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Idiopathic congenital Horner Syndrome. Presentation of a case

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Horner Syndrome results from an interruption of the sympathetic innervation of the eye. This pathway is a chain of three neurons which originate in the hypothalamus, travels down to spinal cord at the level of lower cervical and upper thoracic levels, then traverses the upper chest cavity traveling with the carotid artery, traverse the orbit to innervate pupillary sphincter and accessory muscles for eyelid retraction. The classic clinical triad is unilateral ptosis, miosis and anhidrosis.
There are many causes of Horner Syndrome which can be congenital o acquired. During the first year of life is most often idiopathic in 70%, but the others can be related to neuroblastoma, that's why it is so important to recognize the cause of the syndrome in each patient. In this paper we describe a case report of congenital Horner syndrome, how the diagnosis was made, identification of the causing injury and differential diagnosis.

Key words:
Horner syndrome, congenital, idiopathic, pediatric

Introduction. Horner´s syndrome is a rare condition clinically presented as ptosis, facial anhidrosis and miosis, it results from the disruption of the oculosymphatetic pathway of the innervation to the eye through three orders of neurons, the first one located in the hypothalamus, preganglionic in the thoracic spinal cord and postganglionic in the superior cervical ganglion. Most references classify Horner´s syndrome as congenital or acquired. [1].

Causes of congenital Horner´s Syndrome are anomalies of the carotid artery, birth trauma, tumors and idiopathic occurrence. Acquired Horner´s Syndrome in children is more frequently related to trauma or postoperatively in cervical or thoracic surgery, but there is a percentage of patients of with new onset syndrome in who it can be related to neuroblastoma which is the most commonly associated malignant tumor. [2, 3].

The diagnosis of Horner Syndrome is based on clinical examination, but sometimes the diagnosis can be uncertain, so the pharmacologic testing is indicated, the most used agent is apraclonidine eye drops the test is positive if the pupil dilates as consequence of its action on alfa 1 receptors, which are greater in affected eyes than in normal eyes due to the denervation suffered. [4].

Case presentation

Male patient of 4 months of age, who is the product of normal evolutionary pregnancy, born at term by vaginal route delivery without forceps and with no other previous medical history of interest was brought by her mother to the hospital due to ptosis of the left eye (Figure 1). At the physical examination, right anhidrosis, moderate ptosis of the left eye and central and stable visual acuity were observed;, at the biomicroscope there was heterochromia; brown right eye and blue left eye with miosis, at lighting it was observed that right eye dilated and left eye did not dilated, the time of adaptation in right eye was fifteen minutes while in left eye dilation was not observed and it was difficult to compare because of the age of the patient.

Figure 1

Craneal nerve exploration:

- Third cranial nerve: miosis in the left eye, incomplete ptosis of the left superior eyelid. Adduction, supraduction, infraduction and external rotation ocular movements were normal in both eyes.
- Fourth cranial nerve: internal rotation of both eyes was normal which means innervation of superior oblique was intact.
- Sixth cranial nerve: abduction movements of both eyes were normal.

The clinical diagnosis was Horner Syndrome; to corroborate phenylephrine, eye drops were applied which normally leads to dilation of the pupil, since it blocks the reuptake of norepinephrine, in this patient the test was positive (the ptosis was corrected and pupil did not dilate) for the lack of norepinephrine in this syndrome (Figure 2).

Days later oxamphetamine test was made to locate the cause of miosis, after the application the pupil remained constricted. If the third neuron is intact, amphetamine

Figure 2

causes to empty the noradrenaline vesicles, which leads to persistent mydriasis, so it can be deduced that third neuron was damaged. [5].

The patient was evaluated by a pediatric neurologist who took a Computed Tomography of the brain which is negative for neuroblastoma or other alterations.

Discussion

Differential diagnosis of unilateral ptosis and miosis can have other etiologies such as oculomotor nerve injury in which ocular movements are damaged, pharmacological pupillary blockage, Argill Robertson pupils in which both pupils have abolition of the photomotor reflex in different grades and is usually present in patients with neurosyphilis or systemic lupus erythematosus.

Conclusion

Idiopathic congenital Horner Syndrome is an exclusion diagnosis. It is necessary to do an exhaustive clinical history with emphasis in previous surgery especially thoracic and obstetric trauma and physical examination in patients with Horner Syndrome. In case there is not a clear etiology, we need to exclude space-occupating lesions in children specially in brain because 1 in 10 with neuroblastoma debut with this syndrome.

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Disclosures

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