

Adie Syndrome: A Case Report

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In 1932 Adie¹ described a syndrome in which unilateral accommodation and pupillary contraction were abnormal. It typically was observed in young women with no history or evidence of syphilis and involved a unilateral dilated pupil that was unresponsive to light. He noted that deep tendon reflexes were absent or markedly diminished in these patients. He concluded that it was "a benign disorder *sui generis*." We report a recent case of Adie syndrome.

Case Report

A 37-year-old woman came to the office with right pupil dilatation. It was noticed by a nurse for the first time earlier that morning and had not been present previously according to the patient's report. The patient's only visual complaint was mild photophobia in sunlight. She also had complaints of sinus congestion, headaches, and intermittent mucopurulent rhinorrhea believed to be due to an exacerbation of seasonal allergies. She denied fever, extremity weakness, paresthesias, or a history of sexually transmitted diseases. The patient had a remote history (more than 10 years ago) of an episode of left arm and facial weakness, which resolved.

On a physical examination she had anisocoria with a 5-mm right pupil and a 3-mm left pupil. Pupillary light response was absent on the right and decreased accommodation was present. Extraocular movements were intact bilaterally. Findings on a fundal examination were normal, and her blood pressure was 128/84 mmHg. No other abnormalities were noted. Magnetic resonance imaging (MRI) was ordered and findings were normal except for sinusitis.

Findings on a physical examination the following day were unchanged. Examination of the extremities showed absent ankle or knee reflexes on the ipsilateral (right) side, with decreased (1+) contralateral reflexes. Brachioradialis reflexes

were roughly equal, and the right corneal reflex was decreased. A rapid plasma reagin was obtained and found to be negative. The patient was subsequently referred to an ophthalmologist for confirmation of the diagnosis and to rule out other abnormalities. Confirmation was performed using a dilute solution of pilocarpine.

Discussion

Patients with Adie syndrome have anisocoria characterized by a dilated pupil that is minimally reactive or unresponsive to light. Deep tendon reflexes (knees and ankles) are either diminished or absent.² This disorder is most often seen in young women, who have an average age of onset in their 30s. It usually occurs in one eye, but occurs bilaterally in up to 4 percent of patients.³ Ciliary muscle denervation causes accommodation impairment, while sphincter pupillae denervation causes mydriasis. The resulting anisocoria leads to photophobia in bright light, impaired dark adaptation, and a cosmetic defect.³ Headaches and blurring of vision are also common. It has been suggested that the deep tendon hyporeflexia could be a dysfunction of somatosensory large-diameter afferent fibers at the spinal level. Neural losses of the lumbosacral dorsal root ganglia and dorsal columns have been described.⁴

The differential diagnosis of anisocoria with a dilated pupil includes sphincter muscle damage, third nerve palsy, severe intracranial pathology, unilateral use of a mydriatic, physiologic anisocoria, Horner syndrome, Argyll Robertson pupil, and Adie (tonic) pupil. Adie pupil results from a partial denervation of the postganglionic parasympathetic neurons of the sphincter pupillae muscle and the ciliary muscle.^{3,5} Although most cases are thought to be a result of a viral infection in the ciliary ganglion,⁶ ganglion damage can also result from trauma, tumors, and other infectious agents. Vascular compromise and ischemia from quinine, giant cell arteritis, and reversible ischemia from migraine headaches have also been described.⁶ Other causes include idiopathic disease, orbital trauma or infection, herpes zoster, diabetes mellitus, autonomic neuropathies, and

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Guillain-Barré syndrome.⁷ Adie pupil is thought to represent mild dysfunction of the autonomic nervous system. Other entities can involve more widely spread and troublesome autonomic dysfunction. Ross syndrome involves tonic pupils, altered deep tendon reflexes, and subsequential hypohidrosis.⁸ Acute pandysautonomia and Fisher syndrome are more complicated autonomic illnesses that can also manifest tonic pupils.

The appropriate workup for anisocoria begins with the history. It is vital to ask the patient when the symptoms began, whether trauma was involved, whether drops or ointments were used, and whether there is a history of syphilis or headaches. A tumor, intracranial hemorrhage, or other midbrain abnormality must be ruled out by computed tomography or MRI.⁷ The physical examination first includes a determination of the affected pupil. The affected pupil is usually the larger one in bright light and the smaller one in dim light. Evaluate the pupil margin with a slit lamp for irregularity, to rule out ptosis, and to test ocular motility. Clinical diagnosis can be made by demonstrating the denervation hypersensitivity of the pupil to parasympathomimetic agents such as 0.125 percent pilocarpine.⁹ When such an agent is instilled onto the affected eye, the tonic pupil will noticeably constrict.¹⁰ If the patient is younger than 1 year, a pediatric neurologic consultation is necessary to rule out familial dysautonomia (Riley-Day syndrome).⁸

The condition is benign, and there is no effective treatment. Miotic agents such as pilocarpine 0.125 percent applied three times a day¹¹ have been used to reduce the anisocoria in some patients, but side effects including ciliary muscle spasm have been reported.³ Currently there are no treatment guidelines established in the literature. Within a few weeks of onset, there is often

an improvement in accommodation, which does not always return to normal because of only partial regeneration of the damaged neurons. The affected pupil might become smaller than the normal pupil with time. If the diagnosis is established with certainty, follow-up is routine.⁷

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