Experience In The Management Of Dandy-Walker Syndrome In An Adult Patient

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Abstract

Dandy-Walker Syndrome (DWS) is a congenital brain malformation involving the cerebellum and the fluid spaces around it. There is atresia of foramenofMagendie and Lushka resulting in a complete absence of the part of the brain located between the two cerebellar hemispheres (cerebellarvermis) and cystic dilatation of the fourth ventricle. It may be associated with other anomalies in the brain and other parts of the body. Symptoms such as delayed developmental milestone, bulging of skull may present early in life or may present later in life. We present a case of DWS diagnosed in adulthood with features of raised intracranial pressure (ICP) (headache, vomiting, neck stiffness, and convulsion) and poor academic performance. Difficult airway was anticipated due to the presence of micrognathia and reduced neck mobility.The patient was managed successfully with cysto-periotoneal shunt insertion under general anaesthesiawithout postoperative ventilation.

Keywords: Dandy-Walker Syndrome, Hydrocephalus, VP-Shunt, Airway, Adulthood.

Introduction

Dandy-Walker syndrome is a rare sporadic genetic disorder characterized by a neuropathological triad comprising of hypoplasia of the cerebellarvermis, cystic dilatation of the fourth ventricle and hydrocephalus as a result of atresia of the exit foraminal of Magendie and Luschka. The syndrome was first described by Dandy and Blackfan in 1916. The syndrome has a slight female predominance and an incidence of around 1 in 25,000-30,000 life births.[1,2]The clinical manifestation usually appear in the first year of life, but can also occur in the neonatal period.[3]

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The symptoms that usually become apparent in early infancy include slow motor development, bulging of the anterior fontanel and progressive enlargement of the skull due to the obstructive hydrocephalus. Associated congenital anomalies may include craniofacial anomalies such as cleft palate, micrognathia, hypertelorism, cardiac, renal and skeletal malformations. Cerebral anomalies include agenesis of corpus callosum with poor intelligence and interference with medullarycontrol of respiration which can lead to respiratory failure. We report our experience in the management of a twenty one year old adult diagnosed with Dandy- Walker Syndrome who had cyto-peritoneal shunt on account of raised intracranial pressure.

Case Report

A twenty-one year old boy weighing 55 kgspresented with history of persistent headache and progressive loss of vision of 2 weeks duration. There was associated neck pain, photophobia and vomiting. He had an episode of tonic clonic convulsion which lasted for about 3 minutesduring which he hit his head on the floor. The seizure aborted spontaneously prior to presentation to the hospital. There was past history of convulsionat3 years of age and was been managed for seizure disorder with phenobarbital. There was no history suggestive of cardiac or renal anomalies. No history of delayed developmental milestone. He was withdrawn from secondary school due to poor academic performance.

Pre-anaesthetic examination, revealed a conscious boy with micrognathia, ptosisof left eye (figure 1) with dilated unreactive pupil and todd's palsy affecting the right upper and lower limb. There was neck stiffness with positive Kernig's sign. Mouth opening was adequate(Figure 2) and has Mallampati II. Haemogram and electrolyte result were within normal range. Brain CT-scan revealed cystic dilatation of 4th ventricle with mild to moderate dilation of other ventricles and agenesis of cerebellarvermis. (Figure3) An assessment of Dandy-Walker syndrome was made with secondary hydrocephalus. Consent was obtained for anaesthesia and surgery and fasting guideline was observed.

Intraoperatively, multi-parameter monitor was attached, peripheral saturation, pulse rate, blood pressure, was monitored. An intravenous access was secured on the left hand using size 18 G cannula. He was pre oxygenated with 100% oxygen for 5min with a

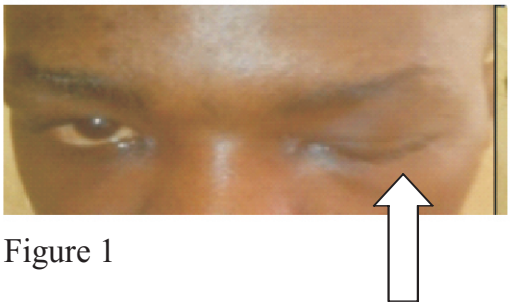

Figure 1

Left eye ptosis

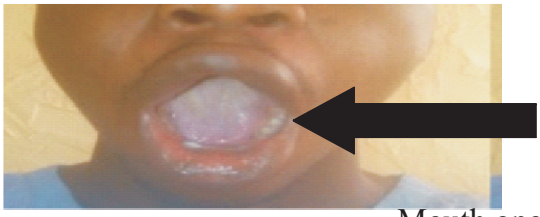

Figure 2

Mouth opening

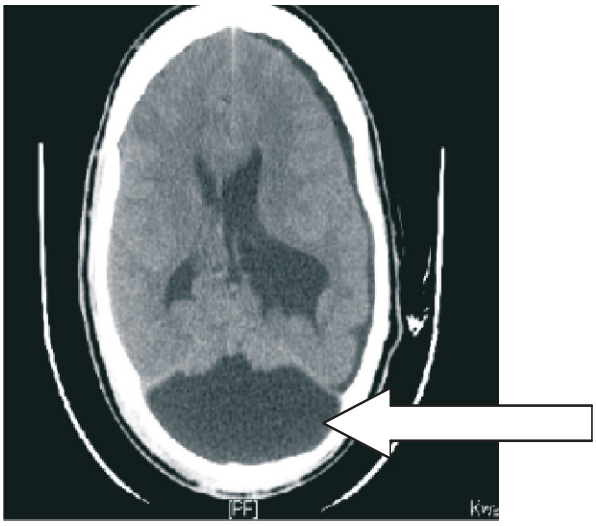

Figure 3

Cystic dilatation of 4th ventricle

tight fitting face mask. Anaesthesia was co-induced with I.V fentanyl 2 µg/kg and propofol 2 mg/kg. Laryngoscopy and tracheal intubation was facilitated with I.V suxamethonium 1 mg/kg.Tracheal was intubated with size 6.5 mm cuffed endotrachealtube and correct tube placement was confirmed with equal air entry on auscultation.

Anaesthesia was maintained with propofol using TIVA, intermittent pancuronium for muscle paralysis, paracetamol and intermittent fentanyl for analgesia. During the course of the surgery, he was haemodynamically stable with SpO2-100%, end tidal CO_2 was maintained between 30 and 35mmHg pulse rate maintained between 70-90/min, and blood pressure between 100-130/60-80 mmHg. Patient had cysto-peritoneal shunt insertion. The procedure lasted for 2 hours 30 min and went uneventful.

At the end of the surgery, patient regained spontaneous respiratory effort. Residual neuromuscular paralysis was reversed with neostigmine 0.05 mg/kg and atropine 0.02 mg/kg. He was extubated after he regained full consciousness and SpO2 of 96-98% in room air. Patient was transferred to recovery room for further monitoring and was maintained on I.V phenytoin sodium postoperatively to prevent seizure.

Patient was discharged home on 4th day postoperatively.

Discussion

The most common clinical presentation of Dandy-Walker syndrome are enlargement of the occiput , macrocrania, mental retardation, cerebellar ataxia and raised intracranial pressure.[4] In older children with Daddy-Walker syndrome, symptoms of increased ICP, including irritability and vomiting, signs of cerebeller dysfunction, including gait disturbance and lack of muscle coordination, may occur.[5]This patienthad been managed as a case of seizure disorder since the age of 3 years with no suspicion of intracranial lesion. The patient presented to our facility with features of raised ICPand worsening neurological status. No radiological evaluation was done probably because of lack of image facility, lack of awareness or ignorance of the managing team since the onset of the symptoms.Accidentally, the diagnosis of Dandy-Walker syndrome was madewith cranial CT scanwhen he was being evaluated for traumatic brain injury after a seizure episode at age 21.

In the present case, the patient had raised ICP and micrognathia. Presence of micrognathiacan make airway management challenging as reported by Ewart and Oh.[6]This patient had mallampati II on airway assessment and he is not at risk of difficult intubation.The important anaesthetic concern in our patient was the management of the ICP and the airway. When difficulties in airway management are anticipated, awake intubation is required despite the increased ICP. In our case, intubation was not difficultdespite the presence of micrognathia, although, difficult airway tray was made available. This finding is similar to that reported by Ji Su Jang et al [5]and Shweta M[7]respectively. Important management strategies should include prevention of further increase in ICP; hence,induction and maintenance of anaesthesia were attained by continuous infusion of propofol which reduced the ICP. End tidal CO_2 pressure was also maintained between 30 and 35mmHg to control the ICP. Intermittent boluses of fentanyl and IV paracetamol were used for analgesia and BP control.In this patient, normocapnia,normotension was also employed to reduce ICP and this is similar to Ramesh et al[8]study.Anaesthesia was maintained with controlled

ventilation, analgesia and muscle relaxants and these has been reported by Kim et al .[9]Endotracheal intubation can be difficult due to the large size head in hydrocephalus and micrognathia. Intubation was made smooth by placement of shoulder roll. Emergence from anaesthesiawas smooth and the patient regained spontaneous respiration soon after surgery and did not need ventilatory support as respiratory pattern was good. Postoperatively convulsion was prevented by maintaining patient on intravenous phenytoin which was converted to oral on the second day postoperatively. He was discharged to out-patient clinic for follow-up on oral phenytoin.

Conclusion

Dandy-Walker syndrome may present outside infant age with increase ICP, attention to anaesthesia details is necessary for successfulmanagement.

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