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Case Report : a Rare Case of Adie Tonic Pupil in a Female Teenager
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A RARE CASE OF ADIE TONIC PUPIL IN A FEMALE TEENAGER

ABSTRACT

Introduction: Adie tonic pupil of Holmes-Adie Tonic Pupil Syndrome is a rare condition which is commonly found in adult females.
Purpose: to report a case of Adie Tonic Pupil in a female teenager.
Case Report: A 17-year old female was referred to neuro-ophthalmology clinic complaining of glare in right eye, which has been felt in the last 2 years. There was no blurred far or near vision nor was there any history of spectacles. Color and contrast vision was within normal limit. There was anisocoria which was greater in light and segmental paresis on iris sphincter of right eye. Pupil diameter of right eye was 5 mm in light and reduced to 4 mm after instillation of 0.1% pilocarpine. There was decreased direct and consensual pupillary light reflex of right eye without reverse relative afferent pupillary defect (RAPD). Patient underwent magnetic resonance imaging (MRI) and the result was within normal limit. Patient was then referred to neurologist for further evaluation of any reflex deficit.
Conclusion: Adie tonic pupil may appear in a female teenager with the classic symptom of photophobia. Evaluations are required to exclude any underlying disease.

Keywords: adie tonic pupil, female, teenager.

I. INTRODUCTION

Tonic pupil refers to any poor pupillary response to light with segmental paresis of the iris sphincter. Tonic pupil is due to damage of the ciliary ganglion or intraorbital ciliary nerve. According to its cause, tonic pupil is classified into local tonic pupil, neuropathy pupil, and Holmes-Adie pupil syndrome.[1,2]

Adie tonic pupil is tonic pupil disorder with no specific etiology in a healthy person. Adie tonic pupil is commonly found in adult female. Its incidence is at highest in 30 year-old females. This case report presents a rare case of adie tonic pupil in a female teenager.[2,3]

II. CASE PRESENTATION

A 17-year old female patient came to neuro-ophthalmology outpatient clinic complaining of glare in right eye, particularly in bright places, which has been felt in the last 2 years. There was no blurred near or far vision and no history of spectacles. She denied having symptoms of headache and diplopia, no history of eye drops, and no history of any alcohol consumption. She had no history of herpes zoster, chickenpox, diphtheria, pertussis. There was also no history of joint pain or

skin rashes. She had never undergone any prior surgery including ocular or dental surgery. There was no prior trauma or injury to the eye. She had never visited any ophthalmologists before.

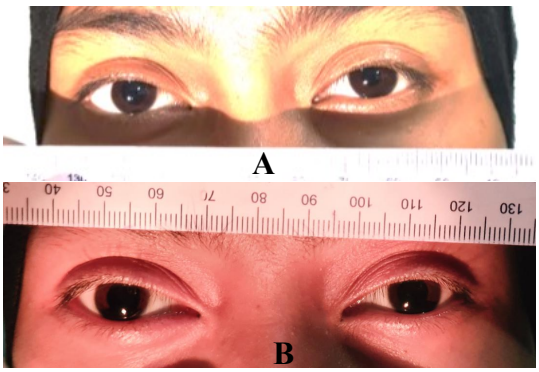

Figure 1. Pupil diameter difference in (A) bright light and (B) dark.

Visual acuity of right eye was 0.8 and left eye was 1.0, measured by Snellen visual acuity chart. Contrast sensitivity, evaluated by MARS chart, showed normal contrast sensitivity. Color vision was evaluated using Ishihara color chart and was within normal limit. Intraocular pressure of both eyes was within normal limit. Ocular motility was within normal limit without any gaze palsy.

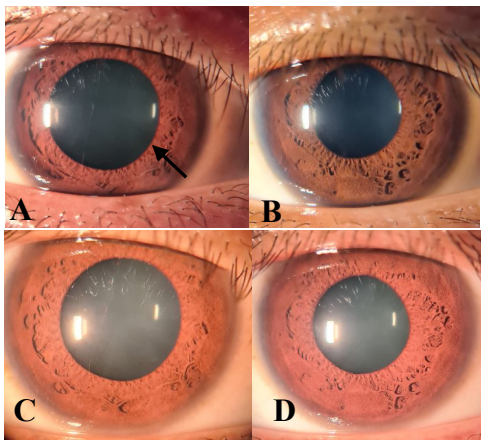

Figure 2. Anisocoria of both eyes, (A) right eye, (B) left eye, segmental paresis is shown in arrow, (C) reduced pupil diameter of right eye after instillation of 0.1% pilocarpine, (D) left eye

There was anisocoria which was greater in light. Pupil diameter of right eye was 5 mm in bright light and 7 mm in dark. Pupil diameter of left eye was 3 mm in bright light and 7 mm in dark as compared in figure 1. Pupil diameter of the right

eye was also reduced during convergence. Slit lamp biomicroscopy also revealed segmental paresis of right iris sphincter in nasal part, as shown in figure 2. There was no signs of infection or inflammation in both eyes, posterior segment examination was within normal limit.

She was given one drop of pilocarpine 0.1% and after 45 minutes, pupil diameter of the right eye became 4 mm in bright light as seen in figure 1. This patient was then diagnosed with Adie Tonic Pupil of right eye and was referred to neurologist to evaluate any reflex deficit.

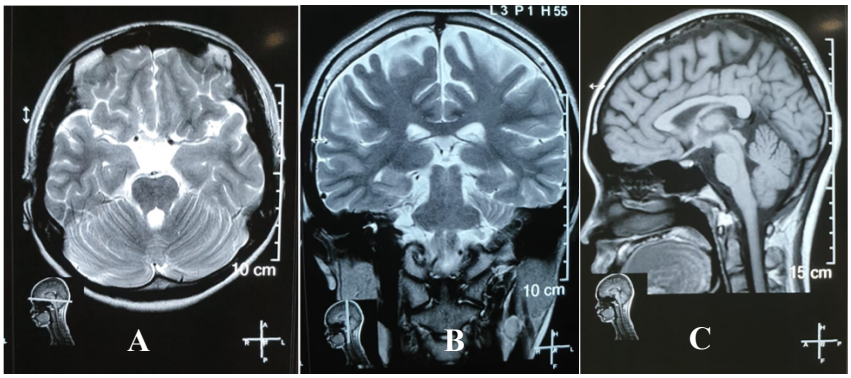

Figure 3. Normal brain MRI result (A) axial plane (B) coronal plane (C) sagittal plane

She underwent brain magnetic resonance imaging (MRI) and the result was within normal limit as shown in figure 3. She was then referred to neurologist for evaluation of any reflex or neurologic disorder. This patient was given explanation about her condition and that there was no specific treatment required for the condition. She was asked to come for follow up visit after visiting neurologist. She was also asked to come to neuro-ophthalmology outpatient clinic regularly, particularly if there is deteriorating condition of the right eye, or when the same symptom is felt on the left eye. The prognosis of this patient was *quo ad vitam dubia ad bonam*, *quo ad functionam ad bonam*, *quo ad sanationam dubia*.

III. DISCUSSION

Anisocoria refers to asymmetry of the pupils between both eyes with diameter difference greater than 0.4 mm. Anisocoria needs to be evaluated further whether

the asymmetry is larger in bright conditions or in dark conditions as the causes may be different. Variable causes of anisocoria are shown in table 1. This patient had more anisocoria in light without any history of trauma and no ocular motility disorders, thus the anisocoria may be caused by tonic pupil syndromes.[1,2]

More Anisocoria in Darkness
Simple (physiologic) anisocoria
Inhibition of the sympathetic pathway • Horner syndrome • Pharmacologic (dapiprazole, thymoxamine)
Stimulation of the sympathetic pathway • Tadpole pupils • Intermittent dilation of one pupil caused by sympathetic hyperactivity • Pharmacologic (cocaine, eye-whitening drops, adrenergic drugs)
Pharmacologic stimulation of the parasympathetic pathway (eserine, organophosphate esters, pilocarpine, methacholine, arecoline)
More Anisocoria in Light
Damage to the parasympathetic outflow to the iris sphincter muscle • Oculomotor nerve paresis • Tonic pupil syndromes (including Adie) • Intermittent dilation of one pupil caused by inhibition of the parasympathetic pathway
Trauma to the iris sphincter
Acute glaucoma siderosis
Pharmacologic inhibition of the parasympathetic pathway (atropine, scopolamine)

Table 1. Causes of Anisocoria
Source: Miller *et al*[1]

Tonic pupil syndromes are pupil which react poorly to light and can be seen as regional iris palsy seen by slit lamp biomicroscopy. Patients with tonic pupil syndromes mostly complain of photophobia and blurry near vision due to poor accommodation. This is caused by lesions which damage the ciliary ganglion or ciliary nerves in the intraorbita.[1,3]

Tonic pupil syndrome, according to Miller *et al*, is further classified into local tonic pupil, neuropathic tonic pupil, and Holmes-Adie Tonic Pupil Syndrome. Local tonic pupil is tonic pupil caused by infection, inflammation, or infiltration which may damage the ciliary ganglion. The causes of local tonic pupil are herpes-zoster infection, chickenpox, measles, syphilis, scarlet fever, sarcoidosis, rheumatoid arthritis, Vogt-Koyanagi Harada Syndrome, viral hepatitis, metastatic or primary intraocular tumors which also have other systemic manifestations. Local

tonic pupil may also be caused by blunt or penetrating trauma to the eye or prior ocular surgery.[1,4,5] Our patient denied any history of trauma to the eye or any systemic conditions related to infections and inflammation, prior to the symptom presentation.

Neuropathic tonic pupil is a tonic pupil condition related to history of chronic alcohol consumption, diabetes mellitus, spinocerebellar ataxias, or Miller Fisher variant of Guillain-Barre Syndrome. In a case report by Venkataraman *et al*, the tonic pupil condition is presented in a patient with multiple sclerosis. Neuropathic tonic pupil is part of a generalized or autonomic neuropathy which will show other systemic manifestations.[1,4] Our patient denied any history of chronic alcohol consumption or any systemic symptoms.

Holmes-Adie Tonic Pupil Syndrome or Adie Syndrome is unilateral or bilateral tonic pupil in healthy person without any systemic conditions. Most patients with Adie Syndrome also have deep tendon reflexes disturbance, which is most likely caused by lesion in central spinal cord. Holmes-Adie Tonic Pupil Syndrome is uncommon, can be familial or sporadic, mostly found in adult female patients between 20-50 years old. Mean age of onset in Adie Syndrome is 32.2 years old while our patient is 17 years old.[1,5,6]

Tonic pupil in Adie Syndrome is caused by damaged postganglionic parasympathetic nerve fibers of the ciliary ganglion. The damaged nerve fibers will cause segmental paresis of iris sphincter, as seen in this patient, and also supersensitivity to parasympathomimetic agent. Our patient was given one drop of diluted pilocarpine eye drop (0.1%) in each eye, and the pupil diameter was remeasured after 30 to 45 minutes. Due to supersensitivity of the damaged postganglionic ciliary nerve fibers, diameter of pupil with Adie Syndrome will reduce after being given a small concentration of parasympathomimetic.[2,7,8] Our patient had long-standing condition and after instillation of parasympathomimetic agent, pupil diameter was reduced by 1 mm.

Adie Tonic Pupil is most commonly unilateral and in 4% of cases will progress to the fellow eye and cause bilateral tonic pupil. In another case report it is mentioned that Adie Tonic Pupil may be an early manifestation of oculomotor

disorders. Our patient had complained of photophobia, as onset of symptom, which had presented for 2 years. Other ophthalmological examination showed there was no disorder of oculomotor nerve. Our patient also underwent MRI and the result showed no lesions or any abnormalities potential to cause oculomotor disturbance. After 2 years, patient also had no complain of any systemic abnormalities. Our patient was then referred to neurologist for evaluation of any neurological reflex disturbance.[1,9,10]

Adie tonic pupil requires no specific medications, unless there is any underlying diseases present. Adie tonic pupil patients need to be reassured about their condition and the eccentricity of this condition. Follow up visit is also required, particularly if there is any deteriorating condition to the same eye or if the same symptom is felt in the fellow eye.[1,10]

IV. CONCLUSION

Adie Tonic Pupil most commonly occur in adult female, but can also be found in a female teenager. Thorough evaluations need to be done to confirm that the tonic pupil is not an initial symptom of any systemic conditions.

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