Evolution of benign episodic pupillary dilation to Adie's tonic pupil

Benign episodic pupillary dilation (BEPD) may cause significant concern for both patients and clinicians alike. In general, BEPD does not require any work-up or further evaluation beyond simple reassurance. We describe a case of BEPD that evolved into a typical neurologically isolated ipsilateral and idiopathic tonic pupil also known as *Adie's tonic pupil*. After reviewing the PubMed database in search of similar reported cases, we found that this case is a unique presentation in the English-language ophthalmic literature.

A 21-year-old man presented with 9 months of intermittent pupillary dilation OD. The episodes occurred randomly, 3 to 4 times a week, and were accompanied by photophobia and retroorbital pressure OD. He denied any trauma. A head computed tomography scan without contrast material and brain and orbit magnetic resonance imaging with and without contrast material were negative. Past medical history included attention-deficit hyperactivity disorder treated with dextroamphetamine-amphetamine. The patient had idiopathic pericarditis that resolved on serial echocardiograms after treatment with nonsteroidal anti-inflammatory drugs, steroids, colchicine, and supportive therapy. He was taking no topical medications and had no known drug allergies. His family history included Sjögren syndrome and systemic lupus erythematosus. The patient had no residual anisocoria between episodes and was diagnosed with BEPD. No treatment was initiated. He then developed an anisocoria and was referred to the Houston Methodist Hospital.

On neuro-ophthalmologic examination, the patient's uncorrected visual acuity was 20/20 OU. His pupils measured 6 mm OD and 5 mm OS in the dark and 5 mm OD and 3 mm OS in the light. The right pupil was sluggishly reactive to light but had a tonic near reaction (light-near dissociation OD). There was no relative afferent pupillary defect. Motility, external, intraocular pressure, slit-lamp biomicroscopic, and funduscopic examinations were unremarkable. Cranial nerves II–XI were intact. Muscle strength was 5/5 in all extremities. A complete blood count, sedimentation rate, C-reactive protein level, homocysteine level, prothrombin time, activated partial thromboplastin time, and vitamin B_{12} and folate levels were normal, and Lyme titre, interferon gamma release assay, antinuclear antibodies, and syphilis testing were negative.

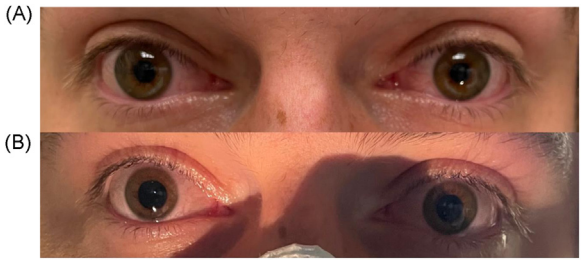

Fig. 1—(A) Pupils measured 5 mm OD and 3 mm OS in the light. (B) Pupils measured 6 mm OD and 5 mm OS in the dark.

Six months later, the pupil was slightly irregular OD with sector paresis, and now bilateral light-near dissociation was noted. Anterior-segment and funduscopic examinations remained unremarkable. Instillation of pilocarpine 0.1% eye drops OU yielded bilateral pupillary constriction consistent with denervation supersensitivity and tonic pupils OU.

The tonic pupil is typically idiopathic (i.e., Adie's tonic pupil) but may occur secondary to trauma, orbital or ocular surgery, infection, or autoimmune disease.[1] The pathophysiology of the tonic pupil is believed to be aberrant regeneration after damage to the ciliary ganglion.

In contrast, the mechanism of BEPD remains ill-defined. Some authors have postulated increased sympathetic or decreased parasympathetic activity in BEPD.[2,3] Our case presented as typical BEPD but evolved to a pharmacologically confirmed bilateral tonic pupil. This suggests that BEPD may be ocular parasympathetic rather than sympathetic in origin.

Several case reports have described episodic unilateral mydriasis with migraine. One study outlined 9 cases of unilateral mydriasis developing during migraine attacks in which the pupillary dilation lasted for 1 or more months and became permanent in 2 patients.[4] While our patient's history of pupillary changes is similar to those described in the above-mentioned study,[4] his case is unique in that his unilateral mydriasis episodes were not associated with migraines.

Clinicians should be aware that BEPD may transition from episodic to constant anisocoria. After a thorough PubMed review, we conclude that this is a unique presentation in the English-language ophthalmic literature. (Fig. 1)

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Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this correspondence.