

## Sudden unexpected death in a young adult with Chiari I malformation

Madam, Chiari I malformation is a dysraphic congenital disorder, frequently associated with other malformations of the same kind, including syringomyelia, Klippel-Feil syndrome and tethered cord.<sup>1,2</sup> Chiari I malformation can be a cause of sudden unexpected death in a group of patients.<sup>3</sup> We report an extremely rare case of Chiari I malformation who developed posterior fossa pneumocephalus after surgery resulting in fatal outcome. Thirty year gentleman presented with loss of sensation over right half of body since 1 month, headache and giddiness of same duration. There was no history of loss of consciousness or trauma. General and systemic examination was normal. Higher mental functions were normal. Neurological examination revealed paralysis of the 9th, 10th and 12th cranial nerves; distal paresis, muscular atrophy and areflexia of the upper limbs; abolition of left abdominal cutaneous reflexes and hyper-reflexia in the lower limbs. Sensory examination revealed loss of position and vibration sense in lower limbs and loss of pain and temperature over upper chest. Laboratory tests and the chest radiograph were normal. X-ray cervical spine was normal. Magnetic resonance imaging (MRI) (figure-1A) showed a

large tonsillar herniation (Chiari I malformation), with a syringomyelic cavity extending from cervical cord to the T 8 vertebra. He underwent suboccipital craniectomy (foramen magnum decompression), C1 arch excision, C2 laminectomy and duroplasty. He was apparently alright in immediate post-operative period. On third day he developed slowly progressive headache followed by deterioration in sensorium. There was no fever or meningeal signs. On

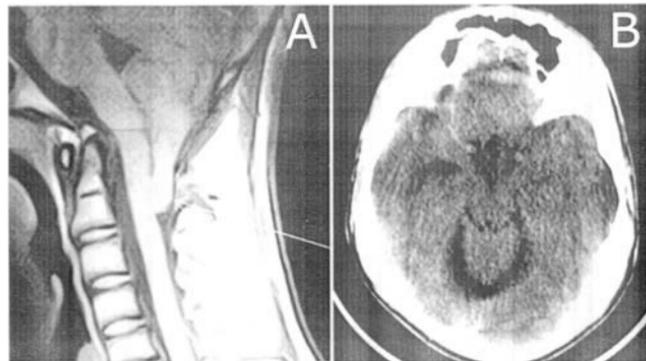


Figure-1. (A) Sagittal craniocervical magnetic resonance imaging (MRI) section T1W image showing herniated cerebellar tonsils (up to C2 level) with an extensive syringomyelia extended from the lower medulla to the level of the T8 vertebra (not seen in the figure) and (B). Post-operative CT scan showing pneumocephalus in the posterior fossa

repeat CT scan there was no evidence of hydrocephalus but it showed pneumocephalus in the posterior fossa with compression of surrounding structures (Figure-1B). He suddenly deteriorated and developed cardio-respiratory arrest with fatal outcome. Tension pneumocephalus after posterior fossa surgery though uncommon but is a well known complication.<sup>4,5</sup> The literature shows that air may enter or diffuse through multiple eroded or thinned bone areas during surgery.<sup>3</sup> It may manifest as deterioration of consciousness with or without localizing signs, severe restlessness, generalized convulsions or focal neurological deficits.<sup>4</sup> Posterior fossa pneumocephalus can be a cause of sudden cardiac arrest in immediate post-operative period.<sup>4</sup> Lethal brainstem compression may accompany even relatively minor trauma associated with chronic cerebellar tonsillar herniation in patients with Chiari I malformation.<sup>3,4</sup> In the present case pneumocephalus aggravated already compromised brain stem dysfunction and resulted in fatal outcome.

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### References

1. Paul KS, Lye RH, Strang FA, Dutton AJ. Arnold-Chiari malformation: review of 71 cases. *J Neurosurg* 1983; 58: 183-187.
2. Dyste GN, Menezes AH, Van Gilder JC. Symptomatic Chiari malformations: an analysis of presentation, management and long-term outcome. *J Neurosurg* 1989; 71:159-168.
3. Rickert CH, Grabellus F, Varchmin-Schultheiss K, Stoss H, Paulus W. Sudden unexpected death in young adults with chronic hydrocephalus. *Int J Legal Med* 2001; 114: 331-7.
4. Dash HH, Bithal PK, Muley, Vishnoi N, Saini SS, Tandon PN. Tension pneumocephalus following posterior fossa surgery in sitting position. *J anaesth Clin Pharmacol* 1990;6;207-12
5. Hernandez-Palazon J, Martinez-Lage JF, de la Rosa-Carrillo VN, Tortosa JA, Lopez F, Poza M. Anesthetic technique and development of pneumocephalus after posterior fossa surgery in the sitting position. *Neurocirugia (Astur)* 2003; 14: 216-21.

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## Announcement

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