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A. García-Molina[a,b,c,*], J. García-Férnandez[a,b,c], C. Aparicio-López[a,b,c], T. Roig-Rovira[a,b,c]

[a] *Institut Guttmann, Institut Universitari de Neurorehabilitació adscrit a la UAB, Badalona, Barcelona, Spain*

[b] *Universitat Autònoma de Barcelona, Bellaterra, Cerdanyola del Vallès, Barcelona, Spain*

[c] *Fundació Institut d'Investigació en Ciències de la Salut Germans Trias i Pujol, Badalona, Barcelona, Spain*

* Corresponding author.
E-mail address: agarciam@guttmann.com (A. García-Molina).

Preganglionic or postganglionic efferent pupillary defect? Clinical versus neuroimaging diagnosis☆

Defecto pupilar eferente, ¿preganglionar o posganglionar? Diagnóstico clínico vs neuroimagen

Dear Editor:

Tonic pupil syndrome is a disorder secondary to lesion to the parasympathetic pathway at the ganglionic or postganglionic level.[1] Manifestations include mydriasis with abolished or minimal pupillary reflex (with vermiform movements of the iris due to contraction of normally innervated segments of the iris sphincter, only visible with a slit lamp). Constriction with near effort is present (light-near dissociation) and results from subsequent aberrant reinnervation of the iris sphincter by fibres which were originally destined for the ciliary muscle. This reaction is tonic, that is, slow and sustained. Furthermore, the syndrome also manifests with cholinergic supersensitivity due to postganglionic denervation.[2]

The origin of the injury may be local (viral ciliary ganglionitis, open or closed trauma, or orbital tumours), or systemic-neuropathic (syphilis, alcohol abuse, diabetes, amyloidosis, or paraneoplastic alterations). However, its most frequent variant is idiopathic tonic pupil or Adie syndrome,[1] which predominantly affects women (70%) in the third to the fifth decades of life. It is caused by a painless degeneration of the ciliary ganglion and posterior funiculi, sometimes associated with a slow viral infection.[3]

We present the case of a 44-year-old woman with flash blindness and a 7-year history of anisocoria. She reported having suffered head trauma 20 years previously and migraine-like headache that was being treated with flunarizine. The brain magnetic resonance imaging (MRI) ordered by her neurologist showed a kink at the ostium of the right posterior cerebral artery, originating from the carotid system. Since the kink was touching the superior branch of the ipsilateral oculomotor nerve (OMN), doctors suspected compression of the parasympathetic fibres and referred the patient to our department. We examined her and observed anisocoria due to right mydriasis that was more pronounced under photopic conditions. Direct photomotor reflex and consensual response were abolished in the right eye (RE) but remained intact in the left eye (LE). The near response was normal with a tonic reaction. Extrinsic eye movements were normal with no pupillary response to isolated RE adduction. Biomicroscopy results showed an irregular right pupil with minimal sectoral movements in response to light. Instillation of 0.125% pilocarpine into the conjunctival sac provoked miosis in the RE and no response in the LE (Fig. 1). Bilateral osteotendinous hyporeflexia was also observed. In view of these findings, we proposed the diagnosis of Adie syndrome. Neurologists requested a new gadolinium contrast MRI with thinner slices which revealed displacement of the right OMN without compression (Fig. 2). We therefore opted for watchful waiting.

Pupil abnormalities are examined relatively frequently in neuro-ophthalmology clinics. Finding the lesion location and cause is necessary to assign an accurate diagnosis and subsequently, an appropriate treatment approach. Non-reactive mydriasis may be caused by lesion to the parasympathetic pathway, which can be located at one of several levels: nuclear, preganglionic, ganglionic, and postganglionic.[1]

In this case, differential diagnosis includes lesion to the OMN (preganglionic) and Adie's tonic pupil (ganglionic/postganglionic). Furthermore, differential diagnosis was complicated in this case by results of an imaging scan showing an anatomical variant of the origin of the posterior cerebral artery which might compress parasympathetic fibres of the right OMN. A compression injury to this nerve at the point where it leaves the midbrain may affect parasympathetic fibres, causing mydriasis on the affected side due to a preganglionic defect.[3]

However, clinical signs do not indicate compression injury to the OMN at that level. Light-near dissociation, tonic near-response, and vermiform movements of the iris may indicate tonic pupil. Although the supersensitivity test with pilocarpine has traditionally been used to diagnose tonic pupil, positive results are not exclusive to

☆ Please cite this article as: Arciniegas-Perasso CA, Díaz-Cespedes RA, Manfreda-Domínguez L, Toro-Giraldo D. Defecto pupilar eferente, ¿preganglionar o posganglionar? Diagnóstico clínico vs neuroimagen. Neurología. 2016;31:66—68.

Figure 1 Clinical examination: light-near dissociation. a) Anisocoria due to mydriatic right eye. b) Intact near-response (tonic response in the RE). c) Response to instillation of 0.125% pilocarpine (RE, mydriasis caused by cholinergic denervation supersensitivity; LE, intact). d) No miosis with RE adduction (aberrant reinnervation of the OMN is ruled out).

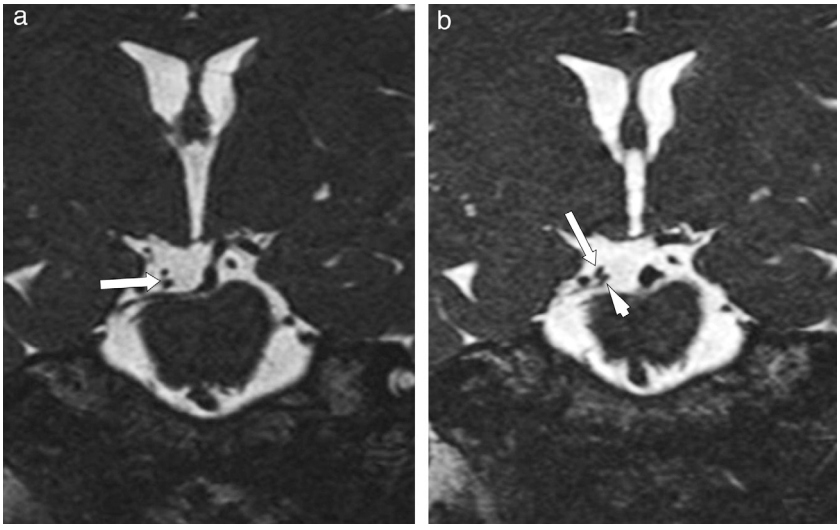

Figure 2 Brain MRI. a) Zoomed view of coronal slice: we observe oculomotor nerve (OMN) in contact with the right posterior cerebral artery (PCA). b) Zoomed coronal slice: OMN (arrowhead), PCA (arrow).

postganglionic parasympathetic lesions but may also be seen with preganglionic lesions.[4,5]

Aberrant reinnervation of the pupil by parasympathetic fibres originally destined for the ciliary muscle depends on the axonal distribution of the ciliary muscle and pupillary sphincter (typically a 30:1 ratio). An abnormal distribution causes light-near dissociation. A similar phenomenon occurs with compressive lesions to the OMN with aberrant reinnervation of the pupillary sphincter by fibres originally destined for the medial rectus muscle. This produces a false light-near dissociation because the pupil responds not only to near objects but also to any eye adduction movement.[1]

Clinical findings associated with bilateral osteotendinous hyporeflexia suggest Holmes-Adie syndrome as a diagnosis for our young female patient, after reconsidering the initial diagnosis suggested by MRI findings. Adopting a multidisciplinary approach to these patients helps determine the diagnosis and an appropriate treatment strategy.

Conflicts of interest

The authors have no commercial interests nor have they received any financial support.

The authors declare that this article has not been published or submitted to another journal for publication. They also confirm transfer of the copyright of this article to the Spanish Society of Neurology.

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C.A. Arciniegas-Perasso*, R.A. Díaz-Cespedes, L. Manfreda-Domínguez, D. Toro-Giraldo

Servicio de Oftalmología, Sección de Neuro-Oftalmología, Hospital Clínico Universitario de Valencia, Valencia, Spain

* Corresponding author.
E-mail address: drcalarpe@gmail.com (C.A. Arciniegas-Perasso).

Asymptomatic colpocephaly and partial agenesis of corpus callosum☆

Colpocefalia y agenesia parcial de cuerpo calloso asintomáticos

Dear Editor:

Colpocephaly (CC) can be observed in radiological findings from brain computed tomography (BCT) and magnetic resonance imaging (MRI). These images show dilated lateral ventricles, specifically in the occipital and temporal horns, and the third ventricle. Frontal horns retain their normal size. This type of hydrocephalus of the posterior half of the brain is a congenital malformation. It can be associated with myelocele, microgyria, cerebellar atrophy, total or partial absence of the corpus callosum, and other structural anomalies. Neurological manifestations occur mainly in children, and include cognitive and motor developmental delay, and epileptic seizures. Diagnosis in adult patients who have learned to perform activities of daily living normally is very infrequent. Imaging studies will show malformations, and differential diagnosis is required to rule out obstructive and non-obstructive ventriculomegaly.[1—5] Case 1 is a 67-year-old man who presented sudden loss of consciousness with no seizures or loss of sphincter control. He regained consciousness, cognitive functions, and motor functions after 1 or 2 minutes. According to his personal history, his developmental progression was normal and he had been working as administrative clerk until his retirement. Ten years before the event, he was diagnosed with arterial hypertension and had since been treated with enalapril dosed at 5 mg twice a day. Clinical, cardiological, and neurological examinations did not show any abnormalities 2 hours after the event, and neither did the electrocardiogram and blood test. The BCT performed in the emergency department revealed very pronounced posterior ventricular dilatation. The patient was then admitted for subsequent studies. A head MRI revealed CC and partial agenesis of the corpus callosum. The electroencephalogram did not show any abnormalities, thus ruling out adult onset of epileptic seizures.[6] On the following day, cognitive tests and exploration of ideomotor, ideational, and limb-kinetic praxis showed normal results, and no interhemispheric disconnection syndrome could be detected.[7,8] A lumbar puncture did not reveal any inflammatory or infectious processes. Given the normal structure of frontal ventricles, we ruled out normal pressure hydrocephalus (gait, sphincter control, and cognition were unaltered). The transient nature of the clinical manifestation led us to consider syncope as the probable diagnosis, regardless of any brain malformations. Fig. 1 shows findings of CC and agenesis of the corpus callosum. Case 2 is a 60-year-old man admitted to our hospital due to respiratory tract infection. He presented confusional syndrome

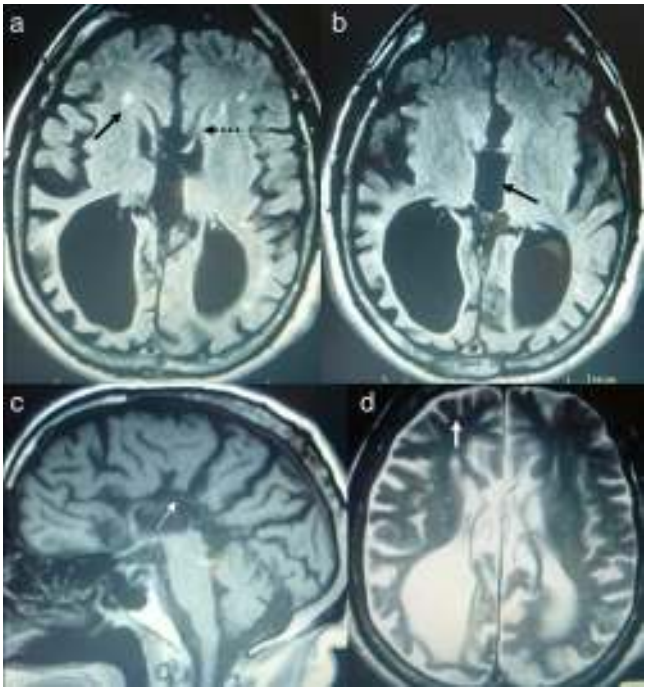

Figure 1 Head MRI scan of case 1. a) Axial FLAIR sequence. Frontal ventricles (dotted arrow) are small, and the frontal parenchyma features small hyperintense images (arrow) that could be associated with arterial hypertension. The third ventricle and occipital horns are dilated (CC). b) Axial FLAIR sequence. The third ventricle appears dilated (arrow) and the upper part of the corpus callosum is very thin. c) T1-weighted sagittal slice: the genu and trunk of the corpus callosum (arrow) seem faint. The fourth ventricle is normal-sized. d) T2-weighted axial slice. Frontal sulci are much smaller than cortical sulci on the posterior part of the brain.

☆ Please cite this article as: Bartolomé EL, Cottura JC, Britos Frescia R, Domínguez RO. Colpocefalia y agenesia parcial de cuerpo calloso asintomáticos. Neurología. 2016;31:68—70.