Definitions

cerebellar tonsils - portion of the cerebellum located at the bottom, so named because of their shape

cerebellum - part of the brain located at the bottom of the skull, near the opening to the spinal area; important for muscle control, movement, and balance

cerebrospinal fluid (CSF) - clear liquid which surrounds, and protects, the brain and spinal cord

cervical - the upper part of the spine; the neck area

Chiari malformation - condition where the cerebellar tonsils are displaced out of the skull area into the spinal area, causing compression of brain tissue and disruption of CSF flow

Dandy-Walker - congenital malformation which involves an enlarged 4th ventricle, an abnormal cerebellum, and a cyst in the skull base region

decompression surgery - common term for any of several variations of a surgical procedure to alleviate a Chiari malformation

laminectomy - surgical removal of part (the bony arch) of one or more vertebrae

lumbo-peritoneal shunt - a tube, surgically implanted, which drains CSF from the lumbar region of the spine into a space in the abdomen

magnetic resonance imaging (MRI) - diagnostic test which uses a large magnet to create images of internal body parts

Marfan's Syndrome - hereditary condition which affects the connective tissue in the body

pathogenesis - the underlying cause, or mechanism, of a disease

pseudotumor cerebri: also known as intracranial hypertension; chronically high

The Many Faces Of Chiari

Case Studies is a feature designed to highlight interesting patient cases reported in the research. Given the lack of knowledge about CM/SM, much of the published research comes in the form of case studies - doctors describing one or two patients they have seen and treated - as opposed to rigorous scientific studies. While this type of publication doesn't advance the scientific cause as much, it does give us a window into some of the issues surrounding CM/SM, including lasting side effects and related conditions. And hopefully, some of our readers will say, "Hey, that's just like me!" and know they are not alone in what they are going through.

Syringomyelia with Chiari Malformation: 3 unusual cases with implications for pathogenesis

Authors: B.K. Owler, G.M Halmagyi, J. Brennan, M. Besser
University/Hospital: Royal Prince Alfred Hospital, University of Sydney, Australia
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Introduction: Dr. Owler and his colleagues reported on 3 cases of Chiari and syringomyelia, each arising from different circumstances. In addition to providing clues as to the mechanism underlying syrinx formation, these cases also highlight the variety of ways in which Chiari and SM can be found to occur.

Patient 1: The first patient was a 27 year old woman who had been suffering from headaches, neck pain, dizziness, and nausea for 5 weeks. The interesting aspect of her symptoms was that they would only occur when she sat up, and would go away when she was prone. Her medical history was complicated because she had Marfan's syndrome - which affects the connective tissue in the body and can cause problems with growth, bones, skin, nerves, etc. A neurological exam showed no problems with her reflexes, sensations, etc.; however, an MRI showed she had a Chiari malformation, a syrinx from C2-T7, and there was evidence she had low CSF pressure and the doctors suspected she had a CSF leak somewhere.

Treatment: Because of her other health problems, the doctors decided to treat her conservatively and she was confined to bed rest.

Outcome: Her symptoms slowly went away and an MRI four months later showed that the syrinx had collapsed. There was no change in the position of the cerebellar tonsils. The authors believe her CSF leak healed, reversing the syrinx.

Patient 2: Patient 2 was a 42 year old woman who had been diagnosed with pseudotumor cerebri 13 years earlier, and for which she had had a lumboperitoneal shunt inserted. Recently, for four months, she had been suffering from progressively worse balance problems. A neurological exam revealed abnormal reflexes, sensations, muscle weakness, and other problems. An MRI showed a 10mm Chiari plus a large syrinx extending from C1-T11.

Treatment: The doctors chose to treat the Chiari and performed a decompression including a laminectomy and duraplasty.

Outcome: Her symptoms improved and 4 months after surgery the doctors shut off the shunt to see if she still needed it. It became clear she did, so the shunt was turned back on. A follow-up MRI showed that the syrinx had mostly collapsed, although the cerebellar tonsils were still descended. The authors point out that acquired Chiari is a known risk of lumboperitoneal shunts. If the shunts drain too much CSF, the pressure in the spine becomes too low compared to the pressure in the skull and the cerebellar tonsils are forced out of position.

Patient 3: 29 year old woman suffering from occasional headaches and numbness and abnormal sensations in her hands. The symptoms began after she went through labor and delivery. Her neurological exam was normal; however, an MRI revealed she had Dandy-Walker, with a cyst protruding into the cervical spinal canal. She also had Chiari and a syrinx.

Treatment: The doctors performed surgery to decompress the Chiari and treat the Dandy-Walker cyst.

Outcome: Six months after surgery, her initial symptoms were gone and her syrinx had collapsed completely.

Author's Discussion: Owler and his colleagues used these cases (the first two anyway) as a basis for discussing one of the current theories on syrinx formation, namely the piston theory. The piston theory was

CSF pressure

syringomyelia (SM) - neurological condition where a fluid filled cyst forms in the spinal cord

syrinx - fluid filled cyst in the spinal cord

vertebra - segment of the spinal column (see Spinal Anatomy)

developed by researchers at the National Institutes of Health, and holds that with every heartbeat, the cerebellar tonsils are driven down into the spinal area, like a piston. This in turn creates a pressure wave in the CSF and CSF is forced into the spinal cord - through small spaces on the outside of veins and arteries - to form a syrinx. Owler points out that the first two cases of acquired Chiari resulted from a CSF leak and a shunt which was draining too much fluid. Both of these cases would result in less fluid outside the spinal cord, meaning a the pressure outside of the cord would be lower. If the pressure is lower outside the cord, it is not clear how a syrinx would form. Although Owler does not make a strong statement, the piston theory does not seem able to explain why a syrinx would develop for Patients 1 and 2.

Editor's Discussion: I found these cases to be very educational because they encompass so many different aspects of CM/SM. First, as mentioned previously, they highlight the great variety of circumstances that can surround CM/SM. Second, they clearly demonstrate not only acquired Chiari, but acquired Chiari which leads to a syrinx. Third, Patient 3 highlights a phenomena which I have heard anecdotally several times, that the prolonged strain of giving birth can be a triggering event for symptoms. Finally, they highlight the somewhat severe limitations of the prevailing theories regarding syrinx formation. While in general I don't believe case reports are as valuable as structured research, they can be a valuable tool to test existing theories and develop ideas for new ones.