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G. Iacob



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G. Iacob

Professor of Neurosurgery. "Carol Davila" University of Medicine and Pharmacy. Department of Neurosurgery, Emergency University Hospital, Bucharest, ROMANIA

ABSTRACT

Introduction: Aqueduct stenosis (AS) in old people is a rare pathological entity. I report a case of a 66 years old woman with severe ataxia, cognitive deterioration, loss of sphincters control (gatism). Clinical, neuro-radiologic and therapeutic considerations are discussed.

Case presentation: A 66 years old woman with a 6 months history of mild cerebral trauma by car accident without losing consciousness, present 2 months before hospitalization severe ataxia, cognitive disorders, gatism. At the time of trauma, brain scanner performed in another institution showed minimal fronto-basal cerebral contusions and blood collection around the brain stem. One month after a new unenhanced brain scan all previous lesions are gone. At admission, an abnormal enlargement of lateral and third ventricles are remarked both on unenhanced CT and MRI scan of the brain explained by cerebral aqueductal stenosis. Ventricular open pressure was 350 mm H₂O. A ventriculoperitoneal shunt with a variable pressure valve was installed. The surgery went uneventful and the patient recovered as expected. 6-month follow-up visit the patient was symptom-free, with a fine intellectual recovery.

INTRODUCTION

Abnormal enlargement of lateral and third ventricles due to aberrant accumulation of CSF induced by a AS, generally implies a congenital etiology; to those people that survive, an alternate CSF drainage pathway via the extracellular space (ECS) of the brain, with increasing resistance to CSF outflow with deep white matter ischemia, leads to symptoms onset in 3%-10% of adult hydrocephalus. To old persons, AS is much rarer (1)(2).

PHYSIOPATHOLOGY AND AETIOLOGY

A 66-year-old woman, teacher, has a previous 6 months history of a mild cerebral trauma by car accident without losing consciousness. At the time of trauma, brain scanner performed in another institution showed minimal fronto-basal cerebral contusions and blood collection around the brain stem. One month after a new unenhanced brain scan

Keywords

aqueductal stenosis (AS),
different pressure
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old people,
surgical techniques,
ventriculoperitoneal shunt
with variable pressure valve



Corresponding author:
G. Iacob

Bucharest University Emergency
Hospital, Romania

gbrl_cb@yahoo.com

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all previous lesions are gone. 2 months before hospitalization the patient present: headaches, dizziness, loss of coordination, severe ataxia, cognitive disorders, loss of sphincters control. Upon physical examination, the patient has an oral temperature of 37°C (98.6°F), blood pressure was 154/90 mm Hg, pulse was regular with a rate of 76 beats/min, her heart sounds are normal. She is unable to stand, due to a severe feeling of imbalance. She has a severe ataxia, the power was normal in all 4 limbs, with mild spasticity, no Babinski sign. Sensation to pinprick and temperature, the joint position and vibration sense are intact bilaterally. She has a sustained horizontal gaze-evoked nystagmus looking to the left and right, with a down beating nystagmus on downward gaze. The gag reflex is diminished. The tongue movements are normal. Routine laboratory analysis findings, including a complete blood cell count, a basic metabolic panel with renal function tests, blood sugar level, lipid profile, prothrombin time and concentration were normal. A fine ophthalmologic, psiyhatic and psyhologic clinical evaluation was performed: fundus oculi was normal, but severe cognitive disorders, especially affecting memory, attention were reported. Noncontrast CT scan of the head and cerebral & spine MRI to exclude a possible obstructive cause of hydrocephalus an AS was performed (Fig. 1).

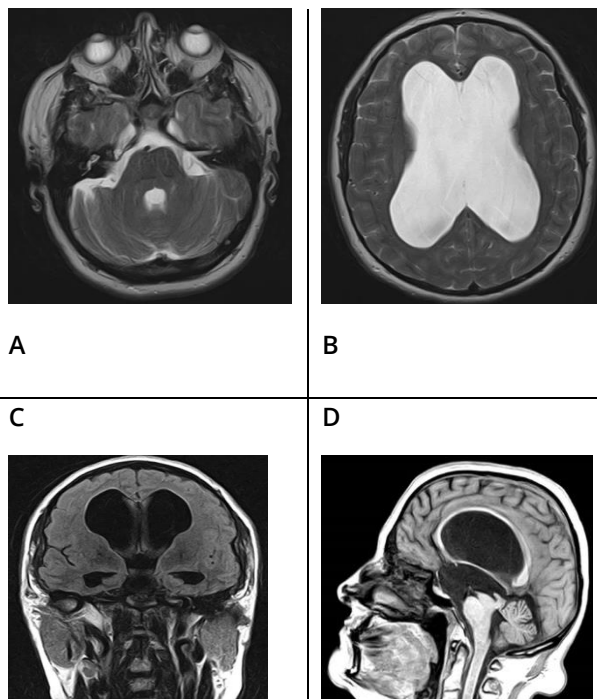


Figure 1. Pre-operative MRI A & B T2 axial, highlighting the 4th ventricle and the lateral ventricles, C & D T1 coronal and sagittal demonstrates the extent of hydrocephalus by AS.

A fine treatment tunnig using surgical shunt diversion with variable valve was installed (a Strata II variable valve – Metdronic was used); ventricular open pressure was 350 mm H₂O. The surgery went uneventful and the patient recovered as expected. 6 month follow-up visit the patient was symptom free, with a fine intellectual recovery. Unenhanced brain scanner demonstrate significant remission of the ventricular size (Fig. 2).

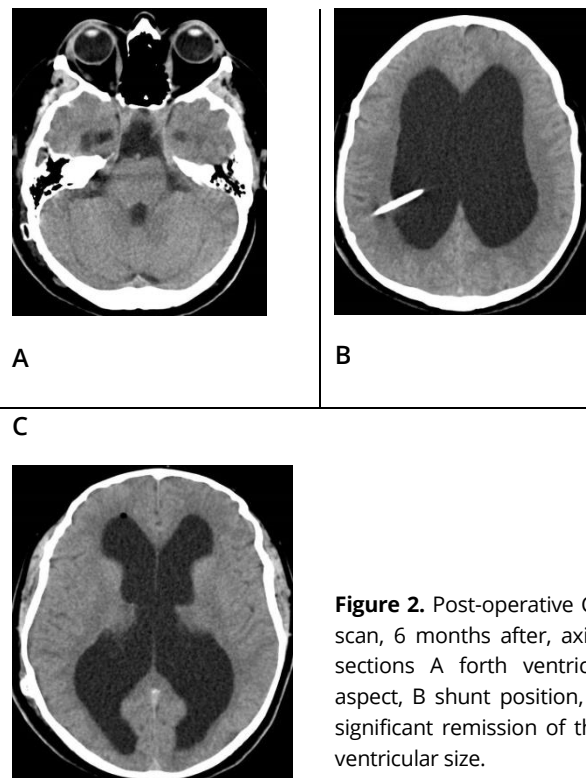


Figure 2. Post-operative CT scan, 6 months after, axial sections A forth ventricle aspect, B shunt position, C significant remission of the ventricular size.

DISCUSSIONS

Aqueductal stenosis (AS), is a pathological entity, generating abnormal enlargement of lateral and third ventricles due to aberrant accumulation of CSF, with normal 4th ventricle responsible for 20% of cases of hydrocephalus (3). More frequent in children 16-60% - its incidence ranges from 0.5 to 1.0 in 1,000 births, with a recurrence risk in siblings of 1.0% to 4.5%, it may be inherited in an X-linked recessive manner, see Bickers-Adams-Edwards syndrome (4). AS appears less in adult 3-10% - as an acquired abnormality and is rare in elderly people.

Historical data: LIAS - late-onset idiopathic aqueductal stenosis (5) was first described by Spiller in 1902, Schlapp and Gere in 1917 suggested the pathological relationship between AS and congenital hydrocephalus. Dandy resume this idea in 1920, 1945 (6)(7). From 1930 to 1977 there are more than 20 bibliographic references with this subject (8), subsequently, many works were performed elucidating the diagnosis and treatment.

Concerning etiology, there are several extrinsic and intrinsic causes for AS. However there is no correlation between aqueduct caliber and hydrocephalus: a partial stenosis may coexist with normal-sized ventricles; also complete stenosis it is difficult to admit at least on a microscopic scale, in a patient who has reached adulthood, in the absence of a macrocephaly - weighty argument for congenital origin, because there are possible alternate CSF drainage pathway via the extracellular space (ECS) of the brain, also late onset of congenital origin of AS (1)(5)(8)(9). The main causes of AS (1)(3-5)(8-12) are:

- congenital with large range incidence between 3.7:1,000,000 to 1:2000, rarely it may be inherited in an X-linked recessive transmission Bickers Adams Edwards syndrome: with congenital AS, corpus callosum and corticospinal spinal tracts agenesis, absence of medullary pyramids. Congenital AS can present later in adult age due to compensation of CSF flow dynamics during childhood and normalization of clinical symptoms (10);

- acquired with different causes: extrinsic and intrinsic. Extrinsic causes can generate AS by mechanic compression;

- tumors: posterior third ventricle mass, tectal plate glioma, pineal and posterior fossa tumor: metastasis, hemangioblastoma, glioma - even a subependymal astrocytoma, vestibular schwannoma, meningioma in contact with tentorial incisura;

- posterior fossa arachnoid cyst;

- cerebral vascular malformations: Galen malformation, basilar megadolico basilar trunk lifting the floor of the third ventricle;

- malformative causes: Chiari and Dandy Walker malformations;

- acquired causes: Paget's disease - by venous compression of bone origin, Guillain Barré syndrome, syphilis, sarcoidosis.

Intrinsic causes can be multiple:

- idiopathic or "simple", late-onset idiopathic AS especially in the elderly, apparently without a precise cause, not uncommon; aqueduct caliber could be obliterated by subarachnoid-villous system age alteration, or even narrowed with normal ependymal cells in the lumen, without gliosis of the surrounding tissues;

- reactive gliosis with glial cell and fibers proliferation, without ependyma (the residual lumen is not outlined by ependyma) as a reaction to:

1. subarachnoid hemorrhage (SAH) after an aneurysm rupture, posttraumatic - as in our case, neurosurgical interventions in elderly subjects after surgical sacrifice of CSF. resorption pathways, heavy bleeding, posterior cerebral fossa with opening of the fluid system;

2. toxic agents: a widespread ependymitis and ventriculitis generating AS at the time of scarring viral (urlian), bacterial, tuberculous, lymphomatous, carcinomatous, toxoplasmosis;

3. increased resistance to CSF flow, see deep white matter ischemia in late adulthood, venous hypertension, developmental venous anomaly draining bilateral medial thalami (12).

- intraventricular high pressures may generate vascular problem, decreased secretion, gradual increase in resorption capacities: trans-ependymal, choroid plexuses, optic nerve sheaths, large vessel adventitia, lymphatic pathways, residual gradient.

- biochemistry changes and physical properties of enclosures with visco-elastic walls, under persistent residual gradient generating structural changes of cerebral mantle.

According to the histopathological classification of Russell in non-tumoral AS there are four types (13-15):

1. "congenital atresia" - generating global caliber stenosis, with normal histology, due to a malformation of abnormal development.

2. "forking" aqueduct"- 2 distinct channels, in a sagittal plane, one behind the other, may communicate, may enter the ventricle independently, or end blindly are separated by normal nervous tissue expression of an incomplete fusion of the median fissure, reducing the lumen and/or perturbing the laminar flow of CSF.

3. "septum formation"- a webs or diaphragms, more frequent at the lower end of the aqueduct, gradually becomes a tiny sheet from prolonged pressure and dilatation of the canal above.

4. "gliosis" - characterized by proliferation of glial cells and overproduction of glial fibers, the residual lumen is not outlined by ependymal, usually a reaction to irritant agents, such as hemorrhage – see our case, infection, or toxic agents, and is often part of a widespread ependymitis of the ventricles. In such cases transition to chronicity involves different pathophysiological mechanisms acting simultaneously (10).

Clinical features in AS reflects the acute or chronic picture of the evolution of the disease (4)(5)(8)(10)(16)(17), due to alterations of CSF dynamics, with acute or compensated stages, explaining in congenital origin, undiagnosed aspects during several years: enlarging infant head size, bulging fontanelles, gaping cranial sutures, setting sun phenomenon, in X-linked form (Bickers-Adams-Edwards syndrome) profound intellectual disability, bilateral adducted thumbs. Subjective aspects in acute or subacute stages: headaches in attacks or continuous, variable seat, vomiting especially in posterior fossa tumor, decreased conscious state; in compensated stages episodes of headaches and nausea more severe in the morning and diplopia by abducens nerve palsy in intra-cranial hypertension syndrome, dizziness, somnolence, in adults with late-onset idiopathic AS more commonly have chronic onset of neurological symptoms, also pure akinesia with gait freezing (18). Objective aspects are variable in acute decompensated stage – see the present case, with: comitial crises, episodes of graying out of vision, papillary stasis on fundus oculi, Parinaud syndrome - upward gaze and accommodation failure, pupillary contraction abnormality, nystagmus, paralysis of extrinsic musculature, intracranial hypertension syndrome with Cushing triad - hypertension, reflex bradycardia, respiratory irregularities, rhinorrhea, "bobble head doll" syndrome with repetitive antero-posterior head movements; sphincter disorders are generally limited to urination disorders of varying severity: from pollakiuria to urinary incontinence in the final stage, rarely accompanied by anal incontinence, impaired consciousness, disturbance of the conscious level up to coma by brain engagement. In chronic stages: walking apraxia, standing disorder, with widening of the support polygon, retropulsion, falls; head wearing in a slight lateral inclination, ataxia, gait disturbance, bipyramidal and cerebellar syndrome by white matter tracts pressure

surrounding the ventricles, increased reflexes, paroxysmal attacks with hypertonicity of the axial muscles and limbs in opisthotonus, spastic weakness of lower limbs with gait disturbance, akinetic mutism is the most serious ultimate form of motor disturbance, neuro-psychological tests detect degradation, with visual-spatial or constructive difficulties associated with motor performance disorders; so called "the nonverbal learning disabilities syndrome" Fletxher 1995. There are also: behavioral changes of cognitive function, mental fatigue, attention disorders, depression, disinterest, temporal disorientation, sluggishness up to 40%, also abulia; vegetative disorders: tachypnea, bradycardia /tachycardia, hyperthermia, hypopituitarism disorder; diabetes; obesity.

In adults, with A.S. there are 3 possible situations (19)(20)

1. active hydrocephalus with PIC average > 12 mm Hg - unclear decompensation mechanism with possibly congenital aqueduct stenosis, after head trauma, infection, hemorrhage, even stenosis secondary to hydrocephalus, itself secondary to a more distal obstacle, thus forming a vicious circle hydrocephalus-stenosis-hydrocephalus.

The disjunction of ependymal cells under the effect of ventricular distension explains the classic transependymal resorption see LOVA concept: long standing open ventriculomegaly in adults; important congenital hydrocephalus found in adults, even with transependymal resorption, justified by an AS, with macrocephaly, cognitive troubles with subnormal QI, intracranial hypertension syndrome, spastic weakness of lower limbs with gait disturbance, loss of sphincters control (gatism).

2. compensated chronic hydrocephalus (symptomatic condition, with non-progressive hydrocephalus): average PIC ≤ 12 mm Hg, with A or B waves. The pressure is normal only in appearance, because above a low mean value there are pathological waves (A and / or B). The transependymal pressure gradient persists, maintaining or worsening ventricular distension.

The triad of HAKIM & ADAMS (1965) bringing together movement disorders, psycho-intellectual disorders and sphincter disorders, classic "normal pressure hydrocephalus". Bi or unisymptomatic forms (isolated motor or mental disorders, exceptionally sphincter disorders) are not rare. Bret-Chazal (18) appreciate that the terminology "normal

pressure" is ill suited, because the intracranial pressure in this condition is not always normal, see our case.

3. arrested chronic hydrocephalus (asymptomatic, inactive with non-progressive ventriculomegaly when the intracranial pressure (ICP) returns to normal, despite the remaining dilated ventricle): average PIC ≤ 12 mm Hg, without wave A or B, is a chronological stage in the natural history of a hydrocephalus not a constant situation, has applicability in all progressive, active hydrocephalus in adults, the evolution is in bursts, appear after shunt or endoscopic third ventriculostomy (ETV)

In AS the main studies (20-22) are:

-unenhanced brain scanner identifies a possible cause of hydrocephalus, the ventricular dilation, permit reproducible measurements such as the bifrontal index, highlight parenchymal abnormalities, peri-ventricular hypodensities, the degree of visibility of the subarachnoid spaces: dilated, normal, erased, see basal cisterns, Sylvian valleys. Brain scanner in dynamic may quantify hydrocephalus, evaluate hydrocephalus after treatment.

-cerebral MRI may precise topography of a possible obstruction; may show funnelling superiorly of Sylvius aqueduct, distinguish the extent of obstructive hydrocephalus of the lateral and third ventricles with the 4th ventricle not dilated, transependymal resorption and signal abnormalities in the subependymal zone, images of leukoencephalopathy reflecting vascular processes more distant from the ventricles, monitorise the hydrocephalus during treatment. Useful MRI protocols in AS are: sagittal T2 with the absence of flow-void signal intensity at the aqueductal level, obstructing web; on sagittal CISS and three-dimensional constructive interference in steady-state (3D-CISS): decreased aqueductal stroke volume; phase-contrast MR imaging: peak systolic velocity; cine cardiac-gated phase-contrast MRI: aqueductal CSF flow after aqueductoplasty with stenting (21)(22). In patients with AS and in controls, the apparent diffusion coefficient (ADC) in four regions in the centrum semiovale on midsagittal FIESTA or CISS image proof deep white matter ischemia (DWMI), increasing resistance to CSF outflow through the ECS of the brain and in late adulthood may contribute to the development of

symptoms in adult onset AS (9). Another useful studies are PET to prove cerebral functional activity and angiography: to demonstrate megadolico basilar trunk, aneurysm of the ampulla of Galen, or for the study of venous return circulation, intimately linked to the dynamics of CSF.

In AS surgical treatment (5)(23-25) is based on:

1.treatment of the cause: excision of the responsible tumor

2.ventriculo-peritoneal shunt with valve, as in my case, with possible shunt complications - infection, obstruction and overdrainage.

3.endoscopic third ventriculostomy (ETV) can be considered the best surgical procedure for obstructive hydrocephalus caused by AS, restoring physiological circulation of CSF; to patients with obstructive triventricular hydrocephalus, with increased intracranial pressure, translucent membranous stenosis or aqueduct obstruction, prestenotic dilatation of the aqueduct.

4.aqueductal plasty (AP) could be performed alone or as an adjunct to third ventriculostomy, with or without a silastic stent, with few indication today. This procedure it's a logic, but also a high risky procedure with important complications rate after the tectal plate dorsal and midbrain tegmentum ventral trauma: diplopia, dysconjugate eye movement, trochlear palsy.

5. the endoscopic trans-fourth ventricle - retrograde aqueductoplasty with stent placement especially in cases of supratentorial slit ventricles.

CONCLUSIONS

This case could be a plea to demonstrate that AS may generate not only a chronic noncommunicating hydrocephalus with a potential normal pressure, as seen later in adults and very rare in elderly people, but also active hydrocephalus installed after a posttraumatic subarachnoid hemorrhage.

The lack of anatomo-pathological confirmation of Sylvius aqueduct to explain AS, may generate speculations to explain active as like chronic noncommunicating hydrocephalus by gliosis or even by the presence of a small and slow growing periaqueductal microglioma.

An active patient survey should be mandatory.

ABBREVIATIONS

CSF - cerebro-spinal fluid

AS - aqueductal stenosis
 LIAS - late-onset idiopathic aqueductal stenosis
 ECS - extracellular space of the brain
 PIC - intracranial pressure
 AP - aqueductal plasty
 ETV - endoscopic third ventriculostomy
 RA - retrograde aqueductoplasty by the endoscopic trans-fourth ventricle, with stent placement
 VP - ventriculo-peritoneal shunt

FOUNDING AND CONFLICTS OF INTEREST

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