A 48-year-old Caucasian woman presents with the recent development of blurry vision in her left eye. The patient reports no recent ocular injury and no pain, headache, or other neurological symptoms. She wears corrective lenses but is otherwise healthy, with no exposure to anticholinergic medications. Examination reveals anisocoria: the left pupil is twice as large as the right. There is no ptosis. Both direct and consensual pupillary light reflexes were intact on the right, but they are absent on the left. Monocular vision is also abnormal on the left. Fundoscopy reveals normal retinas bilaterally; however, slit-lamp examination reveals vermiform movements of the left iris. Extra-ocular movements are normal.

**What is your diagnosis?**

a. Hutchinson’s Pupil  
b. Argyll Robertson Pupil  
c. Marcus Gunn Pupil  
d. Adie’s Pupil

**Answer: Adie’s pupil**

Adie’s pupil was first described in two separate papers in 1931. It is also commonly known as a tonic pupil. The etiology of the tonic pupil is not clearly defined; however, the pathophysiology appears to involve damage to the ciliary ganglion with resultant aberrant regeneration of the parasympathetic nerve fibres. It has an incidence estimated at between 4 and 7 cases per 100,000. It occurs sporadically but is found most often in women between the ages of 20 and 40 years. The clinical presentation is one of gradual onset of a unilateral dilated pupil that fails to respond to light and, if early in its presentation, accommodation. With time, the pupil begins to respond to accommodation, owing to the inappropriate reinnervation of fibres from the ciliary ganglion to the sphincter muscle. Patients who also develop absent deep tendon reflexes are said to have Holmes-Adie syndrome.
Although the majority of cases occur in isolation, Adie’s pupil has been associated with a variety of conditions, including rheumatoid arthritis, connective tissue diseases, paraneoplastic syndrome, and peripheral neuropathies. A unilateral dilated pupil can represent a benign condition or be an early sign of significant pathology. A thorough history and physical examination, excluding the presence of other neurological signs and symptoms, is usually all that is required to make the diagnosis. Pupillary constriction in response to local pilocarpine (0.125%) administration can help confirm the diagnosis. Where clinical uncertainty exists, a specialist referral is appropriate. Besides patient reassurance, there is no specific treatment for this condition.

**Resources**

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