

Isolated Simultaneous Bilateral Adie's Pupil

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Adie's pupil, an efferent pupillary defect is caused by injury to parasympathetic pupillomotor nerve supply to Iris sphincter muscle. More commonly seen in young adult females with up to 80% cases having unilateral involvement. According to literature, Adie's Pupil may occur alone or as a part of a systemic condition or may be associated with a syndromic presentation. Bilateral Adie's pupil in a female at initial presentation with mild visual symptoms and no syndromic association is sparsely reported in literature.

Keywords: Adie's pupil, Anisocoria, tonic pupil.

Adie's pupil is a common cause of anisocoria. It is an efferent pupil defect, which causes loss of reaction to light of iris sphincter muscle, the reason being the injury to the post-ganglionic parasympathetic nerves. Such patients may be asymptomatic but suffer from accommodative symptoms or photophobia with difficulty in focusing. Signs are anisocoria with light near dissociation and loss of accommodation and abnormal response to light, which may be missed or misdiagnosed in a busy clinical setting. Reports of tonic pupils in literature date back to early 20th century, with half of the patients being females. In later years, Adie, Moore and Holmes reported 46 more cases out of which 42 were females. According to reports, this disease has a predominance of women with 20-40 years old age group but has also been reported in children with a possibility of familial incidence¹. In 20% of the 220 cases from literature, both eyes were involved. The incidence of the condition is reported to be 4-7 per 100,000 per year².

We present a case of a 30 years old female with bilateral Adie's tonic pupils.

A 30 years old female presented with a 3-month history of photophobia and blurring of near vision, both of which had gradually increased over time. The patient had no other ocular or systemic complaints. No significant ocular history was present. Her medical history was insignificant and there was no history of usage of any systemic or topical medication.

On examination her visual acuity was 6/6 for far and N6 for near in both eyes (OU). Although she was N6 OU, she required 15-20 seconds of constant effort at the near target to see the line clearly. The pupils showed anisocoria with a pupillary diameter of 7mm on right side and 8mm on left (**Fig. 1**) with patient focusing on a distant target in a moderately lit room with no response to light stimulus bilaterally. Segmental contractions of iris were not seen in either eye. The patient had light near dissociation on both sides with a pupillary diameter of 4mm on right side and 5mm on left side when looking at near target for 60 seconds, which reverted to previous pupillary size

CASE REPORT



Fig 1: Picture showing bilateral dilated pupils prior to instillation of topical 0.1% Pilocarpine drops.

after 30 seconds of cessation of fixation to near target. In a dimly lit room, after instillation of 0.1% pilocarpine, examination after 30 minutes revealed a pupillary diameter of 5 mm on right side and 4mm on left (**Fig. 2**). Rest of the examination was unremarkable and no evidence of iris trauma or pigment dispersion were found.



Fig 2: Picture showing bilateral constricted pupils 30 minutes after instillation of topical 0.1% Pilocarpine drops.

Detailed neurological examination did not reveal any abnormality. Opinion of the neurologist was sought, radiological and lab investigations were done, including MRI Brain and Visual Pathways with contrast, with none of them revealing any abnormality. Based on history, examination and investigations, a diagnosis of bilateral Adie's pupil was made.

The patient was counselled regarding the condition and was offered prepared topical 0.1% pilocarpine eye drops. The patient perceived resolution of her complaint of photophobia with the usage of the offered eye drops in both eyes once daily and was satisfied with the treatment offered. She was also offered hyperopic correction for near vision, but was not interested in using near glasses.

DISCUSSION

Adie's syndrome is mostly a disease of young adults. Most patients with Adie's pupil have a variable accommodative paralysis at the onset, which is the

main cause of their complaints. In 1812, Ware³ described a 30 to 40-year-old woman with a right pupil showing light near dissociation.

In 1902, Saenger⁴ described a 34-year-old woman with bilateral light-near dissociation, her left pupil was larger than the right and after accommodation, it took around 10 minutes for left pupil to return to its original size. Also in 1902, Strasburger⁵ reported a patient whose both pupils were not reacting to light, however, both pupils reacted to near target but very late, and a delay was noted in relaxing accommodation when refocusing back to a far target. Strasburger also noted other neurological signs and symptoms with an early stage of multiple sclerosis considered the most likely diagnosis. Drouet et al. and Millar et al⁶ reported bilateral Adie's pupil in a patient during an attack of migraine. The authors discussed postganglionic dysfunction as a cause of transient mydriasis. Our patient had no history of migraine and the pupils were not transiently dilated.

Jivraj and Johnson⁷ reported a case of a young girl who had acute unilateral tonic pupil, which became bilateral 2 months later and found a rare association of neurosyphilis with the condition. Our patient reported with simultaneous bilateral involvement and was found to be seronegative for syphilis. An association between Bilateral Adie's pupil with Sjögren's syndrome has been reported in many cases⁸ with high prevalence in females. Our patient was not found to have any associated feature of the syndrome.

Several cases with Bilateral Adie's pupil along with Autonomic dysfunction and diminished or absent tendon reflexes have been reported in literature. Holmes G et al.⁹ described bilateral Adie's pupil in 3 patients, who had signs of autonomic dysfunction (known as Holmes-Adie Syndrome). Our Patient had no evidence of any Autonomic dysfunction and had normal deep tendon reflexes. Presence of bilateral Adie's pupil is a relatively rare initial presentation and in such bilateral cases are commonly reported to present unilaterally and proceed to involve the other pupil after an interval of few weeks to few months. Our case is relatively rare that our patient had simultaneous bilateral Adie's pupil on initial presentation but with mild visual symptoms. On the other hand, majority of the cases with bilateral involvement occur in females and in a younger age group, which is similar to our case. A similar case was reported by Indranil and Shroff¹⁰ but their patient was a male with a longer duration of more severe visual symptoms which is not commonly

reported. Patient was not found to have any other systemic predisposing conditions or associations and was supported and evidenced by relevant radiological and lab investigations. Keeping in mind the differential diagnosis and after proper evaluation, a final diagnosis of bilateral Adie's pupil was made.

REFERENCES

1. **Lambert SR, Yang LL, Stone C.** Tonic pupil associated with congenital neuroblastoma, Hirschsprung disease, and central hypoventilation syndrome. *Am J Ophthalmol* 2000; 130: 238-240.
2. **Martinelli P.** Holmes-Adie syndrome. *Lancet*. 2000; 356: 1760-1761.
3. **Ware J.** Observations relative to the near and the distant sight of different persons. *Philos Trans R Soc London*, 1812: 31-50.
4. **Saenger A.** Ueber myotonische Pupillenbewegung. *Neurol Zentralblatt*. 1902; 21: 837-839.
5. **Strasburger J.** Pupillenträgheit bei Accommodation und Convergence. *Neurol Zentralblatt*. 1902; 21: 738-740.
6. **Millar E, Habib M, Gnanaraj L.** Bilateral tonic pupil secondary to migraine in a child. *J Pediatr Ophthalmol Strabismus*, 2010; 47: e1-2.
7. **Jivraj I, Johnson M.** A rare presentation of neurosyphilis mimicking a unilateral Adie's tonic pupil. *Semin Ophthalmol*. 2014; 29: 189-91. [PubMed: 23952008].
8. **Bachmeyer C, Zuber M, Dupont S, Blance P, Dhote R, Mas JL.** Adie syndrome as the initial sign of primary Sjogren syndrome. *Am J Ophthalmol*. 1997; 123: 691-692.
9. **Holmes G.** Partial iridoplegia associated with symptoms of other diseases of the nervous system. *Trans Ophthalmol Soc UK*, 1931; 51: 209-28.
10. **Saha I, Shroff CM, Gupta C, Verma R.** An interesting case of simultaneous bilateral Adie's tonic pupil. *Oman J Ophthalmol*. 2018; 11 (1): 82-84.

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