Bilateral Tonic Pupils Secondary to Ross Syndrome: A Case Report

Discussion

The iris is innervated by the parasympathetic and sympathetic nervous systems. A defect in the parasympathetic nervous system may result in dilation of the pupil in bright light, while a defect of the sympathetic nervous system may result in miosis of the pupil in the dark. Light-near dissociation is caused by many pathologies, among which are Adie’s and tonic pupils. Many diseases may cause tonic pupils, such as diabetes, herpes, sarcoid, injury, infec-
tion, syphilis, Guillian-Barre syndrome, Shy-Drager tumor, Charcot-Marie-Tooth, and Holmes-Adie syndrome. Argyll Robertson pupils are bilateral small pupils with a rapid con-
striction to near, but with slight or absent constriction to light. Argyll Robertson pupils may have an association with syphilis, so optometrists should order the appropriate lab tests such as the VDRL and FTA-ABS.

Adie’s syndrome, or Holmes-Adie syndrome, refers to cases in which the etiology of the tonic pupils is unknown. Holmes-Adie syndrome is characterized by poor light reaction, paralysis of accommodation, hypersensitivity to cholinergic drugs, pupils that respond to near vision slowly and decreased tendon reflexes. Typically, the syndrome is unilateral and typically occurs in women more than men. Holmes-Adie syndrome tends to progress; the fellow pupil becomes involved and the deep tendon reflexes decrease. The cause of the progressive miosis remains unknown. Patients with Holmes-Adie syndrome may present with blurred near vision as the chief complaint.

Ross syndrome, while benign, is a progressive autonomic dysfunction, which can occur in patients of any age, ethnic background, or gender. Of the twenty patients with Ross syndrome described in the literature, slightly more were male than female. The typical age at the time of diagnosis of Ross syndrome patients was 36 years. The tonic pupil was bilateral in a majority of cases.

Ross syndrome is characterized by tonic pupils, decrease or loss of deep tendon reflexes, and segmental loss of sweating. Tonic pupils show progressive miosis; this miosis progresses more quickly than the normal miosis of aging. Damage to the sympathetic ganglion cells or the postganglionic projections could account for the loss of sweating. The loss of deep tendon reflexes may stem from the damage in the dorsal root ganglia or spinal interneur-

n loss. The mechanism of injury is unknown. The peripheral autonomic nervous system and dorsal root gan-
glia all develop from neural crest cells. One author suggests...
that some commonality makes these tissues prone to injury.⁹

Patients with parasympathetic and sympathetic dysfunction may have systemic symptoms in addition to the ocular symptoms. Patients with Ross syndrome or Holmes-Adie syndrome may present with the following: orthostatic hypotension, headache, psychiatric disorders, reduced heart rate responses to Valsalva maneuvers, diarrhea.⁹ Ross syndrome overlaps with Holmes-Adie syndrome and may represent different manifestations of the same disorder.⁶,⁹ However, one author has suggested that Ross syndrome can be differentiated from Holmes-Adie syndrome by the loss of sweating, since both conditions present with tonic pupils and decreased tendon reflexes.⁹

Conclusion

Tonic pupils may indicate complex systemic disease.¹⁰ Optometrists should order appropriate lab tests to rule out systemic disease in cases of tonic pupils, although Holmes-Adie syndrome is idiopathic. Holmes-Adie syndrome is characterized by tonic pupils, paralysis of accommodation, and decreased deep tendon reflexes.

Ross syndrome is a rare, but progressive autonomic disease that causes tonic pupils, loss of deep tendon reflexes, and anhidrosis. Ross syndrome can be differentiated from Holmes-Adie syndrome by the lack of anhidrosis in Holmes-Adie syndrome. Patients with Ross syndrome have autonomic dysfunction and may show systemic symptoms such as headache, psychiatric disorders, and abnormal heart rate responses.⁹ Patients have difficulty regulating body temperature and should avoid very hot or cold conditions.⁷ Optometrists need to be aware of Ross syndrome and refer to other practitioners as necessary.

References