Abstract: Congenital malformations of various sorts account for a large percentage of childhood hydrocephalus but associated Craniovertebral junction (CVJ) anomaly has not been reported earlier. Though the anaesthetic concerns for isolated arrested hydrocephalus and CVJ anomaly has been reported but the concomitant occurrence of both and its anaesthetic implications is not mentioned in literature. Here we present the anaesthetic management of a child with arrested hydrocephalus along with CVJ anomaly leading to compression of cervicomedullary junction and myelopathy scheduled for decompression and fixation of craniovertebral junction.

Key Words: Hydrocephalus; Craniovertebral junction (CVJ) anomaly; anaesthesia; surgery.

Introduction

Hydrocephalus can be defined as a disturbance of formation, flow, or absorption of cerebrospinal fluid (CSF) that leads to an increase in volume occupied by this fluid in the central nervous system (CNS). In some cases, ventricular dilation terminates spontaneously, at which point it is termed as “compensated” or “arrested” hydrocephalus (1, 2).

The craniovertebral junction have close relation of the bony, vascular and nervous elements. So, any congenital malformation at this junction may produce a variety of symptoms and signs which can be structural deformities usually with a wide spectrum of neurological deficit (3, 4).

Congenital malformations of various sorts account for a large percentage of childhood hydrocephalus but associated Craniovertebral junction (CVJ) anomaly has not been reported earlier. Though the anaesthetic concerns for isolated arrested hydrocephalus and CVJ anomaly has been reported but the concomitant occurrence of both and its anaesthetic implications is not mentioned in literature. Here we present the anaesthetic management of a child with arrested hydrocephalus along with CVJ anomaly leading to compression of cervicomedullary junction and myelopathy scheduled for decompression and fixation of craniovertebral junction.

Case report

A 5-year-old, 15 kg girl was scheduled for posterior decompression and occipito-C2 fixation for craniovertebral junction (CVJ) anomaly with arrested hydrocephalus. She presented in the neurosurgical clinics with the chief complaints of difficulty in walking for six month. In the perinatal history, she was detected to have intrauterine growth retardation. She was born at term with no history suggestive of perinatal asphyxia. She had a history of generalized tonic-clonic convulsions at 18 months of age. She was on oral phenytoin. The seizures recurred along with loss of consciousness 5 times in last 6 months. There was no history suggestive of trauma to head or spine.

On examination, she had bilateral congenital talipes-equino-varus. Pulse rate was 92 beats/minute, respiratory rate was 20/min. On auscultation, respiratory system and cardiovascular system revealed no abnormality. Neck movements were not assessed in view of CVJ anomaly along with neurological deficit. Head circumference was increased. Cervical X-rays revealed no obvious abnormality. Magnetic resonance imaging showed hypoplastic C1 arch posteriorly, impinging upon thecal cord dorsally with compression of cervical medullary junction and marked hydrocephalus.

No premedication was administered preoperatively. Child was shifted to the operation table...
and folded towel was kept beneath the torso with head fully supported. Routine monitors (electrocardiogram, pulse oximeter, non invasive blood pressure) were attached. Difficult airway cart including paediatric fiberoptic bronchoscope was kept ready. Anaesthesia was induced with incremental concentration of sevoflurane (2-8%) in 100 % oxygen. Intravenous access with 22 G cannula was secured. Fentanyl 30 mcg and rocuronium 15 mg was administered intravenously. An assistant applied manual-in-line-stabilization and airway was secured with cuffed endotracheal tube size 5. Temperature and capnography monitoring was initiated. Arterial cannulation was done in dorsalis pedis artery for invasive blood pressure monitoring. Head was fully supported with manual in-line stabilization (MILS) while positioning the patient prone to Wilson’s frame. Surgical duration was five hours. Blood loss of 100 mL was replaced with colloid. Intraoperative analgesia was provide with top ups of injection fentanyl (15 mcg). Intraoperative course was uneventful with maintained hemodynamic parameters (ie within 20% of baseline values). At completion of surgery trachea was extubated after reversal of residual neuromuscular blockade. The difficult airway cart was kept ready at extubation and also in paediatric neurosurgical intensive care unit where child was shifted for further management. The respiratory and hemodynamic parameters remained stable. She was shifted to ward on 3rd postoperative day and was discharged home on the 7th postoperative day uneventfully.

**DISCUSSION**

Hydrocephalus is not a disease but a dynamic process which proceeds with changes of the ventricular system size. Causes and clinical symptoms of hydrocephalus are changing with the patient’s age but 55% of all cases have congenital origin. The most frequent cause of hydrocephalus with elevated intracranial pressure (ICP) in children before age of two is intraventricular haemorrhage in perinatal period. The next causes are the congenital inflammatory process of the central nervous system such as TORCH (toxoplasma, rubella, cytomegalay, herpes simplex) which leads to brain damage. Hydrocephalus as a symptom can coexists with such developmental anomalies like Dandy-Walker syndrome, myelomeningocele or narrowed Sylvian aqueduct. Hydrocephalus is often the result of head injury with intracranial haemorrhage in this group of age (5). Schick and Matson reported that this spontaneous termination occurs in 10-15% of cases of hydrocephalus. The arrest in hydrocephalic progression generally occurs at the age of 1-2 yrs and is due to resumption of the proper balance between the production and absorption of CSF due to the development of a minor CSF pathway (6). This pathway involves an increase in circumference in the circumventricular blood vessels and the absorption of CSF by intraxial blood vessels. Many factors like fever, infections can upset this balance, leading to sudden decompensation of the hydrocephalus and elevated ICP. These patients may also decompensate, especially following minor head injuries. So care is warranted to take full aseptic precautions during invasive procedures and to treat any suspected infections at the earliest.

A wide spectrum of congenital, developmental and acquired abnormalities arises at the cranio-cervical border. This is because of the complex developmental anatomy, the transition between the spinal cord and the brain, and the junction between the highly mobile upper cervical spine and the skull. This results in compression and distortion of neural structures, the vertebral basilar vascular system and cerebrospinal fluid pathways. Intermittent attacks of altered consciousness (as seen in our patient too), confusion and transient loss of visual fields, as well as vertigo, occur in 25% of children with abnormalities of the craniovertebral junction. This is provoked by rotation or extension of the head, or manipulation of the head and neck (7). Atlas hypoplasia is rare anomaly that is strongly associated with upper cervical cord compression and myelopathy due to spinal cord deformity (8, 9).

Patients with CVJ anomaly with arrested hydrocephalus present distinct challenges to anaesthesiologist. The main concerns in the anaesthetic management includes difficulty in securing airway, maintaining immobility at cervical spine during airway management and positioning apart from concerns related to hydrocephalus, compressive myelopathy and unstable cervical spine. The concern of exaggerated haemodynamic fluctuations due to surgical manipulation near medulla in patient with hydrocephalus. Sedative premedication should be considered carefully as it may exacerbate or mask signs of neurological dysfunction. Conversely, an anxious combative patient may have a detrimental rise in ICP during induction. Gaseous induction is an acceptable alternative with a non-irritant volatile anesthetic agent such as sevoflurane or halothane. A rapid gaseous induction is preferable to prolonged attempts at cannulation in...
a distressed and unwell child. Once anaesthetized, intravenous access should be established quickly to allow the use of a neuromuscular blockade to facilitate control of the airway. Hypercarbia should be avoided as it causes cerebral vasodilatation and may worsen raised ICP especially in combination with laryngoscopy and airway manipulation. Positioning and intubation requires meticulous attention. Cervical spine anomalies reduce mobility of the neck and produce difficulties in managing the airway for the anaesthesiologist (7) As in our patient hypoplastic C1 arch was impinging upon thecal cord dorsally and there was compression of cervical medullary junction. During positioning and intubation this impingement may increase further morbidities including problems with the cardiovascular (e.g. congenital cardiac disease) and respiratory systems (e.g. bronchopulmonary dysplasia, kyphoscoliosis, recurrent respiratory infections secondary to neurological dysfunction). Patient with congenital CVJ anomaly may have decreased pulmonary reservoir and these values may further decrease during intraoperative and postoperative period (12).

We conclude the concomitant occurrence of arrested hydrocephalus and CVJ anomaly requires great vigilance in the perioperative period including airway management and haemodynamic fluctuations.

Children with hydrocephalus are prone for hypertension and bradycardia (Cushing’s response) and compression at medullary junction (as in our patient) may lead to further hemodynamic instability. Moreover any vascular compression at CVJ can further compromise the cerebral perfusion in already compromised perfusion because of hydrocephalus. Hypotension should be avoided because of the risk of decreasing cerebral perfusion pressure in the face of raised ICP. That is why we used invasive blood pressure monitoring to maintain the mean arterial pressure and hence the cerebral perfusion. Particular attention should be paid to the possibility of comorbidities including problems with the cardiovascular (e.g. congenital cardiac disease) and respiratory systems (e.g. bronchopulmonary dysplasia, kyphoscoliosis, recurrent respiratory infections secondary to neurological dysfunction). Patient with congenital CVJ anomaly may have decreased pulmonary reservoir and these values may further decrease during intraoperative and postoperative period (12).

References

