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Abnormal Involuntary Movements and Hydrocephalus

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RESUMEN: Antecedentes: Se han descrito movimientos involuntarios anormales en pacientes con hidrocefalia; sin embargo, su etiopatogenia no ha sido suficienemente esclarecida, especialmente en lo que respecta al temblor. En el presente trabajo estudiamos las disquinesias observadas en pacientes con hidrocefalia, sus características clínicas y demográficas y la relación entre ambos problemas. Métodos: Serie de casos clínicos observados en un período de 10 años en un Servicio de Neurología de un hospital de referencia de tercer nivel. Nueve enfermos, 6 varones y 3 mujeres (edad media: 67 años) en los que la hidrocefalia antecedió a la aparición de disquinesias. Resultados: La hidrocefalia antecedió por 2.33 años a la aparición de las disquinesias y estas fueron: temblor en 6, parkinsonismo en 1 y distonía en 2. Cinco de estos enfermes tenían antecedentes de disquinesias entres sus padres o hermanos. En 4 de ellos, el tratamiento de la hidrocefalia con válvula de derivación ventrículo-peritoneal hizo desaparecer los movimientos anormales. Conclusiones: La hidrocefalia puede ser causa desencadenante de disquinesias (temblor, parkinsonismo y distonía cráneo-cervical) en un grupo de pacientes con una determinada susceptibilidad, que se encuentren en la sexta década de la vida y que tengan antecedenes familiares de trastornos del movimiento. Posiblemente la hidrocefalia por distorción mecánica o la alteración del flujo sanguíneo a los ganglios basales, o ambos, ocasione un desbalance entre los impulsos centrales y periféricos para el aparecimiento de temblor y parkinsonismo; y por otro lado cause desinhibición del control que los ganglios basales ejercen sobre las motoneuronas de los núcleos motores trigeminal y facial, para el aparecimiento de la distonía cáneo-cervical.

ABSTRACT: Background: Abnormal involuntary movements have been described in patients with hydrocephalus. However, the etiophatogenesis of this association has not been clarifed. We study the presence of dyskinesia, as weel as its clinical and demographic characteritics in patients with hydrocephalus. Design and patients: Series of cases studied during a 10 year period in a neurologic service of a third-level reference hospital. Nine subjects, 6 men and 3 women (mean age: 67 years) in whom hydrocephalus proced dyskinesia. Results: Hydrocephalus preced in 2.33 years the appearence of dyskinesia. Dyskinetic symptoms included tremor in 6 patients, parkinsonism in 1, and dystonia in 2. Five of these patients had family history of dyskinesia in parents or siblings. In 4 of them, the placement of a ventriculoperitoneal shunt improved the abdominal movements. Conclusion: Hydrocephalus may trigger dyskinesia (tremor, parkinsonism, and cranial-cervical dystonia) in a group of susceptible patients who are in their sixties and have a familiy history of movement disorders. It is possible that hydrocephalus due to mechanic distrotion or to alteration of blood flow to the basal ganglia or both, causes an unbalance between the central and the peripheral impulses for tremor and parkinsonism to appear; on the other hand, ti might unlock the control that basal ganglia exert on the motor-neurones of the trigeminal and facial motro nuclei thus triggering the cranial-cervical dystonia.

Hydrocephalus has been considered as the cause of abnormal involuntary movements (AIM), including parkinsonism, (1-3) blepharospasm, (4) blepharospasm, palatine myoclonia, (5) and tremor. (6) Oppenheim and Bing were probably the first authors who made reference to this association, Oppenheim to acute hydrocephalus and the presence of action/intention tremor, adn Bing to the presence of tremor in congenital hydrocephalus (quoted by Keane). (6) It has been considered that dystonia associated with hydrocephalus is related to a disinhibition of the control that basal ganglia exert on motor-neurones in the brainstem, specially on facial nerve nuclei. (7) Such dystonia is classifield as secondary from disorders involving brainstem and diencephation. (7,8) Tremor-related hydrocephalus is usually present in the context of parkinsonism. Since resting, postura, and action/intention tremor have been observed after midbrain disorders as well, (9,10) it is difficult to explain their pathogenetic mechanism in such cases.

We presen 9 patients who developed hydrocepahlus and abnormal involuntary movements to beter understand the pathophysiology of this association. All patients were examined in the Neurology Service at the Carlos Andrade Marin Hospital betwen September 12, 1987, and November 27, 1997. The Short Test of Mental Status (11) was used to asses the mental state in every case. Normal pressure hydrocephalus was diagnosed according to Adams & Victor (12) with the triad of clinical signs including progressive walking impairment, mental funcion perfomed in patients with tremor, determining the tremor frecuency (Hz). All patients were assessed at the beginning and during their owen individual follow up period by at least one of the authors.

CASE DESCRIPTIONS

Patient 1. A 74-year-old man was evaluated because of a 7-year history of a progressive walking disorder associated with distraction, forgetfulness, urinary incontinence, and head tremor presented both at rest and during chore/work execution. There was no personal or family history of neurologic disease. He had been taking biperiden (4 mg/day) and clobazam (10 mg/day) without improvement. Neurological examination revealed tremor in both sides specially in hands (resting tremor 1/3, postural tremor 3/3, and action/intention tremor 1/3). Less severe tremor was also observed in the head and legs, and there was a bilateral Froment´s sign. TEr was 7Hz. Mental status assessment was 19/38 and a CT of the head revealed hydrocephalus that was considered to be normotensive in natura. An acute teste with L-Dopa (250 mg) was negative. Drainage of CSF through a lumbar puncture did not improve the walking difficulty or the sphincter distrubance, but ameliorated the tremor. He has been followedup for 4 years, with a adequate control of tremor with repeated CSF drainage (+-25cc) every 45 days.

Patient 2. An 80-year-old black woman was evaluated because of a 7-year history of progressive memory loss asociated with generalized tremor, walking difficulties, and urinary incontinence. She had a sister with tremor. The patient had been terated with primidone (750 mg/day) without improvement. Neurological examination revealed generalized tremor (resting tremor 1/3,

postural tremor 3/3, and action/intention 23/38. CT of the head showed normal pressure hydrocephalus. Drainage of CSF through a lumbar puncture did not improve tremor contrll. He was followed-up during 3 years, without improvement.

Patient 3. A 75-year-old man presented with an 8 year history of progressive walking disorder associated with intellectual deterioration and urinary incontinence. During the last year he had been experienced hand tremor. He has a 59-year-old sister with diagnosis of esential tremor.The patient had been treated with pronolol (160 mg/day) with no results. Neurological examination revealed hand tremor (resting tremor 1/3, and postural tremor 1/3). Ter was 7 Hz. There were no bradykinesia or rigidity. A CT scan of the head revealed hydrocephalus as well as multiple cystic lesions located over the cerebral hemisheres (cortical and subcortical areas) without involment of basal ganglia. Anticysticercal antibodies were present in CSF. With a diagnosis of neurocysticercosis, he received albendazole (15 mg/kg/day for 8 days) and underwent the placement of a ventricular shunt for relief of hydrocephalus due to shunt dysfunction. The shunt was replaced and he began to improve. Three months after the second he began to improve.Three months after the second surgery, tremor improved and 6 months after, it disappeared. Oney year later, the tremor has not recurred.

Patients 4. A 66-years-old man complained a slowing progressive walking disorder during the las 10 years. During the year prior to admission, he had been experienced moderated resting tremor in the 4 limbs that did not respond to L-dopa therapy (up to 1250 mg/day). One month before admission, he had been experienced moderated resting tremor in the 4 limbs that did not respond to L-dopa therapy (up to 1250 mg/day). One month before admission he developed headache, vomiting, and diplopia. Neurological examination revealed paresis of the right VI cranial nerve, generalized tremor (resting tremor 2/3 and postural tremor 1/3), a bilateral Froment´s, generalized rigidity, and symmetric bradykinesia. CT scan of the head showed obstructive hydrocephalus related to a fourth ventricle cysticercuso. In addition, there were cystic lesions in the left sylvian fissure, and the right patietal lobe, and multiple calcifications disseminated in the brain parenchyma. CSF examination revealed anticysticercal antiboide by ELISA. He was treated with albendazole (15 mg/k/day for 8 days) and underwent a posterior fossa craniotomy for removal of the fourth ventricle cysticercus. In addition, a ventricular shunt was placed for relief of hydrocephalus. Hedache disappeared an diplopia improved after surgery. Tremor and bradykinesia also improved, and the patient was discharged taking clonazepam (2 mg/day). A 3-year follow-up confirmed the good prognosis noticed at the time of the hospital discharge.

Patient 5. A 40-year-old woman was evaluated because of acute headache and vomiting. He had a 4-year history of mild headache associtaed with slow and progressive disminution of visual acuity. She also complained of postural and action/intention tremor that started in the left hand and latter comprised the righ hand and legs. Tremor had been improved with clonazepan (16 mg/day); however, side-effects of that drug made him to withdraw therapy. The mother of the patient (63-year-old) has essential tremor. On admission, the patient was lethargic without loalizing signs. The EEG showed bilateral fronto-temporal theta waves. CT scan of the head revealed hydrocephalus due to a third ventriclecysticercus as weel as multiple calcifications and parenchymal brain cystic lesions that involved the cerebral cortex and subcortical white matter of both cerebral hemispheres without compromise of basal ganglia. He received albendazole (15 mg/kg/day for 8 days) and prednisone (50 mg/day). Control CT scan revealed disappearance of all cystic lesions without change in the size of the ventricular system. A ventricular shunt was placed and 15 days later he experienced improvement in tremor which disappear in 7 more days. A 4-year followup shoed no recurrence of symptoms.

Patients 6. A 60-year-old man was evaluated because of a 6-year history of slowly progressive memory distrubances, disorientation, a walking disorder characterized by increase in the support base and short steps, and urinary incontinence. Three years before admision, he expereinced speech ailment-enunciation difficulty and interrupted pronunciation that also progressed slowly. He also noticed that his head tended to hyperextensión, that increased slowly until it reached a fiexed posture. He received clonazepan in doses of 12 mg/day, without response. A 67-year-old brother of the patient had essential tremor. On admission, neurological examination revelaed a fixed retrocollis that and diminished during sleep. His voice was interrupted by dystonic emissions. A CT scan of the head hydrocepalus associated hypo- and hyperdense cysted lesions located in the cerebral cortex and subcortical white matter that did non involve the basla ganglia. A lumbar puncture yeldied a turbid CSF, and the cytochemical analysis revealed 35 ce per mm3, 56 mg/dl proteins, and anticysticercal antibodies by ELISA. The patient was treated with albendazole 15 mg/kg/day for 8 days and undewent the placement of a ventricular shunt. Ten days after surgery his memory and walking distrubance began to improve . One month after surgery his urinary control improved. One month later, the speech and pronuntiation also improved, and by the 8^{th} month after surgery, the position of the head began to improve. We was asymtpomatic at the end of a one year follow-up period.

Patient 7. A 46-year-old woman was evaluated because of a 5-year history of a walking disturbance. Four years before admission, she experienced hand tremor, urinary incontinence and memory impairment as weel as urinary incontinence progressed over the time. On admission, the neurological examination revealed a short-step walk with increase of sustentation, apraxia, and generalized tremor (postural tremor 2/3 and intentiomor frecuency was 9 Hz. Mental status assessment was 25/38. A CT scan of the head revealed normal pressure punctures and CSF evacuation; however, she did not improved in the 2-year follow-up period.

Patient 8. A 77-year-old man presented with a 4-year history of walking difficulties, memory impairment, urinary incontinence, and bilateral hand tremor. He was a 74-year-old brother with Parkinson diseases. At 70 the patient presented walking torridness, memory disorders and lack of sphincter control thath progressed slowly. He had been treated with propanolol (120mg/day) and clonazepan (8mg/day) with minimal improvement. On admission, neurological examination revealed generalized tremor (resting tremor 1/3 postural tremor 3/3, and action/intention tremor) and bilateral Froment´s signs. A CT scan of the head revealed normal pressure hydrocephalus. He underwent the placement of a ventricular shunt. He began to improve after surgery, including urinary control, memory, and tremor. The latter resolved 4 months after the shunt. There has been no evidence of recurrence in tremor during the 3-year follow up.

Patient 9. A 88-year old man began with progressive memory loss at 81. Sometime later, he presente walking difficulties and urinary incontinence, and by the age of 84 he developed retrocollis. He was treated with biperiden (6 mg/day) but treatment was complicated with several event of urinary tetention. Neurological examination revealed a fixed position retrocollis. Mental status assessment was 22/38. A CT scan of the head revealed normal pressure hydrocephalus. Evacuation of CSF throught a lumbar punctures induced to 29/38, and there was an important walking recuperation. He was no amelioration of the dystonic posture of the head. He was treated with clonazepam (3mg/day) that was well tolerated. However, there has been no change durigng a 2-year follow-up.

RESULTS

The study included 9 patients (6 men and 3 women) with ages ranging from 40 to 88 years old (mean age: 67.33+-21.4 years) who had hydrocephalus and an involuntary movement disorder. Etiology of hydorcephalus include normal pressure hydrocephalus in 5 patients an neurocysticercosis in the ramaining 4. No patient had been treated with neuroleptics or any other drug that may induce the develpment of involuntary movements. Six of the 9 patients had tremor, 2 had dystonia, and one had parkinsonism. Clinical manifestations of hydrocephalus preced the develpment of involuntary movents for a mean of 2.33 years.

Five or the 9 patients were treated with a ventricular shunt and the remaining 4 were managed with repeated tumbar puctures and CSF drainage. One of the patients olso undewent surgical removal of a fourth ventricle cysticercus. Beforme surgery, all patients had been treated with different drugs for the control of involuntary movements disappeared after the shunting procedure in 3 patients with tremor and in one with retrocollis, and partially improved in the patient with parkinsonism. Three of the 4 patients who had complete recovery had hydrocephalus related to neurocysticercosis and the other hand, drainage of CSF through repeated lumbar puncture only improved the involuntary movemnts in one of 4 patients. Interstenly, all the patients who improve after either ventricular shunt placement of CSF drainage, had a family history of involuntary movement disorders.