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Adie-Holmes Syndrome

Síndrome de Adie-Holmes

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Abstract

The Holmes-Adie syndrome is characterized by the presence of tonic pupil associated with absence or diminution of deep tendon reflexes. In some cases there may be autonomous nerve dysfunction. The mechanism that causes the disorder is not fully known, but is believed to be caused by denervation of the postganglionic supply to the sphincter of the pupil and the ciliarymuscle which can occur following viral disease. Typically it affects young adults and is unilateral in 80% of cases, although it may develop in the contralateral eye in months or years. We report a case of a woman presenting typical signs of this syndrome, in which pharmacological test was essential for diagnosis.

Keywords: *Adie syndrome/diagnosis; Pupil disordes/diagnosis; Physical examination; Case reports*

Resumo

A Síndrome de Holmes-Adie É caracterizada pela presença de pupila tônica associada à diminuição ou ausência dos reflexos tendíneos profundos. Em alguns casos pode haver disfunção nervosa autônoma. O mecanismo que causa a desordem não é totalmente conhecido, mas acredita-se que seja causada pela desnervação do suprimento pós-ganglionar para o esfíncter da pupila e para o músculo ciliar, que pode ocorrer após doença viral. Tipicamente afeta adultos jovens e é unilateral em 80% dos casos, embora possa se desenvolver no olho contralateral em meses ou anos. Nós relatamos o caso de uma mulher apresentando sinais típicos desta síndrome, em que o teste farmacológico foi fundamental para o diagnóstico.

Descritores: Síndrome de Adie/diagnóstico; Pupila tônica; Exame Físico; Relatos de casos

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Introduction

The Holmes-Adie syndrome was described simultaneously in 1931 by Gordon Morgan Holmes and William John Adie. It is characterized by the presence of tonic pupil associated with absence or diminution of deep tendon reflexes. In some cases there may be autonomous nerve dysfunction.[1]

The mechanism that causes the disorder is not fully known, but is believed to be caused by denervation of the postganglionic supply to the sphincter of the pupil and the ciliarymuscle which can occur following viral disease. Typically it affects young adults and is unilateral in 80% of cases, although it may develop in the contralateral eye in months or years.[2, 3]

Case Report

Female, 43 years old, white, searched an ophthalmologist complaining her left eye pupil was "bigger" than the right eye one for over a month. No other ophthalmological complaints. She reported associated vertigo. She searched for neurological aid and a MRI of the brain was performed without changes. She went to the otorhinolaryngologist, where she was diagnosed with Meniere's Syndrome, and she began treatment with Labirin® (betahistine di hydrochloride). She abandoned treatment after developing headache as a side effect of the drug. Personal background reports thalassemia minor, flu shot one year ago and infection treatment of H. pylori for 6 months. She is a folic acid user.

Ophthalmologic exam showed visual acuity of 20/15 in the right eye (RE) and 20/20 in the left eye (LE) without correction. Biomicroscopy shows significant anisocoria, left greater than right. Pupillary diameter measured by iTrace Ray Tracing Corneal Topography and WavefrontAberrometer (Tracey Technologies) is shown in Table 1 and Figure 1.

Fundus biomicroscopy was unchanged. Direct pupillary reflex present in the right eye and slow in the left eye. Slow consensual reflex in both eyes, with no relative afferent defect in either eye.(Figures 2 and 3).

The pupillary near reflex was slow in the left eye, with slow dilation. (Figure 4)

The patient also had absent tendinous reflex in the right and normal in the left.

Table 1

Pupillary diameter measurement in both photopic and mesopic conditions

	Photopic		Mesopic	
	Prior to pilocarpine	Post pilocarpine 0.125%	Prior to pilocarpine	Post pilocarpine 0.125%
RE	3.37mm	3.12mm	3.86mm	3.30mm
LE	5.34mm	4.07mm	5.47mm	4.36mm

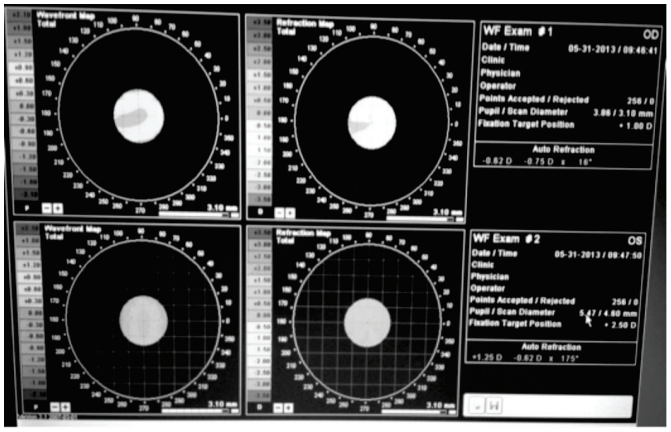

Figure 1: iTrace in physiological mesopic conditions

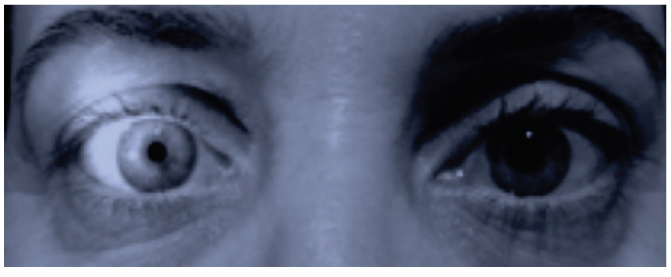

Figure 2: Normal pupillary light reflex of the right eye

Figure 3: Slow pupillary light reflex of the left eye

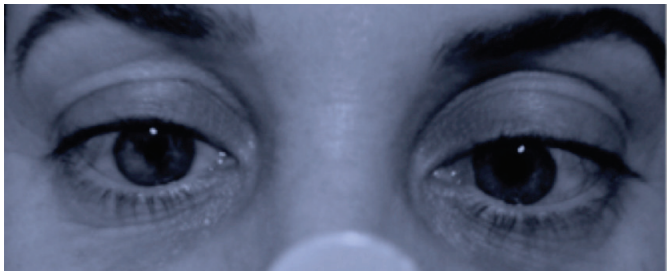

Figure 4: Pupillary near reflex

Discussion

The tonic pupil is usually noticed by others and not by the patient himself. The difference in pupil size (anisocoria) can be found in a variety of ophthalmic disorders, which may

be benign or not, such as physiological anisocoria, Horner's syndrome, Adie's pupil, pharmacological anisocoria, pupillary supranuclear disorders, problems of the iris, systemic diseases or paralysis of the third cranial nerve.[4]

In our case, after clinical and complementary orbit USG and cranial MRI, which was normal, was done testing the instillation of pilocarpine 0.125%[5], which was positive for the left eye. We believe that drug tests, such as that performed above, assist in laboratory diagnosis of tonic pupil syndrome.

References

1. Martinelli P. Holmes–Adie syndrome. Lancet. 2000; 356(9243):1760–1.
2. Ulrich J. Morphological basis of Adie's syndrome. Eur Neurol. 1980; 19(6):390–5.
3. Mak W, Cheung RT. The Holmes-Adie plus syndrome. J ClinNeurosci. 2000; 7(5):452
4. Wilhelm H. Neuro-ophthalmology of pupillary function—practical guidelines. J Neurol. 1998;245(9):573-83.
5. Antonio-Santos AA, Santo RN, Eggenberger ER. Pharmacological testing of anisocoria. Expert OpinPharmacother. 2005; 6(12):2007-13.

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