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CASE REPORT

Acquired Chiari I malformation following a lumbo-peritoneal shunt– William's hypothesis revisited

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Abstract

Background: Syrinx formation can occur in congenital conditions like Chiari malformation, split cord malformation and also in acquired conditions like trauma, infections and malignancy. Several hypotheses have been put forward to explain the pathogenesis of syrinx formation; William's pressure dissociation hypothesis is one amongst them. We report a 55 year old female who developed Chiari I malformation with syrinx formation, two months after lumbo-peritoneal shunt for communicating hydrocephalus as an evidence to support William's hypothesis.

Case report: A 55 year old female with no comorbidities was operated two years ago for a left frontotemporal fungal granuloma. She was asymptomatic until two months back when she developed headache and difficulty in walking. CSF analysis was normal and CT head showed communicating hydrocephalous. A Lumbo-peritoneal shunt was performed and patient improved symptomatically following surgery. Two months into the follow up period, she had recurrence of symptoms. MRI brain with CV junction showed communicating hydrocephalus with Chiari I malformation and syrinx formation. Patient underwent a right ventriculo-peritoneal shunt with ligation of lumbo-peritoneal shunt following which she was relieved of her symptoms. A follow up imaging showed resolution of hydrocephalus and obliteration of syrinx.

Conclusion: Acquired Chiari I malformation with syrinx formation after lumbo-peritoneal shunt and resolution of the same after ligation of shunt tube supports William's hypothesis.

Keywords: Chiari; syrinx; shunt

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Introduction

Lumbo-peritoneal shunt is one of the treatment options for communicating hydrocephalus. Being an extracranial procedure, it has a lower complication rate. Wound hematoma, shunt obstruction and shunt infection are known complications following this procedure. Acquired Chiari I malformation following lumbo-peritoneal shunt is a rare complication which is being detected with increasing frequency because of the wide spread availability of imaging techniques. This case illustration provides evidence in support

of William's pressure dissociation hypothesis as the pathogenesis behind acquired Chiari I malformation and syrinx formation following lumbo-peritoneal shunt.

Case report

A 55 year old female was operated two years ago for fungal granuloma in the left frontotemporal region. She received antifungal treatment post operatively and remained asymptomaticuntil two months back when she experienced headache, nausea and difficulty in walking of one week duration.She denied history of fever, visual blurring and loss of consciousness.

On examination, patient was conscious, coherent with stable vital parameters. Neurological examination did not reveal any focal deficits. CSF analysis showed sugar levels of 27 mg, protein 316 mg and Indian ink stain for cryptococcus was negative. CT brain showed communicating hydrocephalus. (Figure A and B) A lumbo-peritoneal shunt was performed and patient improvedsymptomatically after surgery.

She had recurrence of symptoms two months after the CSF diversion procedure and clinical examination did not reveal any deficits. MRI brain with CV junction showed communicating hydrocephaluswith Chiari I malformationand syrinx formation (Figure C). Patient underwent ligation of lumbo-peritoneal shunt and placement of aright ventriculo-peritoneal shunt which relieved her symptoms. A repeat MR imaging performed three months later showed resolution of hydrocephalus and obliteration of syrinx (Figure D).

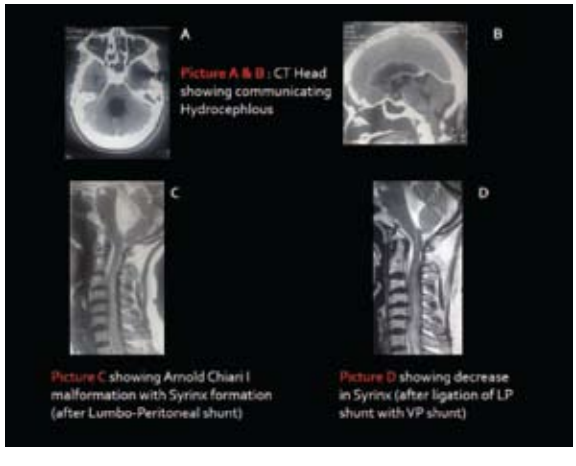

Figure: A, B, C and D

Discussion

The incidence of asymptomatic Chiari I malformation in normal population is 14%, but its incidence may be more in patients having pseudotumorcerebri, communicating hydrocephalus and CSF leaks [1]. A CT based study by Chumas et al found a 70% incidence of acquired Chiari I malformation after a lumbo-peritoneal shunt, with an associated syrinx in 50-70% [2]. Payner et al followed up 10 children with lumbo-peritoneal shunt; fourchildren developed symptomatic acquired Chiari I malformation after 0.75 to 4.5 years and three of them developed asymptomatic acquired Chiari I malformation after 1.5 to 4.5 years [3]. There are no definitive studies regarding true incidence of acquired Chiari I malformation, but studies comprising of less number of patients showed an incidence of 4.3% for symptomatic and 11.4% for asymptomatic acquired Chiari I malformation with a 1.4% incidence of syrinx formation [4].

Lumbo-peritoneal shunt is a preferred CSF diversion procedure for communicating hydrocephalus. Being an extracranial procedure, it has a low complication rate, although shunt infection, shunt malfunction and CSF overdrainage are known to occur. Till 1990s, only few cases of acquired Chiari I malformation with syrinx formation following lumbo-peritoneal shunt have been documented in literature.

There are two hypotheses explaining acquired Chiari malformation following alumbo-peritoneal shunt. The "cephalocranialdisproportion"hypothes is pertaining to the pediatric age group states that arrest of growth of the skull and a compensatory increase in growth of brain parenchyma after ventricular decompression leads to descent of tonsils below the level of foramen magnum.[5,6] This theory was not widely accepted because there have beenno cases of acquired Chiari I malformation after placement of a shunt tube into the ventricles. The "cranio-spinal pressure gradient" theory states that tonsillar descent occurs as a result of pressure gradient between cranium and spinal regions after alumbo peritoneal shunt. Drainage of CSF to peritoneum leading to low pressure in lumbarcanal anda decreased CSF flow across the foramen magnum increases the intracranial pressure which favors tonsillar descent [7].

In both the hypotheses discussed earlier, descent of tonsils causes obstruction to circulation of CSF across the foramen magnum. According to William's pressure dissociation hypothesis, sudden increase of subarachnoid pressure during coughing, sneezing or Valsalva maneuvers promotes tonsillar descent. The tonsils act as a "one way valve" favouring the passage of CSF from the fourth ventricle into the spinal canal but not the reverse because the increase in intracranial pressure persisted even after the spinal subarachnoid pressure had returned to normal [8].

Levine also hypothesised that in the presence of subarachnoid obstruction at the foramen magnum, a variety of activities, such as assuming the erect posture, coughing or straining, and pulsatile fluctuations of CSF pressure during the cardiac cycle, produce transiently higher CSF pressure above the block than below it. There are corresponding changes in transmural venous and capillary pressure favouring dilation of vessels below the block and collapse of vessels above the block. The spatially uneven change of vessel caliber produces mechanical stress on the spinal cord, particularly caudal to the block. The mechanical stress, coupled with venous and capillary dilation, partially disrupt the blood-spinal cord barrier, allowing ultrafiltration of crystalloids and accumulation of a protein-poor fluid in the syrinx cavity [9].

The pathogenesis of acquired Chiari I malformation with syrinx formation in our patient can be explained from the discussions above. Ligation of lumbo-peritoneal shunt and placement of a ventriculoperitoneal shunt would have restored the cranio-spinal pressure gradient to its physiological levels and eliminated the transmural venous and capillary pressure differences leading to resorption of syrinx. However, foramen magnum decompression may be needed in patients with persistence of symptoms, as was observed in some case reports.

Conclusion

Acquired Chiari I malformation with or without syrinx formation following alumbo-peritoneal shuntsupports William's hypothesis. Although most of patients remain asymptomatic, ligation of lumbo-peritoneal shunt with placement of ventriculoperitoneal shunt needs to be done in symptomatic cases. Those with persistent symptoms despite shunt ligation may benefit from foramen magnum decompression.

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