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A rare case of persistent idiopathic adie's tonic pupil



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ABSTRACT

Background: Adie's Tonic Pupil is a condition of a tonically dilated pupil, which is unresponsive to bright light followed by moderate responsiveness to accommodation. This condition will resolve with time, ranging from months to years. Some patients may experience increased accommodation for near vision but the pupillary light response does not recover and become persistent after 2 years.

Case Presentation: A 26-year-old female patient showed anisocoric pupil in a response to bright light in the left eye. The size of the pupil is 7 mm with the absence of direct and indirect

light reflexes and near reflexes. She was diagnosed with Adie's tonic pupil. It is found to be persistent in the 3 years of follow up, the involved pupil was showing anisocoria, 5 mm in diameter, poor constriction mechanism in direct, indirect, and near reflexes examinations.

Conclusion: Adie's tonic pupil is a benign neuro-ophthalmology disorder of unknown etiology. This condition mostly will spontaneously resolve. But, at some patients, it could be persistent, which may be related to degree of myelination of neural fibers.

Keywords: Corneal ulcer, dematiaceous fungus, keratectomy

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INTRODUCTION

Adie's tonic pupil is characterized of a tonically dilated pupil, which is unresponsive to bright light followed by moderate responsiveness to accommodation. It is caused by damage to postganglionic fibers of parasympathetic innervation in ciliary ganglion. Most cases of the tonic pupil are idiopathic. Tonic pupil is usually unilateral, but it can rarely be seen in both eyes. Adie's tonic pupil has a prevalence of 2/1000 cases and are reported in young female predominance with mean age of onset of 32.2 years from 20 to 40 years of age. Diagnostic features of tonic pupil are pupillary light-near dissociation, slow re-dilatation after near effort, and denervation supersensitivity to dilute cholinergic drugs.¹⁻³

The Adie's pupil is mostly benign and idiopathic (70%). Accommodative paresis may resolve with time, ranging from months to years. Eventually, an Adie's tonic pupil becomes miotic. However, some patients may experience an increase reflex responding to near accommodation but the pupillary light response does not recover and become persistent after 2 years.⁴⁻⁶

eye clinic upon noticing her left pupil was bigger than right pupil when it was flashed with a flashlight by her friend. She also complained a glare in sunlight and having difficulty when reading. There was no history of any systemic disease, a chronic use of any medication, nor trauma. Her best corrected visual acuity was 6/6 bilateral with spherical and astigmatism lens.

The eye examination revealed that she had anisocoria and her left eye was dilated. Under illumination, we found that her right pupil and left pupil were 3 and 7 mm in diameter, round and regular (**Figure 1**). Direct and indirect reflexes were observed in the right pupil but not in the left. The left pupil was mildly tonically constricted on near vision under ophthalmic instillation of topical dilute pilocarpine (0.1%). In the examination performed in 30 minutes, we found the left pupil constricted to 3 mm, but the right pupil remained unchanged (**Figure 1**). Neither ptosis nor impaired ocular movement was observed.

The laboratory tests were within normal limit and there was no abnormalities in her brain blood vessels examined with magnetic resonance angiography (MRA) (**Figure 2**). Patient was diagnosed with anisocoria pupil caused by Adie's tonic pupil and was given diluted pilocarpine (0.1%) to be administered twice daily.

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CASE REPORT

A 26-year-old female patient visited our outpatient

In a follow-up occurs three years after (Figure 3) the diagnosis was made, under the eye examination we found the pupils were still in anisocoric. Her left pupil was 5 mm in diameter and reacted slowly

in direct and indirect light reflex examination. In addition, it was poorly constricted in the near response.

DISCUSSION

The cause of tonic pupil is usually idiopathic. It is more common in young female, and 70% of patients in were in their third decade of life. It is rarely associated with a number of ocular and systemic of diseases, such as autoimmune hepatitis, migraine, Guillain–Barre syndrome and dysautonomia (as in Harlequin and Ross syndrome) or ischemia. Tonic pupil is unilateral in 80% of cases, although the second pupil may later become involved (4% per year).⁴ Our patient was a 26-year-old female, without any history systemic disease, chronic medication, nor trauma. Her laboratory examination and magnetic resonance angiography (MRA) result were normal.

Unilateral tonic pupil is usually caused by damaged in postganglionic parasympathetic nerve fibers in ciliary ganglion. The symptoms of Adie's tonic pupils are mostly accommodative symptom of photophobia or blurred near vision. But, they just occasionally have no symptom and most patient say that anisocoria was noticed by a friend or a relative. The involved pupil is dilated and have irregular size compared to the other eye. Light reflex is weak or unresponsive. Moreover, the accommodation process is also impaired. Diagnostic features of tonic pupil are based on clinical observation, the absence of an extraocular abnormality and abnormal pilocarpine test. Neuroimaging is considered to eliminate other causes.^{1,2,6}

Our patient first notified by her friend that her pupils were unequal after accidentally flashed. She also complained a glare in sunlight and had difficulty in reading. The complaints arose because of visual accommodation disturbances. Her left pupil was affected, dilated and had regular size compared to the other eye. The eye showed unresponsiveness of the direct and indirect illumination reflexed. In addition, there was a mild constriction after the pupil when the wye was examined for near reflex.

Topical dilute pilocarpine is used as a diagnostic test. One drop is placed in each eye and the pupillary diameter is measured in approximately 30 to 45 minutes. Normal pupils are not expected to constrict when dilute concentrations of pilocarpine are used. The denervated iris sphincter is supersensitive to weak cholinergic or parasympathomimetic solution such as dilute pilocarpine eyedrop (0.1%). About 80% of patients with tonic pupil show cholinergic denervation supersensitivity. Our patient was examined with diluted pilocarpine test 0.1% for



Figure 1. (a) The left pupil was unresponsive to direct and indirect light stimuli. (b) After topical dilute pilocarpine was installed into both eyes, 30 minutes, the left pupil showed marked constriction (Courtesy by Permadi).

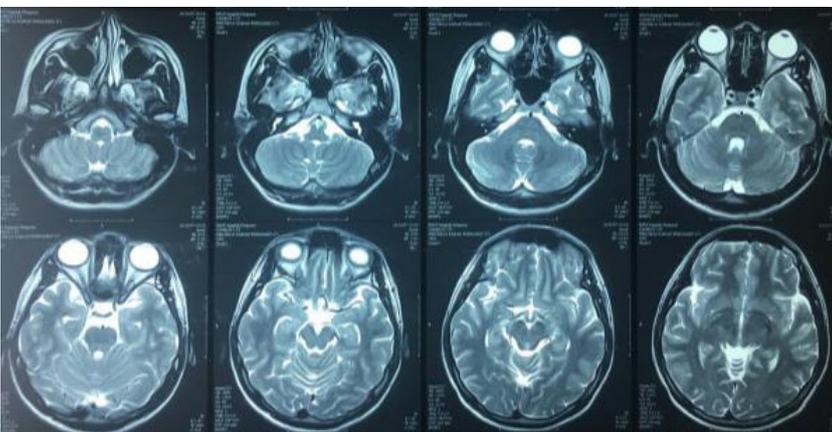


Figure 2. This MRA did not show any pathologies and abnormalities in blood vessels in this patient.



Figure 3. Pupil anisocoria persisted in the follow-up in three years (Courtesy by Permadi).

both eyes and found to have a supersensitivity denervation in the left eye with reduction of its pupil size to 3 mm after 30 minutes. Meanwhile, the normal pupil did not constrict with this dilute dose of pilocarpine.^{3,4}

Adie's tonic pupil is a benign condition. Accommodative paresis may resolve with time, ranging from months to years. Some patients may experience increased near accommodation but the pupillary light response does not recover and become persistent after 2 years. In this case, we found a persistent Adie's tonic pupil. The effected pupil remained unresolved after 3 years of diagnosis. The pupils were still anisocoria. The effected one showed slow reaction to light and poor constriction in accommodation. The isolated postganglionic-fiber involvement may be related to the size and degree of myelination of neural fibers.^{1,4,6}

CONCLUSION

Adie's tonic pupil is a benign condition and the etiology are mostly unknown. The pupil should be examined thoroughly to exclude other differential diagnosis. Adie's tonic pupil becomes persistent if during two years of follow-up no significant improvement for pupillary reaction was found. This condition may be related to the degree of myelination of neural fibers.

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CONFLICT OF INTEREST

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