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CASE REPORT
ΕΝΔΙΑΦΕΡΟΥΣΑ ΠΕΡΙΠΤΩΣΗ

Extrapyramidal symptoms as a ventriculoperitoneal shunt complication in a 12-year-old male child

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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2020, 37(3):387–390

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Hydrocephalus is one of the most common clinical conditions affecting the central nervous system, with an incidence of congenital hydrocephalus of 3–4 per 1,000 births. Placement of a ventriculoperitoneal shunt (VPS) is an established treatment to regulate the drainage of cerebrospinal fluid (CSF) in hydrocephalus, but several complications may occur. We report here the case of a 12-year-old male child who presented extrapyramidal symptoms following the placement of a VPS for the management of occlusive hydrocephalus. Extrapyramidal symptoms have been reported only rarely as a complication in patients with VPS.

Εμφάνιση εξωπυραμιδικής συμπτωματολογίας ως επιπλοκή κοιλιοπεριτοναϊκής παροχέτευσης σε άρρενα ασθενή ηλικίας 12 ετών

Περίληψη στο τέλος του άρθρου

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Hydrocephalus is a common disorder of cerebral spinal fluid (CSF) physiology, resulting in abnormal expansion of the cerebral ventricles, typically associated with increased intracranial pressure. Hydrocephalus is one of the most common clinical conditions affecting the central nervous system with an incidence of congenital hydrocephalus of 3–4 per 1,000 births. Treatment options include shunt placement and endoscopic procedures, individualized to the child. The long-term outcome for children treated for hydrocephalus varies.[1]

Treatment for hydrocephalus has not advanced appreciably since the introduction of CSF shunts more than 50 years ago. Modern shunting began in the 1950s when ventriculoatrial (VA) shunts set a new standard in hydrocephalus treatment. The first VA shunt was implanted in Norway in 1961, and VA shunts were the standard treatment procedure until 1979. Beginning in 1980, use of ventriculoperitoneal shunts (VPS) became more common, because of a few, but fatal, complications from VA shunting. Since then, VPS have been the standard treatment for patients with hydrocephalus.[2]

The outcome of children with hydrocephalus has been the object for several studies. The risk of shunt malfunction gradually declines over the years, but it persists through life, and children treated for hydrocephalus therefore require life-long follow-up. We report here the rare case of

a 12-year-old male child who developed extrapyramidal symptoms as a complication of a VPS.

CASE PRESENTATION

A 12-year-old boy was admitted to the pediatric department with a 2-week history of persistent dizziness and headache. One year earlier he had received a VPS for management of occlusive hydrocephalus caused by stenosis of the aqueduct of Sylvius. The child had an unremarkable perinatal and family history; he was born full term, weighting 3,500 g, with Apgar score 1^{10} and 5^{10}.

His initial neurological examination on admission revealed anisocoria with the right pupil slight larger than the left, mild cognitive impairment and periodic alteration of alertness, with remarkable bradypsyche. During the next few days, he developed irritability, upward gaze palsy, upper limb intentional tremor and gait disturbances (unsteadiness and ataxia).

The vital signs were within the normal range and the complete blood count, arterial blood gases and biochemical values were normal. Cardiological examination, with electrocardiogram (ECG) and echocardiogram, was unremarkable. The electroencephalogram (EEG) revealed bursts of monorhythmic theta or delta waves in both hemispheres. Fundoscopic examination excluded papillary edema.

Cranial and abdominal X-ray confirmed the position of the shunt. Cranial computed tomography (CT) showed a right-sided VPS, and normal brain structure, with a small calcified region in the left thalamus. Following neurosurgical consultation, brain magnetic resonance imaging (MRI) showed a right-sided VPS, normal brain structures, and a small high intensity region in the left thalamus causing obstruction of a small degree in the aqueduct of Sylvius and slight enlargement of the third ventricle.

Because of worsening of his tremor and development of dysarthria the patient underwent emergency lumbar puncture, showing biochemical, microbiological and cytological analysis of the CSF within normal limits. Antibody titers in the serum and CSF for autoimmune encephalitis and viral encephalitis (antibodies against herpes simplex virus types 1 and 2, mycoplasma, cytomegalovirus) gave normal results. Medication with valproic acid, clonazepam, acyclovir, ceftriaxone was started. Following revision of the MRI scan, emergency CSF drainage from the catheter was conducted, with impressive improvement of the extrapyramidal signs.

The following day a new shunt was inserted and the old shunt was removed. Postoperatively, the child became febrile, with pleocytosis of the CSF and worsening of his general condition and reappearance of the extrapyramidal signs. The new shunt was removed and external drainage was inserted. The CSF cultures were negative, but measurement of the levels of CSF neurotransmitters (5-hydroxyindoleacetic acid, homovanillic acid, 3-ortho-methyldopa, 5-hydroxytryptophan, neopterin, biopterin) revealed an increase in the ratio 5-hydroxyindoleacetic acid/homovanillic acid (4.7, normal range 1.32–3.28). Treatment with levodopa temporarily improved the tremor.

The child became afebrile after few days with antibiotic therapy (teicoplanin, meropenem), but without further improvement in his extrapyramidal signs or dysarthria. On the 26th day of admission the patient underwent placement of a second shunt, which was followed by resolution of his extrapyramidal signs within 72 hours. The patient was discharged a week later with instructions for physiotherapy and under treatment with valproic acid and levodopa. At follow-up after two months he showed no extrapyramidal signs or dysarthria.

DISCUSSION

Hydrocephalus is the most common condition treated by pediatric neurosurgeons. The prevalence of infant hydrocephalus is roughly one case per 1,000 births, but the incidence is probably higher in developing countries. The most common causal mechanisms in high-income countries are post-hemorrhagic hydrocephalus of prematurity, congenital aqueduct stenosis, myelomeningocele and brain tumors.[3]

Shunting became the standard treatment for hydrocephalus in Norway during the 1960s. The most common type of shunt is the VPS, which diverts CSF from the ventricles to the peritoneal cavity, although other distal sites such as the right atrium of the heart and the pleural cavity are sometimes used. VPS is still considered the gold standard for the treatment of communicating hydrocephalus, and is also an option for the treatment of non-communicating hydrocephalus in immature patients or when there are anatomical limitations for endoscopic third ventriculostomy (ETV).[4] A shunt generally consists of silastic tubing that runs subcutaneously from the head to the abdomen, with a valve between the ventricular and distal catheters. Differential pressure (with either fixed or programmable settings) or flow-regulating valve mechanisms are often paired with antisiphon or gravitational devices to prevent CSF overdrainage from posture-related siphoning. Despite technological progress, valve design appears to have little, if any, effect on shunt efficacy or failure rates.[5]

Extrapyramidal motor signs in patients with hydrocephalus include Parkinsonian and Huntingtonian movements. Hydrocephalus may cause extrapyramidal symptoms, most likely due to local pressure on tracts of the nigrostriatal pathway or the cortico-striato-pallido-thalamo-cortical circuit. Parts of these pathways lie in close proximity to the ventricular system and may be subjected to volume effects or ischemic changes secondary to ventriculomegaly, resulting in a hypokinetic rigid syndrome, or tremor.[5] During the course of hydrocephalus, periods of elevated intracranial CSF pressure are hypothesized to result in dysfunction of

the white and gray matter of the brain, including the basal ganglia. These hypotheses are supported by functional imaging studies and by post-mortem histopathological analysis of brain parenchyma.[6] The pathophysiology of Parkinsonian symptoms in hydrocephalus has not been conclusively understood. The abnormal pulsation of CSF flow occurring in hydrocephalus may produce secondary damage to the nigrostriatal dopaminergic pathway and a downregulation of dopamine receptors in the striatum and putamen. The mesolimbic dopaminergic pathway and the ascending reticular activating system may also be injured in some cases of severe and long-standing hydrocephalus.[7]

Children with treated hydrocephalus face many potential long-term complications. Despite advances in technology and precision neurosurgical techniques, VPS failure continues to be a considerable clinical problem globally. Common causes of VPS failure include infection, shunt catheter blockage, and over- or underdrainage.[8] Shunt failure, usually from mechanical obstruction, occurs in almost 40% of children in the first 2 years after shunt placement. The rate of shunt infection is about 5–9% per procedure, and infection usually occurs within 3 months of surgery. Shunt overdrainage can present either acutely, with subdural hygroma or hematoma, or chronically, with the so-called slit-ventricle syndrome. Mortality due to hydrocephalus and its treatments is reported between 0 and 3%, depending on the duration of follow-up. Shunt event-free survival (EFS) is about 70% at one year and 40% at 10 years.[9] Common manifestations of the long-term complications in patients with shunt treated hydrocephalus are motor development disturbances, cognitive and visual impairment, epilepsy, hearing loss and endocrine disorders.[10] Patients with untreated hydrocephalus have progressive brain dysfunction due to ventricular enlargement and increased intracranial pressure.[11]

Review of the literature revealed few previously reported cases of Parkinsonian syndromes associated with hydrocephalus, all of which affected adults. Curran and colleagues reported 9 cases of obstructive hydrocephalus associated with marked parkinsonism in 1994. They discussed the possibility that the pathophysiology of hydrocephalic parkinsonism involves various sites of dysfunction in the nigrostriatal pathway and the cortico-striato-pallido-thalamo-cortical circuit. At certain locations these pathways lie in close proximity to the ventricular system and may be subjected to mass effects and ischemic changes secondary to ventriculomegaly.[12] We report here a case of a 12-year-old male child presenting with extrapyramidal symptoms following placement of a VPS for occlusive hydrocephalus. To the best of our knowledge this is the first case reporting Parkinsonian syndromes associated with hydrocephalus in childhood. Establishing the diagnosis in patients with hydrocephalus presenting with extrapyramidal signs is often difficult and requires further attention.

ΠΕΡΙΛΗΨΗ

Εμφάνιση εξωπυραμιδικής συμπτωματολογίας ως επιπλοκή κοιλιοπεριτοναϊκής παροχέτευσης σε άρρενα ασθενή ηλικίας 12 ετών

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Αρχεία Ελληνικής Ιατρικής 2020, 37(3):387–390

Ο υδροκέφαλος αποτελεί μια από τις συχνότερες παθολογικές οντότητες του κεντρικού νευρικού συστήματος, με συχνότητα που υπολογίζεται σε 3–4/1.000 γεννήσεις. Η κοιλιοπεριτοναϊκή παροχέτευση αποτελεί τη θεραπεία εκλογής, ωστόσο ενέχει τον κίνδυνο εμφάνισης διαφόρων επιπλοκών. Περιγράφεται περίπτωση άρρενος ασθενούς, ηλικίας 12 ετών, με κοιλιοπεριτοναϊκή παροχέτευση λόγω αποφρακτικού υδροκεφάλου, ο οποίος εμφάνισε εξωπυραμιδική συμπτωματολογία. Η εξωπυραμιδική συμπτωματολογία σπάνια περιγράφεται ως επιπλοκή σε ασθενείς με κοιλιοπεριτοναϊκή παροχέτευση.

Λέξεις ευρετηρίου: Εξωπυραμιδικά, Επιπλοκές, Παιδιά, Παροχέτευση, Υδροκέφαλος

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