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Internal hydrocephalus, external hydrocephalus, and the syndrome of intracerebral cerebrospinal fluid entrapment: a challenge to current theories on the pathophysiology of communicating hydrocephalus

Ralph Rahme, Michel W. Bojanowski *

Division of Neurosurgery, Hôpital Notre-Dame du CHUM, University of Montreal, 1560 Sherbrooke E, Montreal, Quebec, Canada H2L 4M1

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SUMMARY

The natural history of external hydrocephalus (EH) in adults is often marked by conversion into internal hydrocephalus. We describe a complication of this conversion, the ICE (intracerebral CSF entrapment) phenomenon, and demonstrate that both EH and ICE represent a challenge to current theories on the pathophysiology of communicating hydrocephalus (CHC). We propose a new model for CHC where the pattern of CSF distribution is largely determined by the intrinsic compliance of each of the intracranial structures. In this model, failure of distal CSF absorption resulting in an excessive intracranial CSF volume is the common denominator and CSF diversion the common solution to all forms of CHC.

Introduction

It is now widely accepted that obstruction of the arachnoid villi leading to reduced cerebrospinal fluid (CSF) absorption as a mechanism for communicating hydrocephalus (CHC) is a rather obsolete concept that should be abandoned [1–4]. Instead, current theories on the pathophysiology of CHC generally fall into one of two major schools of thought: the bulk flow theory where extraventricular obstruction of CSF flow, usually at the basal cisterns, is thought to be the underlying mechanism for CHC [3,4], and the pulsation theory where decreased intracranial compliance leading to increased pulsatile stress on the ventricular walls is believed to be the cause of ventriculomegaly [1,2].

External hydrocephalus (EH) is a well-known complication of surgically managed aneurysmal subarachnoid hemorrhage (SAH) in adults [5–7]. It is believed to result from the combination of two phenomena: decreased absorption of CSF and surgical tearing of the arachnoid membrane which may act as a ball-valve device leading to the subdural trapping of CSF [6–8]. EH often evolves into internal hydrocephalus (IH) over a period of weeks, either spontaneously or following drainage [5–9], and CSF diversion remains the mainstay of treatment for this condition [5–8].

In this paper, we describe a new entity, the ICE (intracerebral CSF entrapment) phenomenon, a complication of rapid conversion from EH to IH in the setting of a disrupted ventricular wall and an adjacent hematoma resorption cavity. We demonstrate that both EH and ICE represent a challenge to current theories on the pathophysiology of CHC and propose a new model to explain our findings.

The ICE phenomenon: clinical case

A 65-year old right-handed female patient was referred for grade 4 SAH. The patient was found in her house lying on the floor, confused, and unable to move her left side. On exam, she was awake but disoriented to time and place, had an incoherent speech, and exhibited profound left-sided hemiparesis and hemineglect. Head CT revealed a large right frontotemporal hematoma with a 7 mm midline shift, along with diffuse SAH predominating in the right sylvian fissure where an underlying calcified aneurysm of the right MCA was suspected (Fig. 1A and B). Cerebral angiography confirmed the presence of a 10 mm aneurysm of the right MCA bifurcation (Fig. 1C and D). The patient was taken to the operating room where evacuation of the intracerebral hematoma and clipping of the aneurysm were performed through a right pterional craniotomy. Given the substantial cerebral edema, duraplasty was performed and the bone flap was not replaced. The patient had a favorable post-operative course with excellent intracranial pressure control and progressive neurological improvement. Unfortunately, her condition was complicated by an aspiration pneumonia necessitating prolonged intubation and a tracheotomy. However, she continued to improve neurologically and was able to recover substantial motor strength in her left side.

Three weeks following the hemorrhage, the patient developed progressive somnolence. On exam, her scalp flap was full to palpation. Head CT revealed the presence of an extra-axial fluid collection overlying the right cerebral convexity and herniating through the craniectomy defect (Fig. 2A). Although not markedly

* Corresponding author. Tel.: +1 514 890 8000x26809; fax: +1 514 412 7816.
E-mail addresses: rrahme@waln.org (R. Rahme), m_bojanowski@yahoo.com (M.W. Bojanowski).

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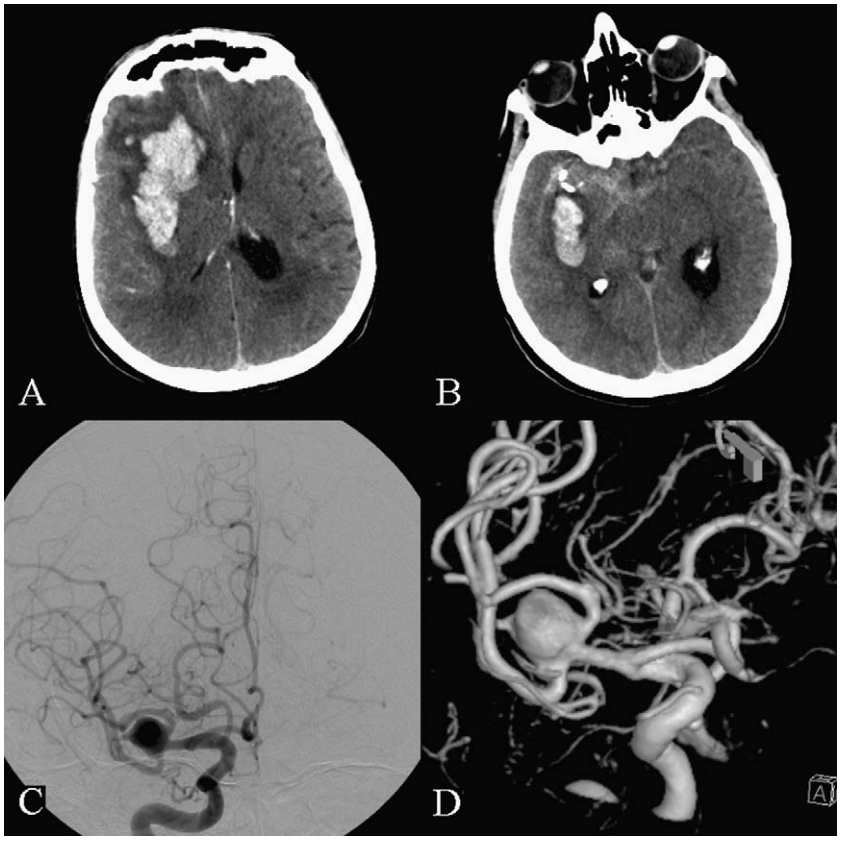

Fig. 1. (A and B) Head CT reveals a right frontotemporal hematoma with significant mass effect and subarachnoid hemorrhage predominating in the right sylvian fissure. An underlying calcified aneurysm of the right MCA is suspected. (C and D) Cerebral angiography demonstrates a 10-mm aneurysm at the right MCA bifurcation.

dilated, the ventricular system was felt to be plump (Fig. 2B and C). Therefore, the diagnosis of EH was made and it was decided to insert a ventriculoperitoneal shunt (VPS). However, the patient exhibited marked spontaneous improvement in her level of consciousness, which made us reconsider our decision and simply monitor her clinically. Unfortunately, four days later, the patient developed sudden neurological deterioration with several episodes of vomiting, severe lethargy, left hemiplegia, and a very tense skin flap. Head CT was urgently performed and demonstrated quasi-complete resolution of the extra-axial collection and the development of tetraventricular hydrocephalus. The intracerebral hematoma resorption cavity, which appeared to communicate with the frontal horn of the right lateral ventricle, was filled with CSF and distended, with marked overlying cerebral edema and herniation of the cerebral parenchyma through the craniectomy defect (Fig. 2D–F). These images suggested conversion from EH to IH and high-pressure ICE within the hematoma cavity with secondary CSF dissection into the cerebral parenchyma. The patient was taken emergently to the operating room where a VPS was inserted. The patient exhibited immediate post-operative neurological improvement with a quick return to her baseline status, and her skin flap became soft. Head CT showed resolution of ventriculomegaly and significant decrease in size of the hematoma cavity and surrounding cerebral edema (Fig. 2G–I). She eventually underwent cranioplasty and was transferred to a rehabilitation facility.

Failure of current theories to account for EH and ICE

In the bulk flow model of CHC, tetraventricular dilation is thought to result from obstruction of CSF flow distal to the fourth ventricle but proximal to the cortical subarachnoid spaces (CSAS), which may explain the effectiveness of endoscopic third ventriculostomy (ETV) in many of these cases [3,4]. However, such a model would be incompatible with the subdural trapping of CSF in EH as this would require substantial CSF flow from the ventricles into the CSAS where the arachnoid tear acting as a one-way valve is presumed to be present [6–8]. Moreover, redistribution of CSF from the subdural compartment into the ventricles and the hematoma cavity would not be expected with this model given the obstruction of the basal cisterns. Instead, it would be more logical to assume that the CSF exiting the subdural compartment would undergo reabsorption through the supposedly patent arachnoid villi into the dural venous sinuses.

In the pulsation model, ventricular dilation is believed to result from the transmission of pulsatile stress through the choroidal arteries to the ventricular walls as a result of decreased intracranial compliance caused by scarring in the subarachnoid spaces (SAS) [1,2]. In this model, ETV is thought to restore normal intracranial compliance by venting the third ventricle [1,2]. However, in EH, the ventricles often have a normal to slightly enlarged size and instead the arterial pulsation-free subdural compartment is markedly dilated. Also, the sudden redistribution of CSF into the ventricles and the hematoma cavity are hard to fit in this model.

Therefore, it is clear that excess CSF volume resulting from an imbalance between production and absorption exists in this condition and that this extra volume tends to move freely between the subdural compartment, SAS, ventricular system, and, in the case of ICE, hematoma resorption cavity and cerebral interstitium [5,10]. In this setting, the major determinant of the pattern of CSF distribution appears to be the intrinsic compliance of each of the intracranial compartments as CSF distribution tends to follow the paths of least resistance [5,7].

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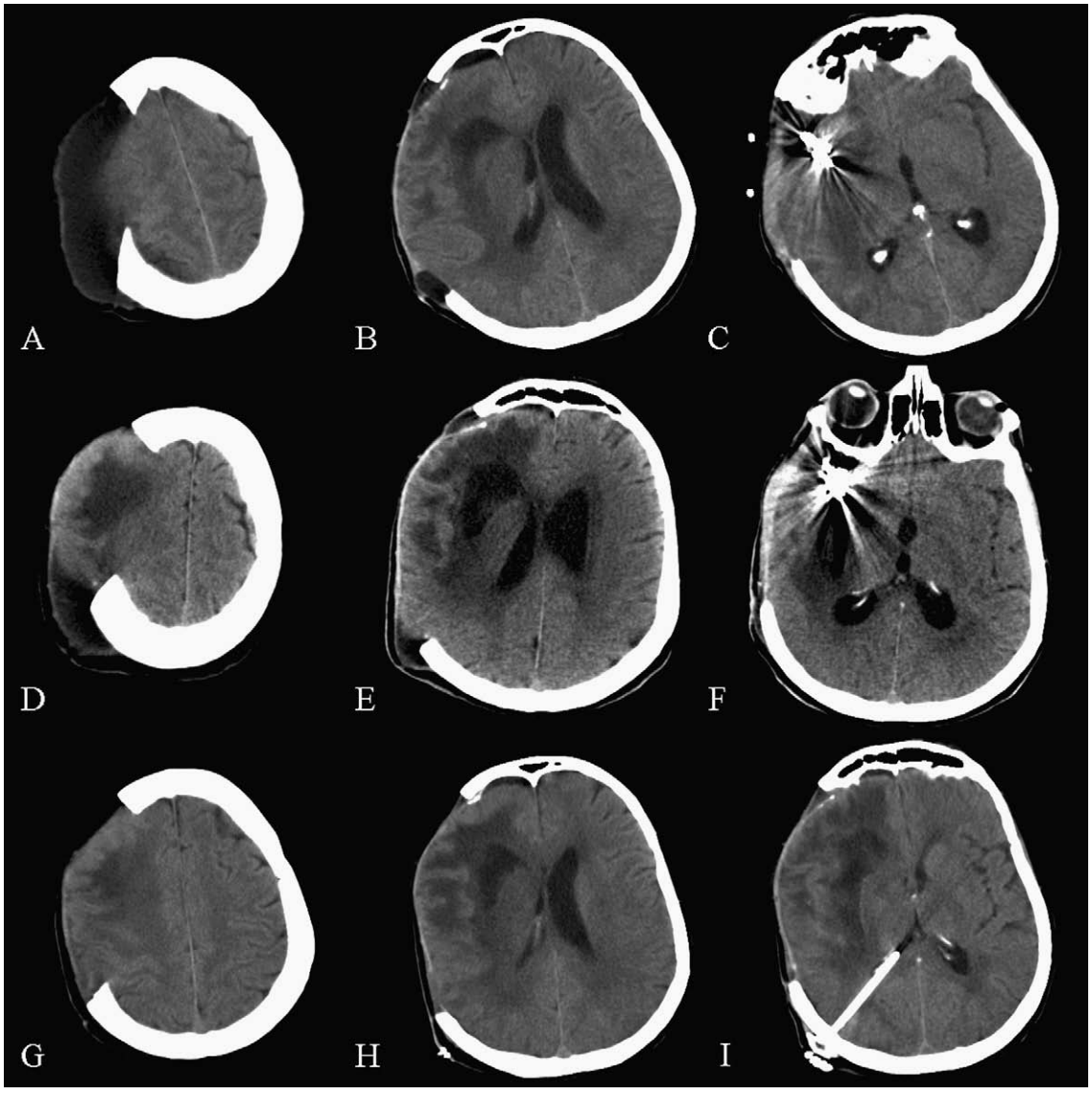

Fig. 2. (A-C) Head CT reveals an extra-axial fluid collection herniating through the craniectomy defect with a slightly dilated ventricular system. This is strongly suggestive of external hydrocephalus. (D-F) Head CT following neurological deterioration demonstrates conversion into internal hydrocephalus and a distended CSF-filled hematoma resorption cavity with marked overlying cerebral edema leading to herniation of the cerebral parenchyma through the craniectomy defect. This is the ICE phenomenon. (G-I) Head CT following CSF diversion shows marked decrease in the size of the ventricles and hematoma cavity with substantial reduction in the amount of cerebral edema.

The alternative hypothesis

We propose that, in the normal situation, the ventricles constitute the area of least resistance in the intracranial cavity. On one end, their ependymal lining appears to protect the periventricular white matter by resisting passage of excess CSF from the ventricles to the cerebral interstitium. In fact, it has been proposed that stretching and disruption of the tight junctions in the ependyma secondary to ventricular dilation in hydrocephalus may result in transependymal CSF flow and interstitial cerebral edema [3,10]. Similarly, transependymal flow of CSF has been implicated in the pathophysiology of pseudotumor cerebri, a condition whose treatment consists in CSF diversion [11]. Finally, ependymal rupture has been shown to result in spinal cord edema and syrinx formation in the setting of experimental hydrocephalus [12]. On the other end, the ventricular system appears to have a higher compliance compared to the CSAS. This may be due to the combination of two factors: the significantly smaller inner (ventricular) surface of the brain compared to the outer (cortical) surface which results in a higher hydrodynamic stress for the same amount of pressure [2], and the higher mechanical resistance of the cerebral cortex relative to the periventricular white matter, partly because of denser cellularity and higher vascularity.

In the presence of a disrupted ependymal lining and an adjacent hematoma resorption cavity, the latter freely communicates with the ventricles and becomes the path of least resistance to CSF flow. At least two factors may account for this phenomenon: first, the surface of the hematoma cavity is smaller and thus the hydrodynamic stress there is higher compared to the ventricular system, and second, this cavity is usually lined by a friable edematous white matter which is probably less resistant and more compliant than the ventricular ependyma. Consequently, CSF is trapped in this cavity as the ventricular system dilates and subsequently dissects into the surrounding cerebral interstitium, thus resulting in the ICE phenomenon. When this happens slowly, as may occur for instance in longstanding hydrocephalus and porencephaly, the cerebral tissue has time to adapt through compensatory mechanisms to the excessive CSF load. However, if ICE develops acutely,

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e.g. following rapid conversion from EH to IH as in this case, the brain's buffering capacity is quickly overcome and significant cerebral edema ensues. In the present case, rapid conversion from EH to IH resulted in acute ventricular dilation and CSF entrapment within the hematoma cavity with secondary cerebral edema leading to herniation of the right frontal lobe through the craniectomy defect. This resulted in severe neurological deterioration with focal deficits which were readily reversed by prompt CSF diversion.

What led to the quick conversion from EH to IH? We postulate that this is the result of a sudden rupture of the arachnoid membrane lining the subdural CSF collection. As EH progresses, this thin membrane is subjected to increasing tension until its capacity to resist is overcome, which leads to its rupture. This releases all the trapped CSF into the adjacent SAS from where it quickly redistributes into the more compliant ventricles and hematoma resorption cavity.

Conclusion

In this paper, we present evidence that challenges current theories on the pathophysiology of CHC. While we acknowledge that the bulk flow and pulsation models may offer an explanation to several aspects of CHC, particularly the high success rates of ETV, we believe that both models have significant shortcomings since none of them can explain the pathophysiology of EH, its conversion into IH, and the occurrence of the ICE phenomenon. We propose that CHC is not a single entity, but rather the common end result of several different disturbances affecting the normal intracranial circulation of CSF. Although the bulk flow and pulsation models may underlie many cases of CHC, thus accounting for the frequent success of ETV, failure of CSF flow and/or absorption distal to the CSAS, resulting in excessive amounts of intracranial CSF, still appears to play a role in the pathophysiology of at least some cases of CHC, particularly EH, and probably accounts for ETV failure in many such cases. In our model, the pattern of CSF distribution in the intracranial cavity and hence the subtype of hydrocephalus (EH, IH, pseudotumor cerebri, or ICE) are mainly determined by the intrinsic compliance of each of the intracranial structures, and CSF diversion remains the common solution for the various forms of CHC.

Conflicts of interest statement

None declared.

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