Medical Hypotheses 85 (2015) 958–961

Contents lists available at ScienceDirect

Medical Hypotheses

journal homepage: www.elsevier.com/locate/mehy

Pulse wave myelopathy: An update of an hypothesis highlighting the similarities between syringomyelia and normal pressure hydrocephalus

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ARTICLE INFO

Article history:
Received 11 May 2015
Accepted 26 August 2015

ABSTRACT

Most hypotheses trying to explain the pathophysiology of idiopathic syringomyelia involve mechanisms whereby CSF is pumped against a pressure gradient, from the subarachnoid space into the cord parenchyma. On review, these theories have universally failed to explain the disease process. A few papers have suggested that the syrinx fluid may originate from the cord capillary bed itself. However, in these papers, the fluid is said to accumulate due to impaired fluid drainage out of the cord. Again, there is little evidence to substantiate this. This proffered hypothesis looks at the problem from the perspective that syringomyelia and normal pressure hydrocephalus are almost identical in their manifestations but only differ in their site of effect within the neuraxis. It is suggested that the primary trigger for syringomyelia is a reduction in the compliance of the veins draining the spinal cord. This reduces the efficiency of the pulse wave dampening, occurring within the cord parenchyma, increasing arteriolar and capillary pulse pressure. The increased capillary pulse pressure opens the blood–spinal cord barrier due to a direct effect upon the wall integrity and interstitial fluid accumulates due to an increased secretion rate. An increase in arteriolar pulse pressure increases the kinetic energy within the cord parenchyma and this disrupts the cytoarchitecture allowing the fluid to accumulate into small cystic regions in the cord. With time the cystic regions coalesce to form one large cavity which continues to increase in size due to the ongoing interstitial fluid secretion and the hyperdynamic cord vasculature.

Introduction

Syringomyelia is the development of a tubular, fluid filled cavity, within the spinal cord. It is most commonly associated with an extramedullary lesion at the foramen magnum such as the tonsillar herniation of Chiari I malformation [1]. Other intradural causes include Dandy–Walker malformations, arachnoiditis secondary to meningitis, trauma or haemorrhage and compressive tumours such as a meningioma [1]. Intramedullary causes such as direct spinal cord trauma and intrinsic tumour are also found [2]. Most hydrodynamic theories for the formation of the syrinx invoke mechanisms whereby CSF from the subarachnoid space is pumped into the spinal cord parenchyma. In an extensive review by Elliott et al., the hydrodynamic theories suggesting CSF fluid accumulation in syrinx cavities is due to it being pumped against the pressure gradient from CSF to cord parenchyma, were all found to be wanting [3]. Greitz was the first to suggest that the fluid in a syrinx may be derived from the capillaries themselves but did not suggest a mechanism as to why the interstitial fluid production should be increased. The mechanism for the fluid accumulation that was put forward by Greitz was a blockage to the outflow of this fluid as it passed into the subarachnoid space [4].

A theory relating the cause of syringomyelia directly to a reduction in compliance and increased pulsation strength, analogous to normal pressure hydrocephalus (NPH), was developed in 2004 [5]. It was noted that both conditions share similar manifestations, i.e. both syringomyelia and NPH are associated with the accumulation of fluid within the centre of their respective neural structures, with displacement of the parenchyma into the subarachnoid space despite the lack of a well-defined pressure gradient [6]. It is also noted that both conditions have reduced compliance of the subarachnoid space surrounding their respectively affected neural tissue [7,8]. Since the initial hypothesis was published by the current author, further evidence has come to light regarding the effects of pulsation energy on the neural tissues. The purpose of

Abbreviations: CSF, cerebrospinal fluid; NPH, normal pressure hydrocephalus; mmHg, millimeters of mercury.

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http://dx.doi.org/10.1016/j.mehy.2015.08.028
0306-9877/

the current paper is to further develop the hypothesis that syringomyelia is a manifestation of pulse wave energy, leading to a form of pulse wave myelopathy.

The hypothesis

The hypothesis put forward is that the primary instigating factor in syringomyelia is a reduction in the compliance of the walls of the small veins draining the spinal cord. This reduces the efficiency of the windkessel mechanism. The windkessel mechanism dissipates the pulsation energy in the arteries supplying the spinal cord by transferring it into the blood flowing out within the small veins draining the cord, thereby bypassing the capillary bed. The resulting increase in pulse pressure within the capillaries will stress their walls leading, to disruption of the blood–spinal cord barrier and thus increase interstitial fluid secretion. An increase in the pulse pressure within the arterioles renders them hyperdynamic and this disrupts the background cytoarchitecture of the cord, allowing fluid to accumulate into small cystic spaces. With time, these spaces coalesce into one large cavity which expands into the subarachnoid space due to the continued accumulation of the interstitial fluid.

Evaluation of the hypothesis

Compliance in normal pressure hydrocephalus and syringomyelia

In NPH there is a generalised reduction in intracranial compliance which leads to an increase in the CSF pulse pressure which is twice normal [9]. This has a direct effect upon the venous compliance. The speed of the pulse waves within the vascular tree in NPH (i.e. the pulse wave velocity), is twice normal, indicating the compliance of the vessel walls is reduced by half [10]. Direct measurement of the pulsation strength within a vessel can be made using the pulsatility index or P.I. The sagittal sinus pulsation is increased from a P.I. of 0.4 in normal young people to 0.7 in NPH, but the cortical veins do not respond in the same way. The cortical vein pulsation decreases from 0.4 in normal young people to 0.3 in NPH (i.e. the cortical vein compliance is much lower than normal) [11]. These findings are similar to those noted in syringomyelia. In one study, the total intracranial compliance was reduced by 20% [12] and in another study, the total intracranial compliance was reduced by 28% in patients with Chiari I malformation with or without syringomyelia [13]. Successful posterior fossa decompression surgery was associated with a 54% increase in compliance in the later patients [13]. However, measuring intracranial compliance may underestimate the effect around the cord, as the spinal subarachnoid space compliance may be more important in this condition than intracranial compliance. In 20 patients with a Chiari I malformation and syringomyelia, the cervical compliance was reduced by 45% compared to controls and there was a 44% increase in CSF pulse pressure [7]. Compliance is also reduced in other forms of syringomyelia rather than just those associated with Chiari I malformation. In 36 patients with syringomyelia due to subarachnoid space block from arachnoid fibrosis, the mean cervical CSF pressure was normal but the pulse pressure was increased from 1.6 mmHg in controls to 2.7 mmHg. The cervical and lumbar subarachnoid compliance was reduced in the patients by 47% compared to controls. When the compliance could be improved surgically, the syrinx improved. If the compliance could not be improved the syrinx remained [14].

One may argue, that not all cases of syringomyelia may be associated with reduced compliance. The development of Chiari I malformation and syringomyelia has been recognised in patients with long term lumboperitoneal shunts [15]. Similarly, tonsillar ectopia and syringomyelia has been found to occur in patients with spontaneous intracranial hypotension secondary to CSF leak from meningeal diverticula at the nerve root sheaths [16]. In both of these later conditions the subarachnoid space pressure is reduced and the compliance of the dura is probably increased. However, the small veins within the spinal cord may not be similarly affected. The compliance of dog veins was measured under varying trans-mural pressures (the trans-mural pressure is the pressure gradient across the vessel wall). The peak compliance was found to occur at a trans-mural pressure of 4 mmHg, however, the compliance was reduced to 50% of the original if the trans-mural pressure were increased by only 1–5 mmHg [17]. Decreasing the CSF pressure around the small veins draining the spinal cord increases the trans-mural pressure across the walls of the veins and therefore may make them less compliant. Thus, reduced venous compliance is a common theme in both NPH and syringomyelia.

The windkessel effect

The classical teaching is that the blood flowing from the arterioles into the capillaries is completely dampened [18]. Therefore, pulsatile arterial flow is converted into non-pulsitile continuous flow before the capillaries. In the peripheral tissues, this dampening depends only upon the compliance of the arteries, which expand and contract, allowing the pulsation energy to be dissipated into the surrounding tissues. The dampening of fluid pulsations is known as the windkessel effect. Arterial dampening depends on expansion of the arteries in systole and contraction in diastole. The windkessel mechanism is more complex within the neuraxis. The CSF surrounding the arteries is incompressible. Therefore, the pulse energy stored in systole must be accommodated by the available compliance of the system i.e. either the walls of the container (the dura mater) must be shifted and/ or the veins passing through the cord or subarachnoid space must be compressed. A breakdown in the windkessel effect will direct greater pulsation energy into the arterial inflow of the neural structures and their capillary beds, possibly leading to parenchymal derangement [19]. In the spinal subarachnoid space, the pulsation energy of the spinal cord is as important as the brain pulsation energy, with regards to the origin of the spinal canal pulse pressure. In one study, the cervical subarachnoid pulsation was equally attributable to the carotid territory (i.e. the brain) and the thoracic aorta (i.e. the spinal cord) [20]. Similarly, the lumbar CSF pulsatility has been found to be predominantly due to local pulsations and not directed from above [21]. Therefore, the cord appears to utilise its surrounding subarachnoid space compliance, at each level, to dampen its pulsations. Failure of this effect must have consequences.

Breakdown of the blood–brain and blood–spinal cord barrier

It has already been noted that an increase in pulse pressure in the subarachnoid space occurs in both NPH and syringomyelia. This is a manifestation of a breakdown of the windkessel effect. This loss of the windkessel effect has been associated with a breakdown in the blood–brain barrier in hydrocephalus. In human infantile hydrocephalus, the capillary wall shows blood–brain barrier dysfunction with increased vesicular and vacuolar transport, open inter-endothelial junctions, thin and fragmented basement membranes and discontinuous perivascular astrocytic end feet. The findings suggested an inter-endothelial route either for hydrocephalic oedema formation or resolution [22]. Others have also found a significant blood–brain barrier breakdown in adults with NPH as well [23]. Many other forms of senile dementia also have a loss of the blood–brain barrier. There is evidence of blood–brain barrier breakdown in Alzheimer/s disease and vascular dementia