USEFULNESS OF APRACLONIDINE IN THE DIAGNOSIS OF HORNER SYNDROME

EL USO DE APRACLONIDINA EN EL DIAGNÓSTICO DEL SÍNDROME DE HORNER

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ABSTRACT

Case reports: We present four cases: two males with Horner Syndrome, who were diagnosed by means of apraclonidine 0.5% and cocaine 4% tests respectively. Two children with anysocoria, one of whom had Horner Syndrome confirmed with apraclonidine 1% and the other, in whom an apraclonidine test ruled out this syndrome but in whom pilocarpine 0.125% confirmed Adie's pupil.
Discussion: Apraclonidine drops (1% and 0.5%) may aid in the diagnosis of Horner Syndrome and are easier to obtain than cocaine *(Arch Soc Esp Oftalmol 2009; 84: 105-108).*

Key words: Apraclonidine, Horner Syndrome, anysocoria, cocaine, diagnosis.

RESUMEN

Casos clínicos: Se presentan cuatro casos: dos varones adultos con Síndrome de Horner clínico, diagnosticados mediante el test de apraclonidina 0,5% y el test de cocaína 4% respectivamente, y dos niños con anisocoria de los cuales, en uno de ellos el Síndrome de Horner se confirma con apraclonidina 1% y en otro el test de apraclonidina 1% descarta el Síndrome de Horner y el test de pilocarpina 0,125% confirma una pupila tónica de Adie.
Discusión: La apraclonidina al 1% y al 0,5% pueden orientar el diagnóstico de Síndrome de Horner, siendo más fáciles de obtener que la cocaína.

Palabras clave: Apraclonidina, Síndrome de Horner, anisocoria, cocaína, diagnóstico.

INTRODUCTION

Lesions in the anatomy of the sympathetic innervation are responsible for the so-called Horner Syndrome. Depending on their location, they may be central, preganglion or postganglion.

Clinical manifestations include mild ptosis; lower eyelid elevation; miosis accentuated in the dark; decreased ipsilateral sweating (if preganglion); and iris hypochromic heterochromia when dealing with congenital or long duration syndrome.

For years, the tests performed to confirm diagnosis included the cocaine test and the hydroxyamphetamine test. Cocaine causes dilation in normal eyes though not in the affected eye due to the lack of noradrenaline in nerve endings. Cocaine inhibits the reuptake of noradrenaline in nerve endings, thus increasing its effect in normal conditions.

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Hydroxyamphetamine causes dilation in both eyes in preganglion lesions though not in those eyes affected by postganglion lesions, therefore limiting the location of lesions. This may be explained by the fact that hydroxyamphetamine boosts the release of noradrenaline in postganglion endings.

In the past, diluted adrenaline and fenilefrine were also used since hypersensitivity caused by denervation associated to Horner Syndrome results in marked mydriasis, not as widely reported in normal eyes (1,2).

More recently, apraclonidine, an a2-agonist commercially available as ocular hypotensor, has been found to induce dilation in eyes affected by Horner Syndrome and have a minimal impact on normal eyes, giving way to a «reversal» of anisochoria in these patients, and providing a considerably reliable confirmation of diagnosis (3).

This seems to be the consequence of its action on a1-receptors, which is greater on affected eyes than on normal eyes due to denervation (2).

CASE REPORTS

Adults

Case 1

Thirty-one year old male who had undergone surgery of the left apical sympathetic schwannoma after repetitive episodes of pneumothorax.

Ophthalmic exploration after surgery was compatible with left-sided Horner Syndrome. The test was performed using apraclonidine 0.5% eye drops, observing 30 minutes later reversal of anisochoria with slight dilation of the left eye, reduced ptosis and decreased elevation of the left lower eyelid.

Case 2

Forty-six year old male, smoker and former drinker arrived in the ER reporting slight right-sided periocular pain, 2-month evolution ptosis and miosis. Systematic neurological exploration yielded normal results and imaging tests (chest x-rays, cervical-thoracic CAT scan, cranial MNR and angio cerebral NMR) were normal.

He did not report facial anhydrosis. Ophthalmic exploration was compatible with right-sided Horner Syndrome, the rest being normal (fig. 1).

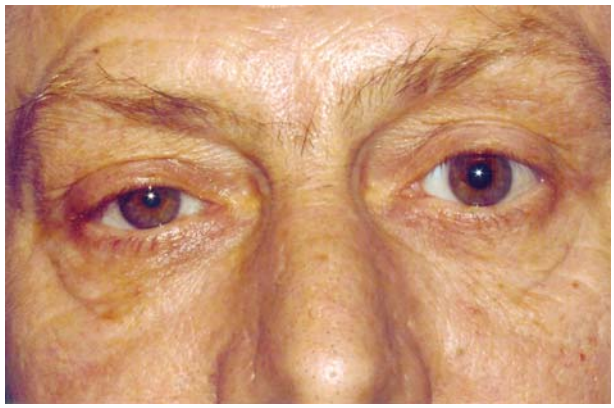

Fig. 1: Right-side Horner Syndrome.

The test performed using phenylephrine eye drops was positive.

Another appointment was scheduled for the 4-percent cocaine test, observing persistent miosis in the right eye and mydriasis in the left eye, thus confirming the diagnosis of Horner Syndrome of unknown etiology (fig. 2).

The hydroxyamphetamine test was not performed in order to «locate» the lesion, as eye drops were not available.

Children

Case 1

Eleven-month-old girl referred for anisochoria with a more pronounced mydriasis on the left pupil.

No relevant background. Weight at birth: 2,800 grams.

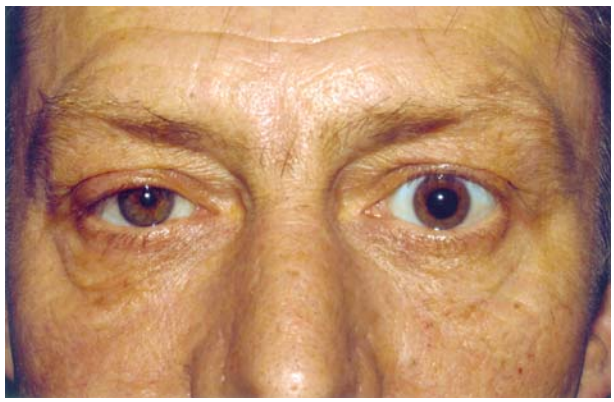

Fig. 2: After administering the 4-percent cocaine eye drops, the non-affected eye dilates and the right eye remains miotic.

Exploration revealed mild intermittent sudden-onset exotropia in the right eye. No ptosis. The fundus picture was characterized by somewhat pale papillae. Cranial CAT scan was normal.

The apraclonidine 1% test was performed, and 30 minutes later no changes were observed in the pupils, thus discarding a right-sided Horner Syndrome.

Another appointment was scheduled to perform the test with pilocarpine 0.125%, which induces miosis in the left eye with the second instillation, thus diagnosing a miotonic pupil.

Case 2

Four-month-old boy referred for ptosis and miosis in the left eye. Background: Caesarean delivery (pelvic presentation.) Born with deviation of labial commisure (potential alteration of the facial nerve at birth).

Ophthalmic exploration compatible with left-sided Horner Syndrome: ptosis, miosis, iris hypochromy and anisocoria greater in dark (fig. 3).

The apraclonidine 1% test was performed; 30 minutes later, anisochoria reverted (fig. 4). The Horner Syndrome diagnosis is confirmed, caused by potential alteration of the brachial plexus at birth.

To date, neurological controls were normal.

DISCUSSION

Apraclonidine 1% and 0.5% may be used at first to guide diagnosis for possible cases of Horner Syndrome (3) both in adults and children (4). It is widely available and no significant side effects have been reported to date. This entails a considerable advantage with respect to the cocaine test due to the wide availability of apraclonidine and to the fact that such tests have positive effects on the affected eye.

Fig. 3: Long-term evolution of left-sided Horner Syndrome: Ptosis, miosis and iris hypochromy.

Fig. 4: After administering apraclonidine 1%, anisochoria reverts and ptosis decreases.

In the present cases, both concentrations (0.5% and 1%) were used indiscriminately since patients had been examined in several departments and this form was available at the time. If both concentrations actually lead to reversal of anisochoria with identical effectiveness, apraclonidine 0.5% should be the form of choice, as it is associated with fewer side effects, especially in the case of children (4).

It is highly unlikely that instillations cause side effects. At any rate, the most common side effects may be: local hyperemia and burning sensation, dry mouth, and more rarely drowsiness, cephalea, dizziness, alterations of cardiac rhythm and digestive function. No adverse reactions were observed in either case.

Very recent lesions of the sympathetic pathway could lead to false negatives due to lack of hypersensitivity. In such cases, the use of conventional tests is advised (5.)

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