Acute respiratory failure after endoscopic third ventriculostomy: A case report and review of the literature

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ABSTRACT

Endoscopic third ventriculostomy (ETV) is a relatively safe procedure. However, postoperative acute respiratory failure may be fatal. The authors report an 8-month-old patient with obstructive hydrocephalus secondary to posterior fossa cyst, and Chiari malformation. After ETV he developed difficulty in breathing, and had to be reintubated and ventilated. The infant recovered fully after craniocervical decompression and insertion of cystoperitoneal shunt. We speculate that respiratory failure is related to relative expansion of the posterior fossa arachnoid cyst, causing significant compression on the brain stem. Supportive care with mechanical ventilation and brain stem decompression were the mainstay of treatment.

Key words: Endoscopic third ventriculostomy, hydrocephalus, respiratory failure

INTRODUCTION

Endoscopic third ventriculostomy (ETV) for the treatment of obstructive hydrocephalus is an alternative to cerebrospinal fluid (CSF) shunting that may allow patients to avoid shunt implantation. The opening made in the floor of the third ventricle allows CSF to flow out of the ventricle to the subarachnoid space bypassing the obstruction.[1]

Mid line posterior fossa arachnoid cysts causing hydrocephalus are rather uncommon, representing approximately 2 - 3% of all intracranial cysts. They usually push the cerebellar vermis anteriorly, while separating the two cerebellar hemispheres. The most frequent clinical manifestations of these cysts in infants are macrocrania and symptoms of intracranial hypertension due to hydrocephalus.[2] Chiari malformation and syringohydromyelia have also been associated with these cysts.[3,4]

Neuroendoscopy has been recently introduced into the management of posterior fossa cysts, and successful endoscopic cyst marsupialization has been reported in limited series of patients.[5,6]

Primary complications of ETV are fever, bleeding and syndrome of inappropriate antidiuretic hormone due to damage to the hypothalamic structures.[1] A variety of serious complications have also been reported including herniation syndrome, arrhythmia, injury to cranial nerves and major vessels, and severe uncontrollable tachypnea.[7-11]

The authors report a case of acute respiratory failure following ETV for hydrocephalus associated with posterior fossa arachnoid cyst and Chiari malformation.

CASE REPORT

An 8-month-old infant, weighing 9 kg, a product of uneventful pregnancy and normal delivery presented with enlarging head and sunset appearance of the eyes. His development was within normal but has difficulty in turning due to heavy head. His head circumference was above the 97th percentile, and the anterior fontanelle was wide, full and slightly tense. He had upward gaze palsy and the rest of neurological examination was otherwise normal. Radiological investigations showed obstructive hydrocephalus caused by a large posterior fossa arachnoid cyst, distorting and occluding the fourth ventricle outlet; and associated with downward herniation of the cerebellar tonsils through the foramen magnum [Figure 1].
He underwent ETV through the right angle of the anterior fontanelle successfully. On an attempt to fenestrate the posterior fossa cyst through the lateral ventricle, he bled and the procedure had to be abandoned leaving an EVD in the lateral ventricle, which was kept opened at a level of 10-cm height from the patient’s head. Anesthesia was induced with 7-8% sevoflurane in oxygen. Ventilation with face mask was found to be easy. Fentanyl 10 microgram (mcg) and Cisatracurium 1 mg were given intravenously (i.v.) to facilitate endotracheal (ET) intubation. First attempt was with 3.5-mm internal diameter noncuffed polyvinyl chloride (PVC) ET tube was not successful as the anesthetist found it to be grade 3 intubation (Cormack and Lehane Classification). The second anesthetist inserted the same ET tube after repositioning the shoulder roll resulting in improved glottic view. There was little leak of air around the ET tube on gentle intermittent positive pressure ventilation (IPPV). After intubation, 125 mg paracetamol suppository was inserted. Anesthesia was maintained with 2-3% sevoflurane in 50% oxygen air mixture and patient received an additional bolus of fentanyl in the dose of 5 mcg. Anesthesia course was uneventful except slight drop in body temperature to 34°C despite of the use of fluid and body warmers. After completion of the surgery, the patient was warmed to 36.5°C and inhalational anesthetic terminated. Residual neuromuscular block was reversed with mixture of neostigmine and atropine. Trachea of the patient was extubated when the patient was fully awake after attaining adequate respiration and shifted to postanesthesia care unit (PACU). During first hour of stay in PACU, the patient’s vital were stable and no additional analgesic was needed. Before shifting the patient to ward, oxygen face mask was removed to assess breathing on room air, but oxygen saturation by pulse oximeter (SPO2) dropped to 90% and mild inspiratory stridor was noted. There were no signs of respiratory distress at that stage. The SPO2 improved to 98% when oxygen face mask with 5 liters/min flow was applied. The cause of inspiratory stridor was not obvious and intubation trauma secondary to two attempts at intubation was considered to be a possibility. There was no change in the vital signs or in the severity of stridor. It was decided to shift him to the pediatric intensive care unit (PICU) for observation and further management as it was not safe to send this patient to the ward. While waiting for PICU admission, the patient received 2.5 mg dexamethasone was given i.v. followed by 2.25% racemic epinephrine nebulization in the dose of 0.25 ml in 3-ml normal saline. It resulted in rise in heart rate from 125 to 150 beats/min; however, other vital signs were unchanged during this period. After 45 minutes of nebulization the patient rapidly developed difficulty in breathing and SPO2 dropped to 80%, which improved to 95% when IPPV and PEEP were applied through anesthesia face mask attached to Ayre’s T-piece circuit connected with 100% oxygen with 8 liters/min flow. The trachea of the patient was reintubated with same size ET tube after administering 10 mcg fentanyl i.v. After ET intubation, the patient developed generalized tonic-clonic seizure, which was controlled with 0.5 mg Midazolam i.v. Another episode of seizure recurred after 10 minutes and controlled with an additional dose of 1 mg midazolam i.v. An emergency CT scan was done which showed marginally smaller lateral ventricles with extra-axial fluid collection, and crowded foramen magnum. There was some blood in the occipital horns and tip of the EVD was in the lateral ventricle [Figure 2]. The patient was then shifted to PICU and connected to mechanical ventilation.

He maintained his vital signs and was stable enough next morning to arrange an MRI scan, which revealed reduction in the size of the ventricles, wide supratentorial subdural space, along with marginal increase in the posterior fossa.
cyst and anterior displacement and compression of the brain stem against the clivus. The brain stem was kinked and compressed, and the foramen magnum was crowded by the cerebellar tonsils compromising the medulla [Figure 3].

He was taken back to theater for craniocervical decompression; suboccipital craniectomy and removal of the posterior arch of Atlas, followed by durotomy and Gor-tex duroplasty, and insertion of cystoperitoneal shunt. Postoperatively, he recovered and his breathing improved dramatically that he could be weaned successfully off the ventilator and discharged of the hospital in a week’s time.

He was seen in the outpatient clinic after 1 month, his parents reported improvement in his behavior and motor development, they also noticed slight reduction in the head size. A CT scan at that time showed stable ventricular size and subdural space, while posterior fossa cyst size was marginally smaller [Figure 4].

DISCUSSION

The authors report an unusual complication of acute respiratory failure following ETV, performed for the treatment of obstructive hydrocephalus secondary to a large posterior fossa arachnoid cyst, and downward descent of the cerebellar tonsils through the foramen magnum.

Although infants with small posterior fossa cysts have subtle symptoms or incidentally diagnosed, most large cysts present with hydrocephalus, Chiari malformation, and hydrocephalyasia, as in our case[4,13-15] At the time of presentation the brain stem was tolerating two relatively equal forces of supra- and infra-tentorial pressure; one exerted by the posterior fossa cyst and the second force by the hydrocephalus. This pressure gradient had changed after ETV and insertion of an EVD that lead to relative expansion of the cyst, probably due to occluded aqueduct of Sylvius. Subsequently the brain stem was pushed against the clivus and more crowding of the foramen magnum and compression of the medulla oblongata occurred. This resulted in early postoperative respiratory deterioration and the need for mechanical ventilation.

Arachnoid cysts are usually said to remain stable in size.[14-16] The onset and rapid deterioration to respiratory failure in this patient is correlated to change in the posterior fossa cyst size and change in its pressure. Upward herniation following insertion of EVD for the treatment of hydrocephalus in the presence of posterior fossa space occupying lesion may lead to rapid and probably fatal respiratory failure.[13,17] Davis et al. 2001[18] also reported acute respiratory failure due to bilateral vocal cord paralysis following insertion of ventriculoperitoneal shunt in a child with obstructive hydrocephalus associated with posterior fossa tumor and Chiari malformation. They attributed the cause to the presence of large brain stem astrocytoma, and upward progression of the tumor. In our case EVD was kept opened at 10-cm height from the patient’s head, which could be attributed to the change in pressure gradient and created pressure on the brain stem, and may explain rapid postoperative respiratory failure.

In our case when, the cause of postoperative inspiratory stridor was not obvious and intubation trauma secondary to two attempts at intubation was considered to be a possibility.[12] However, postoperative scan showed markedly compressed brain stem by the posterior fossa cyst and herniated cerebellar tonsils, craniocervical decompression and cystoperitoneal shunt were decided to be done in an attempt to decompress the brain stem. The

Figure 3: MRI scan taken after ETV and EVD insertion showing wide supratentorial subdural space, along with marginal increase in the posterior fossa cyst (solid white arrows) and anterior displacement and compression of the brain stem against the clivus (black linear arrows). The brain stem was kinked and compressed, and the fourth ventricle was distorted and occluded (white linear arrows). The foramen magnum was crowded by the cerebellar tonsils compromising the medulla.

Figure 4: Postoperative CT scan (a) and 1-month follow-up CT scan (b) showing reduction in the size of the ventricles, persistent wide supratentorial subdural space.
follow-up CT scan showed a reduction in the size of the cyst to correlate to our clinical findings, and showed also extra-axial fluid collection representing subdural hygroma [Figure 4].

The child tolerated the procedure and could be weaned off ventilation and discharged home after a week. He was seen after 1 month and 6 months from discharge and he continued to improve, his head size reduced but the hygroma persisted.

The authors believe that insertion of EVD after ETV in this case contributed to the postoperative complication. Navarro et al. 2006 identified risk factors in their series of 136 ETV procedure. They found a higher incidence of early and late complication rate in children younger than 1 year-old, and patients that had an EVD after ETV, and both risk factors were present in this patient.

CONCLUSIONS

It is not advisable to perform ETV for hydrocephalus secondary to posterior fossa cyst in presence of occluded aqueduct without draining the cyst, e.g. by aqueductoplasty, or cystoperitoneal shunt.

After ETV, the acute relative increase in posterior fossa pressure and posterior fossa cyst size produced compression on the brain stem and the cervicomедullary junction that contributed to the immediate deterioration and resulted in acute respiratory failure.

Supportive care with mechanical ventilation and brain stem decompression were the mainstay of treatment.

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REFERENCES

1. Becker T, Wagner M, Hofmann E, Warmuth-Metz M, Najimi M. Do arachnoid cysts grow? A retrospective CT volumetric study. Neuroradiology 1991;33:341-5.
2. Bernard R, Vallee F, Mateo J, Marsella M, George B, Payen D, Chibbaro S. Uncontrollable high-frequency tachypnea: A rare and nearly fatal complication of endoscopic third ventriculostomy: Case report and literature review. Minim Invasive Neurosurg 2010;53:270-2.
3. Broekmeyer D, Abtin K, Carey L, Walker ML. Endoscopic third ventriculostomy: An outcome analysis. Pediatr Neurosurg 1998;28:236-40.
4. Chazal J. Management of Hydrocephalus in Childhood. In: Sindou M, editor. Practical handbook of neurosurgery from leading neurosurgeons. vol 1, Wien New York: Springer; 2009. p. 524-41.
5. Holst AV, Danielsen PL, Juhler M. Treatment options for intracranial arachnoid cysts: A retrospective study of 69 patients. Acta Neurochir Suppl. 2012;114:267-70.
6. Gangemi M, Seneca V, Colella G, Cioffi V, Imperato A, Maiuri F. Endoscopy versus microsurgical cyst excision and shunting for treating intracranial arachnoid cysts. J Neurosurg Pediatr. 2011;8:158-64.
7. Elgamal EA, Richards PG, Patel UJ. Fatal haemorrhage in medulloblastoma following ventricular drainage. Case report and review of the literature. Pediatr Neurosurg 2006;42:45-8.
8. Galarza M, López-Guerrero AL, Martínez-Lage JF. Posterior fossa arachnoid cysts and cerebellar tonsillar descent: Short review. Neurosurg Rev 2010;33:305-14.
9. Gazioglu N, Kafadar AM, tanriover N, abuzayed B, Biceroglu H, Ciplak N. Endoscopic management of posterior fossa arachnoid cyst in an adult: Case report and technical note. Turk Neurosurg 2010;20:512-8.
10. Handler MH, Abbott R, Lee M. Anore-fatal complication of endoscopic third ventriculostomy: Case report. Neurosurgery 1994;35:525-8.
11. Harsh GR, Edwards MS, Wilson CB. Intracranial arachnoid cysts in children. J Neurosurg 1986;64:835-42.
12. Jain R, Sawlani V, Phadke R, Kumar R. Retrocerebellar arachnoid cyst with syringomyelia: A case report. Neur India 2000;48:81-3.
13. Jamjoom ZA. Intracranial arachnoid cysts: Treatment alternatives and outcome in a series of 25 patients. Ann Saudi Med 1997;17:288-92.
14. King JA, Auguste KI, Halliday W, Drake JM, Kulkarni AV. Ventriceulocystostomy and endoscopic third ventriculostomy/shunt placement in the management of hydrocephalus secondary to giant retrocerebellar cysts in infancy report of 3 cases. J Neurosurg Pediatr 2010;5:403-7.
15. Martinez-Lage J, FAlmagro MJ, Ros de San Pedro J, Ruiz-Espejo A, Felipe-Murcia M. Regression of syringomyelia and tonsillar herniation after posterior fossa arachnoid cyst excision. Case report and literature review. Neurocirugia (Aстur) 2007;18:227-31.
16. Massimi I, Di Rocco C, Tamburrini G, Caldarelli M, Iannelli A. Endoscopic third ventriculostomy: Complications and failures. Minerva Pediatr 2004;56:167-81.
17. Navarro R, Gil-Parras R, Reitman AJ, Olavarría G, Grant JA, Tomita T. Endoscopic third ventriculostomy in children: Early and late complications and their avoidance. Childs Nerv Syst 2006;22:506-13. Epub 2006 Jan 11.
18. Schroeder HW, Niendorf WR, Gaab MR. Complications of endoscopic third ventriculostomy. J Neurosurg 2002;96:1032-40.
19. Tamburrini G, Di Rocco C. Congenital Arachnoid Cysts. In: Lumenta CB, Di Rocco C, Haase J, Mooij JJ, editors. European manual of medicine neurosurgery. Berlin Heidelberg: Springer-Verlag; 2010. p. 305-11.