Mr. B is a 51-year-old male. He is a non-smoker and occasional drinker. He is a farmer and has no significant medical history except strong family history of strokes, Alzheimer’s disease, and cardiac dysrhythmias.

Mr. B developed bilateral hearing loss while he was working with an air compressor in May 2013. His symptoms resolved within a day but re-occurred in June 2013, which was also resolve within 24 hours. However, at this time, his hearing problem was unable to resolve completely. Mr. B was seen by an ear, nose, and throat (ENT) specialist and neurologist and was suggested may have intracranial hypotension. Therefore, Mr. B was referred to a neurosurgeon.

On examination, Mr. B had a normal neurological assessment and without any cerebellar signs (ataxia, tremor, and nystagmus). He has no headaches, nausea, vomiting, diplopia, dizziness, ataxia, or weakness. A magnetic resonance imaging (MRI) was done and it showed a Chiari I malformation. He was referred to neurosurgeon for possible surgical treatment. A pre-operative MRI (see below picture) showed Chiari 1 malformation with a 9 mm inferior ectopia of the cerebellar tonsils and moderate crowding of the foramen magnum. However, no evidence of intracranial hypotension was seen in MRI.

Even Mr. B had no signs and symptoms of Chiari malformation, however, clinical researches indicated if it left untreated, it may lead to problems in the future such as hydrocephalus, decrease in neurological function, and/or intracranial hypertension. Mr. B stated he had formal hearing test conducted before, which showed some impairment of hearing bilaterally. He stated he was aware that surgery may not improve his hearing problem but may prevent complications from the Chiari malformation. He agreed and consented for surgery. A Chiari decompression, duroplasty, and resection of the posterior C1 arch were performed on May 2014.

During surgery, a suboccipital craniotomy with C-1 laminectomy (or small section of skull) is performed to expose the dura, the dura is then opened, the damaged cerebellar tonsils are removed, and a patch (synthetic or a piece of pericranium) is sutured to the dura incision. This patch is to enlarge the space around the cerebellar tonsil and prevent cerebellar ptosis (sagging). Duraseal sealant may be used on the duroplasty site to prevent cerebrospinal fluid (CSF) leak. Lastly, the neck muscle and skin are closed by sutures (Cheng, Nash, & Meyer, 2002; Parker et al., 2013).

Post-operative orders include analgesia (ketorolac, morphine, and oxycodone) for pain control; heparin and sequential compression devices for deep venous thrombosis prophylaxis, bowel routine, antiemetic, and routine neurological and vital signs monitoring.

Post-operative MRI showed changes within the suboccipital soft tissues with packing material.
On post-operative day 1, all neurological signs and vital signs were stable. Prophylaxis antibiotic, analgesia, and bowel routine were ordered. Mr. B complained of headaches but was well controlled with analgesia.

An occupational therapist and physiotherapist were consulted for post-operative activities and possible rehabilitation treatment. It was suggested that no driving for 4-6 weeks, avoiding any stress such as heavy lifting (> 5 lb), bending, straining, and coughing, and gradual increases in normal activities such as walking.

Mr. B was discharged on post-operative day 2. He was instructed to follow up with neurosurgeon in 6 weeks. However, if there were any sign of leakage of CSF such as swelling around the incision site or fluid leaking from the incision site; signs of infection such as increased temperature, redness, and pain on the incision site; or any new neurological sign such as decreased alertness, changes in motor and/or sensory functions, he should seek medical advice immediately.

Case Highlight
Mr. B was presented with hearing lost and a Chiari malformation was discovered incidentally. Common manifestations for Chiari malformation include increase intracranial pressure, headache, altered cerebellar, cranial nerve, brain stem, and/or spinal cord functions. However, 30% of patients with tonsillar herniation of 5 to 10 mm are asymptomatic (Elster & Chan, 1992).

According to Mr. B’s statement, he had history of hearing impairment. Therefore, the cause of hearing lost may not be related to the Chiari malformation. However, Mr. B decided to have the surgery done due to the potential complication from Chiari malformation. Mr. B has an uneventful post-operative course and was discharged home on post-operation day 2.

Reference

Post reading quiz
1) Chiari malformation is the herniation of which part of the brain?
   a) Cerebrum
   b) Cerebellum
   c) Brain stem
   d) Spinal cord

2) Chiari malformation is usually present with which type of signs and symptoms?
   a) Headache, ataxia, weakness
   b) Unsteady gait, diplopia, nausea
   c) Hearing loss, ataxia, vomiting
   d) Dizziness, vertigo, headache

3) What are the common complications of Chiari malformation?
   a) Subdural hematoma, cerebral herniation
   b) Epilepsy, increase intracranial pressure
   c) Hydrocephalus, intracranial hypertension
   d) Subarachnoid hemorrhage, hydrocephalus

4) What is the rationale of avoid straining after surgery?
   a) Prevent increase intracranial pressure
   b) Alleviate pain on the surgery site
   c) Decrease risk of wound bleeding
   d) Improve cerebrospinal fluid flow

5) Herniation of cerebellar tonsil may obstruct CSF flow, which part of the CSF circulation pathway may be affected?
   a) Lateral ventricle
   b) 3rd ventricle
   c) Cerebral aqueduct
   d) 4th ventricle

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