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BILATERAL PERISYLVIAN SYNDROME NOT RELATED TO MALFORMATIONS

REPORT OF TWO CASES

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ABSTRACT - In this case report we present the neuroimaging findings and clinical features of two patients with a bilateral perisylvian syndrome not related to malformations, but probably to ischemic etiology. Evaluations including history, general and neurologic examinations, electroencephalograms, and imaging data were reviewed as recent literature about the subject.

KEY WORDS: bilateral perisylvian syndrome, pseudobulbar paralysis, CT-scan, magnetic resonance imaging.

Síndrome peri-silviana bilateral não relacionada a malformações: relato de dois casos

RESUMO - Neste relato de caso, apresentamos os achados de neuroimagem e os aspectos clínicos de dois pacientes com síndrome peri-silviana bilateral não relacionada a malformações, mas provavelmente a uma etiologia isquêmica. Avaliações, incluindo história, exames físico e neurológico, eletrencefalograma e aspectos de imagem foram revistos, assim como a literatura recente sobre o assunto.

PALAVRAS-CHAVE: síndrome peri-silviana bilateral, paralisia pseudobulbar, tomografia computadorizada, imagem por ressonância magnética.

The anterior bilateral perisylvian or Foix-Chavany-Marie syndrome (FCMS)[3] is characterized by a pseudobulbar syndrome related to bilateral lesions at the anterior opercular region. This was described initially in adults[3,7], due to bilateral infartcs, but it can be found in childhood, due to brain malformations around the Sylvian fissures[1,4-6], occlusive vascular lesions, neoplasms and meningo-encephalitis, as an extremely rare condition[8,9]. The pseudobulbar syndromes can be divided in three clinical presentations: (I) Cortical (Foix-Chavany-Marie), without emotional lability, (II) Striate, with emotional lability and (III) Pontine, with facio-pharyngo-glosso-masticatory diplegia and emotional lability, but without dementia[6,10].

In this report, we present two children who have never developed speech, who have seizures, a pseudobulbar syndrome, and developmental delay, mainly in the cognitive area.

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CASE REPORTS

Case 1. A 16-years-old boy with delayed milestones and difficulty in swallowing. He does not speak but expresses himself through gestures. He presented 2 generalized atonic seizures, with loss of consciousness, the first at 14 months, after which he might have stayed in a coma for days, and the second one when he was 12 years old. The boy has significant drooling and orofacial hypotony. He understands simple orders through gestures and writes some words. His hearing is normal. He has a lowered cognitive level and had 3 normal electroencephalographic studies. He carries a left-sided hemiparesia, mainly in the upper limb. A contrast-enhanced CT scan showed retractile, hypodense, circumscribed lesions, which involves the cortex and white-matter, in both the temporal opercular regions, extending to the parietal lobe on the right side, and another lesion with the same characteristics in the right frontal region. On MRI these lesions were hypointense in T1-weighted images and hyperintense in the T2-weighted (Fig 1) and proton-density images. Nowadays his cognitive difficulties are stable and there has been an improvement related to the motor deficiency. The seizures were controled.

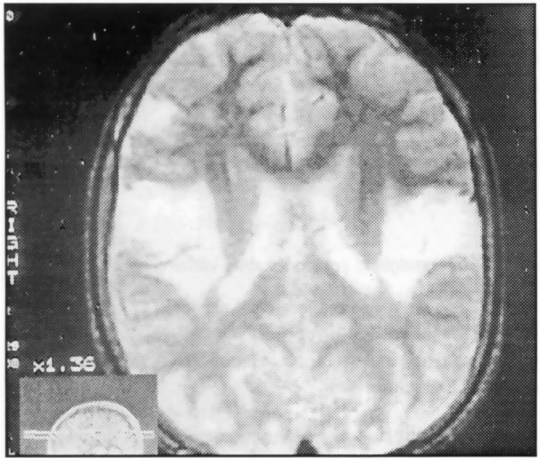

Fig 1. Case 1. MRI T2-weighted axial image (2200/90/ 1 [repetition time/echo time/excitations])demonstrates hyperintense lesions.

Case 2. A 17-years-old girl with speech difficulty, born after a normal pregnancy and delivery. Her father lived in Hiroshima and was nearby when the atomic bomb exploded. There is no such condition in her family, however. She had no developmental delay, except for speech acquisition and nocturne sphincteric control. With 8 years she started having partial versional head seizures,with occasional secondary generalization. The seizures are difficult to control, at irregularly monthly intervals. On examination, she is not able to express herself through speech or any form of written language. She communicates through her eyes, though. She responds to simple orders and does some calculation. She also has a lowered cognitive level and orofacial hypotony. Many electroencephalographic studies showed relevant irritant activity on the right cerebral hemisphere, mainly on the temporal lobe. A contrast-enhanced CT

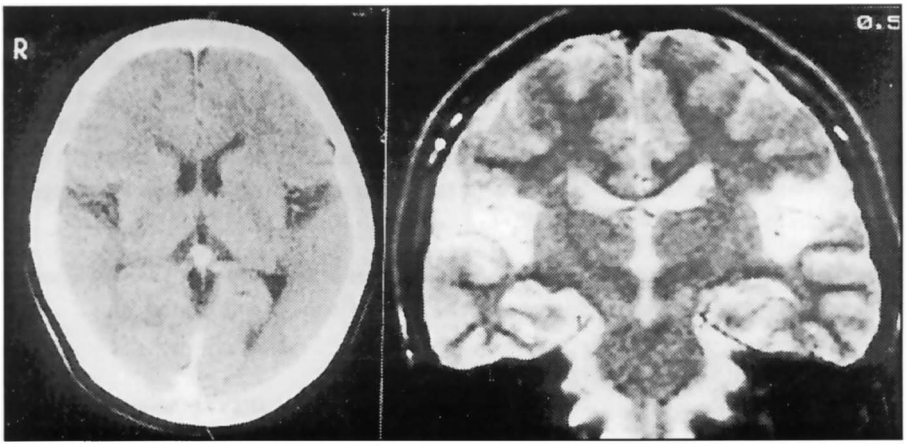

Fig 2. Case 2. A, Contrast-enhanced CT scan study also shows retractile, circumscribed, hypodense lesions on both opercula. B, MRI T2-weighted coronal image (2200/90/1) demonstrates hyperintense lesions at the frontal and temporal opercula.

scan study also showed retractile, circumscribed, hypodense lesions on both opercula (Fig 2A). On MRI there were hypointense lesions on T1-weighted images that were hyperintense on T2-weighted images at the frontal and temporal opercula (Fig 2B). Nowadays her clinical state is stable, despite some difficulty in controlling the seizures.

DISCUSSION

Bilateral perisylvian damage, sufficiently circumscribed so as not to cause major motor or sensory deficiencies of the limbs, or visual field defects, is capable of giving rise to one of two clinical patterns according to its site: (a) the temporal ("posterior") variety marked by "pure word deafness" with or without acoustic agnosia, amusia, or other neuropsychological deficiency, or (b) the frontal ("anterior") variety, i.e., FCMS, with facio-pharyngo-glosso-masticatory diplegia and automatic voluntary dissociation of the affected musculature[7]. The pseudobulbar symptoms are a direct result of the involvement of both the insular and opercular regions[6], although this cortical form can be found also due to deep subcortical infarcts[2]; any lesion of the supranuclear cortico-pontine and/or cortico-bulbar pathways can lead also to this clinical feature.

The anterior perisylvian syndrome may be either congenital or acquired. Mariani et al[7] revised 19 cases of FCMS in adults, with masticatory, facial, pharyngeal and voluntary lingual paralysis with automatic voluntary dissociation, which were bilateral in most cases, and concluded that the etiology is nearly without exception ischemic in nature, always with an acute onset, and stated them in 11 cases as circumscribed softenings in the rolandic and prerolandic distribution of the middle cerebral artery.

The congenital presentation of this syndrome has been emphasized in recent literature[4,6], and its main cause are the cortical dysplasia type malformations. This form frequently includes delayed milestones, variable motor deficiencies, seizures and important cortical pseudobulbar symptoms. The degree of oromotor dysfunction does not seem to correlate with the extension of the malformations on MRI, but it depends on their symmetrical distribution. Patients with asymmetrical opercular and insular abnormalities tended to have milder forms of dysarthria. Those with pyramidal limb motor dysfunction had evidence on MRI of extended malformations into the prefrontal and central regions[6].

Kuzniecky et al[6] have found bilateral perisylvian and perirolandic abnormalities in all of their patients. The lesions were symmetrical in 80% of the cases. CT scans demonstrated thick smooth cortex in the opercular region, but MRI revealed that some cortical areas had multiple small gyri and, in some cases, absence of normal white-matter digitations within the lesions, suggesting, contrary to early belief, polymicrogyria in the opercular and perisylvian regions[5]. Although it is usually not possible to distinguish the different forms of cortical dysplasia on imaging studies, these features and the well-known predilection of the perisylvian region for polymicrogyria and the insular distribution support this hypothesis[1,6].

Prats et al[8] described a 23-month-old infant with opercular syndrome of acute onset and focal seizures involving his face. As time passed, mutism, with normal language comprehension, became the main neurological sequel. He had had a first normal CT scan. A second CT scan, later, showed an area of hyperdensity in both rolandic regions, with slight contrast enhancement. A T2-weighted MRI scan 46 days after the onset of the illness showed bilateral, sharply demarcated areas of increasing signal intensity in the rolandic and opercular regions. The clinical features were considered suggestive of an encephalitic infection of viral origin. Long-term follow-up showed no other underlying cause leading to brain infarction such as a vasculitic process, metabolic disorder, chronic progressive encephalopathy or an epileptic syndrome prone to develop convulsive status.

Moodley and Bamber[8] described a three-year-old African boy who initially presented a meningitis that was later considered to be of tuberculous origin, and two weeks after admisssion, having been on anti-tuberculous chemoterapy, suddenly developed a pseudobulbar syndrome. CT

scan showed bilateral low densities in both opercula and insular regions. Subsequent CT scans performed two months later and other scans one year afterwards showed gross infarctions in both opercular regions, with mild dilatation of all ventricles, considered to be most probably the result of tuberculous vascular occlusive disease in both middle cerebral arteries.

Although having presented it early on infancy, our two patients had no evidence of malformation on CT and MRI scans, which suggest an acquired cause at a later phase of the intra-uterine or even early extra-uterine development.Because perinatal ischemic injury tends to present a pattern that is different from the adult one, with a greater involvement of the periventricular white-matter, the lesions in our patients suggest infarcts caused by oclusion of middle cerebral arteries branches.

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