patient with Parkinson-plus syndrome

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### ABSTRACT

**Purpose:** To report a case of unilateral posterior dislocation of the cataractous lens and subluxation of the lens in the fellow eye of a patient with Parkinson-plus syndrome.

**Observations:** A 67-year-old-man who was a known case of Parkinson-plus syndrome on long-term dopamine agonists and anti-psychotic medications demonstrated apraxia of lid opening associated with moderate-to-severe blepharospasm. He had unilateral posterior dislocation of the cataractous lens and subluxation of the lens in the fellow eye with no prior history of trauma or other known ocular risk factors.

**Conclusion and importance:** This case may represent an unusual example of spontaneous lens dislocation secondary to apraxia of lid opening and concurrent blepharospasm, which is associated with Parkinson-plus syndrome.

### 1. Introduction

The lens is considered dislocated when it lies entirely outside the hyaloid fossa of the vitreous body. In the absence of trauma, dislocation or subluxation of the lens should evoke suspicion for any hereditary systemic disease such as Marfan syndrome, homocystinuria, Ehlers–Danlos syndrome, or associated ocular disorders.<sup>1</sup> Spontaneous luxation occurs due to the rupture of the zonular fibres in degenerative and inflammatory conditions such as long-standing glaucoma, high myopia, hypermature cataract, retinal detachment, and pseudoexfoliation syndrome.<sup>2</sup> However, spontaneous posterior dislocation of the lens with no prior history of trauma or other known risk factors is a very rare occurrence. We report a case of unilateral spontaneous posterior dislocation of the cataractous lens and subluxation of the lens in the contralateral eye of an elderly patient with Parkinson-plus syndrome.

### 2. Case report

A 67-year-old man presented to us with severe headache of 5 days duration and blurring of vision for 3 months. There was no history of any ocular trauma, surgery, or long standing ocular disease. He was a known case of Parkinson-plus syndrome, major depressive disorder, obstructive

sleep apnoea, hypertension, and hypothyroidism, and was on treatment with levodopa (100mg), carbidopa (25mg), escitalopram (10mg), pramipexole (25mg), enalapril (2.5mg), and thyronorm (25µg) for the past 12 years with several modifications in the drug regimen depending on his systemic condition. He had been examined elsewhere and treated with intravenous mannitol (250ml) a day earlier and was prescribed oral acetazolamide (250 mg) thrice a day, topical timolol maleate 0.5% twice a day, brimonidine tartrate 0.2% twice a day, and dorzolamide 2% thrice a day in both eyes.

On examination, he demonstrated apraxia of lid opening (ALO; frequent spasms of the orbicularis oculi muscles, procerus, and corrugators bilaterally, causing forcible eyelid closure) associated with moderate-to-severe blepharospasm (Fig. 1). He had bilateral brow ptosis and dermatochalasis. The corrected distance visual acuity (CDVA) was 20/120 in the right eye and 20/60 in the left eye. Intraocular pressure (IOP) was 27 mmHg and 13 mmHg in the right eye and left eye, respectively, by Goldmann applanation tonometry. The right eye showed circumcorneal ciliary congestion, corneal epithelial oedema, a shallow anterior chamber with grade 2 cells, iris stromal atrophic patches, mid-dilated non-reacting pupil, and nuclear sclerotic cataract. The left eye also had a shallow anterior chamber and nuclear sclerotic cataract. Gonioscopy revealed a 360° and 180° appositional angle

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closure in the right eye and left eye, respectively. The fundus examination was unremarkable with a cup disc ratio of 0.5:1 and healthy neuroretinal rim in both eyes.

A clinical diagnosis of bilateral drug-induced angle closure secondary to escitalopram was made. The angle closure mechanism was likely due to the anticholinergic effects of the drug leading to pupillary dilatation, and increased aqueous production secondary to increased ciliary body blood flow. The patient underwent Nd: YAG laser peripheral iridotomy in both eyes, following which the IOP came down to the normal range. After 1 month, phacoemulsification with intraocular lens implantation in the right eye was planned. A detailed slit-lamp examination was challenging to perform due to ALO. On the operating table, a significant inferior zonular dehiscence more than 6 clock hours and posterior subluxation of the cataractous lens was observed. A pars plana vitrectomy and lensectomy with fragmatome were then performed in the right eye, and he was left aphakic. Postoperatively, his CDVA improved to 20/80 in the right eye with +11.00D, and IOP was 13 mmHg. He was doing well in subsequent follow-ups. During a routine follow-up after 1 year, the cataractous lens in the left eye was found to be dislocated posteriorly in the anterior vitreous with evidence of vitritis. He was treated with topical corticosteroids in a tapering regime, resulting in the reduction of vitritis. However, the patient could not be taken for ocular surgery as he was not systematically fit for surgery.

### 3. Discussion

Parkinson-plus syndrome is a group of heterogeneous degenerative neurological disorders, which differ from the classical idiopathic Parkinson's disease in certain associated clinical features, inadequate response to levodopa, distinctive pathological characteristics, and poor prognosis. Associated clinical features include symmetrical onset, infrequent or atypical tremor, prominent rigidity in axial musculature,

bradykinesia, early postural instability, supranuclear gaze palsy, early autonomic failure, pyramidal affection, cerebellar involvement, alien limb phenomenon, apraxia, and significant early cognitive dysfunction in a few cases. Progressive supranuclear palsy, multiple system atrophy, and dementia with Lewy body disease are commoner disorders. Less frequent disorders are cortico-basal ganglionic degeneration, frontotemporal dementia with chromosome 17, Pick's disease, parkinsonian-dementia complex of Guam, Pallidonigral degeneration, Wilson's disease and a rigid variant of Huntington's disease.<sup>3</sup> In the present case, the patient was suffering from Parkinson-plus syndrome, on treatment with dopamine agonists and anti-psychotic medications for the past 12 years.

Spontaneous posterior dislocation of the cataractous lens in the absence of any trauma and known hereditary systemic associations or ocular risk factors is indeed a rare entity with only one case reported to date.<sup>4</sup> Saikumar et al. described a case of bilateral spontaneous posterior dislocation of the cataractous lens in a 64-year-old man who was on long-term anti-psychotic medications with no prior history of trauma or other risk factors. However, the exact pathophysiology or mechanism of lens dislocation was not clearly understood in this case, though a relation between anti-psychotic medications and zonular weakness has been proposed.<sup>4</sup>

Bilateral spontaneous dislocation of the clear crystalline lens into the anterior chamber in a 17-year-old male has been reported by Jovanović. However, there was no vitreous prolapse or visible defect of the anterior hyaloid membrane, for which the author had no explanation. The case was managed by lens removal, followed by contact lens correction for aphakia.<sup>5</sup> Posterior dislocation of the lens into the vitreous with excursions to the anterior chamber was observed by Schäfer et al. in two patients, one of whom had Marfan syndrome, and the other had a history of blunt trauma, both of which are known risk factors for this condition.<sup>6</sup> Brown et al. reported a case of a 68-year-old woman on anti-psychotic medications who was found to have a dislocated lens in one eye,



Fig. 1. Patient showing apraxia of lid opening associated with moderate-to-severe blepharospasm.

possibly following repeated fist punches on that side of her face in an attempt to rid herself of her distressing auditory hallucinations.<sup>7</sup>

Recently, some studies have shown that nearly 30% of the patients with Parkinson's disease are associated with elevated plasma homocysteine levels, and high homocysteine may be a feasible therapeutic target for cognitive decline in these patients. Raised homocysteine levels may lead to the progression of Parkinson's disease through multiple pathways, including nerve cell apoptosis, oxidative stress, and DNA damage.<sup>8</sup> Also, homocystinuria may be associated with decreased zonular integrity due to the enzymatic defect.<sup>1</sup> In the present case, however, the homocysteine level of the patient was within the normal range.

In our case, the patient had moderate-to-severe blepharospasm with concurrent ALO, which is known to occur as a feature of Parkinson-plus syndrome.<sup>9</sup> Eyelid closure is associated with eye movement generating an extorsional, downward, and inward gaze movement. This is followed by eyelid opening, causing a consistent pulse-like movement in the intorsional, upward, and outward direction.<sup>10</sup> The blink-related oblique eye movements associated with increased blinking for a long period of time in this patient may have generated zonular stress in a nasal and downward direction. The prolonged contraction of the orbicularis oculi muscle might have facilitated the alteration in previously stressed zonules and contributed to lens subluxation and dislocation.

We did not implant IOL in this case, considering the systemic condition of the patient and the possibility of IOL dislocation later due to the same reasons, which can further worsen the general condition of the patient.

ALO coexisting with blepharospasm often respond to botulinum toxin injections into the pretarsal orbicularis muscle or at the junction of the pretarsal and preseptal orbicularis muscle.<sup>11</sup> In patients who are refractory to botulinum toxin injections, other supportive therapies, such as eyelid crutches and surgical myectomy, could be considered.<sup>12</sup> Augmented systemic dopaminergic therapy may also benefit patients who have Parkinson's disease.<sup>13</sup>

A relatively similar case of bilateral intraocular lens (IOL) dislocation has been described in a patient with essential blepharospasm by Moreno-Montañés et al.<sup>14</sup> Spontaneous zonular fibre rupture leading to IOL dislocation has been reported in two cases after exposure to low-frequency vibrations of medium and high intensity (15–40 Hz) for a long time.<sup>15</sup> Also, IOL subluxation or dislocation may occur years after cataract surgery in patients with chronic uveitis.<sup>16–18</sup>

The limitation of this case report is the lack of information regarding the patient's past ocular examination. The details of ocular abnormalities diagnosed before, if any, which could contribute to zonular weakness, were not available.

#### 4. Conclusion

To the best of our knowledge, this is the first reported case of spontaneous cataractous lens dislocation secondary to ALO with concurrent blepharospasm. The present case suggests that patients with atypical or idiopathic Parkinson's disease on long-term dopamine agonists and anti-psychotic medications should be considered as those with a potential risk of having a zonular weakness. The presence of ALO associated with blepharospasm in such patients may have a detrimental effect on zonular fibres, generating zonular stress and leading to spontaneous lens subluxation and dislocation.

#### Patient consent

Informed consent has been obtained from the legal guardian of the patient.

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#### Intellectual property

We confirm that we have given due consideration to the protection of intellectual property associated with this work and that there are no impediments to publication, including the timing of publication, with respect to intellectual property. In so doing we confirm that we have followed the regulations of our institutions concerning intellectual property.

#### Research ethics

Written consent to publish potentially identifying information, such as details or the case and photographs, was obtained from the patient(s) or their legal guardian(s).

#### Authorship

All listed authors meet the ICMJE criteria. We attest that all authors contributed significantly to the creation of this manuscript, each having fulfilled criteria as established by the ICMJE.

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#### Declaration of competing interest

None.

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