Case Report

Successful Treatment of Large Occipital Encephalocele Presenting with Bilateral Vocal Cord Paralysis: A Case Report

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A large subtorcular occipital encephalocele in neonate carries higher risk of associated hindbrain anomalies and secondary process for neurological deterioration which predict the surgical outcomes and long-term prognosis. The dysfunction of the lower cranial nerves often leads to worsening of neurological status from poor respiratory function and repeated aspiration pneumonia. The aims of repairing encephalocele include a good closure of the defect, preservation or restoration of neurological functions and better cosmetic results. The author presented a successful surgical strategy for management of a 4-month-old infant with a large subtorcular occipital encephalocele presented with bilateral vocal cord paralysis and swallowing dysfunction. A step by step approach unlocked the main mechanism(s) of reversible lower cranial dysfunctions in this specific situation, including the increased intracranial pressure and shifting of the axis of lower brain stem.

Keywords: *Occipital encephalocele, Vocal cord paralysis, Lower cranial nerves dysfunction*

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Occipital encephalocele is less commonly found as compared to fronto-ethmoidal encephalocele in Southeast Asian countries[1,2]. Most encephaloceles can be easily detected during prenatal period and at birth. The symptoms and signs of encephalocele depend on the location, size and its contents. The giant size of encephalocele may consist of cerebrospinal fluid (CSF) and other herniated brain tissue, which carry the higher risk of rupture and difficulties of head positioning. Presenting symptoms of giant occipital encephalocele, except from bulging neck mass, include disturbances in respiratory function, swallowing difficulties and delayed development. Hydrocephalus or microcephalic head and other hindbrain anomalies are possible associated findings. The treatment aims at the closure of the defect and restoration of neurological functions with good cosmetic results. The challenges in achieving these aims varies but not excluding establishing a patent airway for anesthesia, positioning for operation, perioperative care of infancy, timing of surgery and type of operation, especially when acute respiratory distress occurs. The author presented a successful strategy for repairing of a large occipital encephalocele in a 4-month-old baby suffering from severe upper airway obstruction. A step by step approach unlocked the main mechanism of delayed lower cranial nerves dysfunction.

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Case Report

A 4-month-old male infant was born with a large occipital encephalocele and had been admitted several times for chronic aspirated pneumonias. His parents gave a history of progressive upper airway stridor over a month before admission. Through clinical examination Global Developmental Delay (GDD) was revealed a remarkable inspiratory stridor with occasionally apnea episodes were documented pre-operatively. The stridor was easily observed with 50 percent retraction of his anterior chest wall and sternum. Lying on his back was restricted because of worsening upper airway obstruction and desaturation of oxygen level. The anterior fontanelle was mildly tense and measured with the size about 2.5x2.5 cm^3. His limbs moved symmetrically with at least grade 3 in motor

power. The size of occipital sac was approximately 8x9x10 cm^3 with positive transillumination test. Pre-operative direct laryngoscopy revealed barely any movement of bilateral vocal cord. The MRI of the brain showed a large occipital encephalocele associated with hydrocephalus, distortion of brain stem, syringomyelia in high cervical cord and possible hindbrain anomalies (Fig. 1). The multidisciplinary care involved a pediatric respiratory team, a pediatric neurological team, a pediatric ENT team and a pediatric anesthesiologist team.

On the day of surgery, the pediatric ENT team was notified for a potential pre-operative tracheostomy if endotracheal intubation failure occurred. The laryngomalacia was another difficult issue for intubation in this case. The large occipital sac was allowed to sit in between the gel cushion and shoulder roll. Fortunately, the tube was safely inserted and secured with the suture because the surgery was performed under prone position. The sac was repaired under operating microscope. A part of the occipital horn was seen during the operation. The pathology showed gliotic brain tissue. The endotracheal tube was extubated 24 hours after surgery. Though improvement of the respiratory stridor was observed during the first few days of operation, the stridor and apnea episodes were still going with small amount of bulging anterior fontanelle. A right ventriculo-peritoneal shunt was performed one week after the first operation. The stridor remarkably disappeared. Furthermore, the swallowing dysfunction gradually improved after one month. The postoperative MRI revealed satisfactory closure of the defect and improvement of brain stem axis, hydrocephalus and stable cervico-thoracic syringomyelia. The parents reported a significant improvement of their child's GDD and nutritional status at a one-year follow-up.

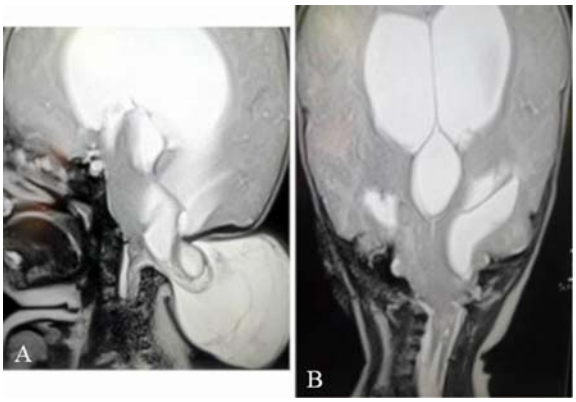

Fig. 1 Preoperative MRI brain from sagittal view (A) and coronal view (B) revealed moderate to severe hydrocephalus, high cervical syringomyelia, large occipital encephalocele with relatively small amount of herniated brain tissue in the sac and dilated of occipital horn with downward herniation causing distortion of brain stem axis.

Discussion

Occipital encephalocele occurs at an estimated rate of one in every 3,000 to 10,000 live births. The term "subtorcular occipital encephalocele" provides the better information about anatomical defect caudal to the torcula, regardless of the presence or absence of the occipital lobe tissue in the sac[3]. From a postmortem study, the associated anomalies were lack of cerebellum or part of cerebellum, distorted brainstem like S-shaped kink, hydrobulbia and hydromyelia[4]. The presence of cerebral tissue herniation into the sac, microcephaly had been associated with poor, long-term outcome[5,6]. A large occipital encephalocele can be presented with an airway obstruction more commonly than the anterior encephalocele because the anatomical defect is close to the important hindbrain structures. The airway problems in the occipital encephalocele seem to be neurogenic in origin rather than mechanical as seen in a basal encephalocele. The pathophysiology of vocal cord paralysis can be result of syringobulbia, hindbrain anomalies itself, displacement of brain stem or increased intracranial pressure[7]. The stridor at birth is associated with poor prognosis as compared to later development of stridor, probably related to hindbrain anomalies[8]. Reversible vocal cord paralysis was reported after relieving of intracranial pressure in patients with hydrocephalus[9,10]. Furthermore, the difficulties for establishing the airway were complicated by a large encephalocele sac and sometimes with other associated abnormalities such as micrognathia, macroglossia[11]. A careful evaluation of anesthetic care is recommended in this special condition such as method of positioning, intubation and perioperative complications[12,13]. The pre-operative tracheostomy is sometimes considered as an option before a definite surgical repair, especially in the respiratory distress condition. However, it is better if tracheostomy can be avoided, especially in a small child. The decision-making had to be discussed with the parents after multidisciplinary team conference. Because of the unknown actual mechanism of airway obstruction, staged operation is recommended at the beginning. The second operation probably relieves pressure

effects to lower brain stem nuclei and hydrocephalus contributing to dramatic improvement of upper airway obstruction and swallowing dysfunction. Alternative option is a shunt operation first and then followed by repairing of encephalocele[14,15]. However, caring about the neck position with large sac can be complicated by infection[16]. The simultaneous surgical repair and shunt operation may be another reasonable option in hydrocephalic patient. Our strategy was done step by step for safety in neonatal surgery, which unlocked the main mechanism(s) of vocal cord paralysis. Indirectly, it relieved the pressure on the lower cranial nerves nuclei which showed the improvement in both vocal cord movement and swallowing function. Relieving the hydrocephalus improved overall alertness, consciousness, posture and limbs movement, which enhanced the restoration of neurological development. Although spine MRI in early post-operative period revealed stable in size and extension of syringomyelia, clinical improvement of diaphragmatic function and chest wall movement can be observed.

Occipital encephalocele is the part of Chiari III malformation, which characterized by herniation of posterior fossa contents through the bony defect around occiput and high cervical area. It is the rarest type of Chiari associating with higher morbidity and mortality[17]. The MRI imaging of Chiari III malformation is important for associated findings such as hydrosyringomyelia, bony defects or anomalies, location of pons and medulla, dysgenesis of corpus callosum[18]. Anomalies of venous drainage of large occipital encephalocele can be seen and often assist in preparing for a massive blood loss in small neonate surgery[19]. Pre-operative MRV, especially around straight sinus and tentorium, had to be carefully reviewed[20]. The herniated brain tissue can be occipital lobe or cerebellar tissue. An innovative reconstruction technique for preservation of visual function was successful performed[21]. Timing of the surgery is recommended to be done as soon as possible to save the child's life-threatening conditions such as respiratory distress, improve child's development and decrease the incidence of infection such as CNS infections, aspirated pneumonias, and irreversible damage of nucleus ambiguous for vagus nerve[9].

In summary, either occipital encephalocele or Chiari III malformation addressed specific problems, which can occur congenitally or later on. Delayed onset of lower cranial nerves dysfunction opens the opportunity to restore neurological function with satisfactory outcomes.

What is already known on this topic?

Multiple factors contribute to airway obstruction in large occipital encephalocele. The prognosis of disease can be varied depending on underlying malformation and its complications.

What this study adds?

Occipital encephalocele is uncommon in Thailand. A successful surgical plan and multidisciplinary team approach are very important for good. long-term outcomes.

Potential conflicts of interest

None.

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รายงานการผ่าตัดแก้ไขความผิดปกติของสมองส่วนท้ายทอยขนาดใหญ่แต่กำเนิดที่มาด้วยอาการอัมพาตของสายเสียงทั้งสองข้าง

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ความผิดปกติของสมองที่ยื่นบริเวณ occipital ขนาดใหญ่ในเด็กเล็กมักพบความพิการของสมองส่วน hindbrain ร่วมด้วยและอาจส่งผลทำให้การหายใจล้มเหลว ซึ่งมีผลโดยตรงต่อการทำงานของระบบประสาท ความผิดปกติของระบบประสาทนี้ส่งผลต่อการพยากรณ์โรคในระยะยาว และยากต่อการรักษาโดยเฉพาะเด็กเล็ก จุดประสงค์การผ่าตัดรักษาเพื่อปิดเยื่อหุ้มสมองกันไม่ให้สมองยื่นออกมา โดยรักษาการทำงานของสมองที่เกี่ยวข้องให้ปกติมากที่สุดและเพื่อความสวยงามของรูปศีรษะ ความพิการของเส้นประสาทสมองส่วนล่างส่งผลต่อการควบคุมหายใจและการกลืน อาจทำให้เกิดปอดบวมตามหลังการสำลักได้บ่อยทำให้การทำงานของสมองแย่ลง ผู้นิพนธ์รายงานผู้ป่วยเด็กเล็กอายุ 4 เดือน ที่มาด้วยก้อนที่ท้ายทอยขนาดใหญ่และมีอาการอัมพาตของสายเสียงทั้งสองข้างและอาการอัมพาตของระบบการกลืน ซึ่งได้รับการวางแผนการผ่าตัดเป็นขั้นเป็นตอน เพื่ออธิบายกลไกการเกิดความพิการในเส้นประสาทสมองส่วนล่าง ซึ่งได้แก่การเพิ่มความดันในกระโหลกศีรษะและการเคลื่อนของแกนสมอง