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SCIENTIFIC LETTER

Bilateral Tonic Pupils Secondary to Ross Syndrome: A Case Report

Pupilas tónicas bilaterales secundarias al síndrome de Ross: informe de un caso

Introduction

In 1958, Alexander Ross described a case with anhidrosis and Adie's syndrome. He considered the Adie's syndrome unrelated to the anhidrosis.[1] Currently, Ross syndrome is described as tonic pupils, decreased or a loss of deep tendon reflexes, and segmental anhidrosis.[2] Ross syndrome is rare, with less than 50 cases existing in the literature.[2]

Case Report

A 44 year-old white male presented to the eye clinic with complaints of difficulty with night driving. His medical history was significant for the following medications: vardenafil, fluoxetine, simvastatin/ezetimibe, tramadol, fish oil, glucosamine, and zolpidem and he had an allergy to percocet. In 1993, he had a history of tick bites from which he developed Rocky Mountain spotted fever (rickettsiosis). After treatment with an antibiotic and recovering from this illness, he noticed an absence of sweating on half of his body. The patient had extensive testing, such as VDRL and FTA-ABS, which was negative. He demonstrated hypersensitivity to dilute pilocarpine and decreased tendon reflexes and was diagnosed with Ross syndrome.

His distance visual acuity without correction was 20/25 in the OD and 20/20 in the OS. Pupils were unreactive to light, with a size of 3–4 mm in bright and dark illumination. Because of the small pupil size, the near reaction was difficult to elicit. The left pupil showed vermiform movements superiorly. The pupils had an irregular shape. Slit lamp exam revealed a normal anterior segment. The field was full by confrontation. Tonometry was measured as 14 in the OD and 14 in the OS. Dilated fundus exam revealed a normal posterior segment, with a cup-to-disk ratio of 0.6 in the OD and OS. He had a loss of sweating on his right side and a loss of deep tendon reflexes.

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.[3] Light-near dissociation is caused by many pathologies, among which are Argyll Robertson and tonic pupils. Many diseases may cause tonic pupils, such as diabetes, herpes, sarcoid, injury, infection, syphilis, Guillan-Barre syndrome, Shy-Drager, tumor, Charcot-Marie-Tooth, and Holmes-Adie syndrome.[2] Argyll Robertson pupils are bilateral small pupils with a rapid constriction to near, but with slight or absent constriction to light.[4] 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.[6] Typically, the syndrome is unilateral and typically occurs in women more than men.[3] Holmes-Adie syndrome tends to progress; the fellow pupil becomes involved and the deep tendon reflexes decrease.[5] The cause of the progressive miosis remains unknown.[7] Patients with Holmes-Adie syndrome may present with blurred near vision as the chief complaint.[5]

Ross syndrome, while benign, is a progressive autonomic dysfunction, which can occur in patients of any age, ethnic background, or gender.[6] Of the twenty patients with Ross syndrome described in the literature, slightly more were male than female.[8] The typical age at the time of diagnosis of Ross syndrome patients was 36 years.[8] 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.[6] The loss of deep tendon reflexes may stem from the damage in the dorsal root ganglia or spinal interneuron loss.[6,7] The mechanism of injury is unknown.[6,9] The peripheral autonomic nervous system and dorsal root ganglia all develop from neural crest cells.[9] One author suggests

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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.[7]

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.

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