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SHORT COMMUNICATION

A case report of anesthesia for a child with Hajdu–Cheney syndrome

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Abstract Hajdu–Cheney syndrome is an extremely rare disorder characterized by progressive skeletal acro-osteolysis, which results in extremity fractures and scoliosis often requiring surgical treatment from childhood. A unique facial structure and deformity of the cervical spine is associated with a difficult airway. We report here a 10-year-old girl with Hajdu–Cheney syndrome who developed progressive basilar impression and medullary compression for which foramen magnum decompression was performed. After slow induction of anesthesia, we were able to perform fiberoptic orotracheal intubation via a VBM bronchoscope airway. This case report contributes to the accumulation of knowledge about anesthesia for this rare syndrome.

Keywords Hajdu–Cheney syndrome · Pediatric anesthesia · Airway management · VBM bronchoscope airway

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Manuscript

Hajdu–Cheney syndrome is an extremely rare disorder, of which no more than 200 cases have been reported. It is characterized by acro-osteolysis, which results in extremity fractures and deformity of the spine from childhood. Hajdu–Cheney syndrome is inherited as an autosomal dominant trait and is sporadic. Such bone degeneration means that individuals with Hajdu–Cheney syndrome are likely to require surgery from an early age [1, 2]. The syndrome is accompanied by maxillofacial anomalies such as micrognathia and mandibular hypoplasia [1] and as such has increased risks associated with a difficult airway.

This 10-year-old girl with Hajdu–Cheney syndrome complained of a persistent headache at the age of 9 years. There was no familial history. At that time, magnetic resonance imaging (MRI) showed a basilar impression, syringomyelia, and obstructive hydrocephalus. Endoscopic fenestration of the third ventricle was performed before foramen magnum decompression (FMD), and this relieved her headache. One year later, she presented with disturbance of gait, sleep apnea, swallowing difficulty, and aspiration pneumonia. Magnetic resonance imaging (MRI) showed medullary compression (Fig. 1), and a second FMD under general anesthesia was planned.

In terms of the patient's surgical history, patent ductus arteriosus was repaired at 10 days of age, and a left forearm fracture was repaired and a palatoplasty performed at 5 years of age. Next, left patella luxation repair, patella tendon reconstruction, and right tibial facture repair were performed under general anesthesia with intubation at the age of 6 years. At 9 years of age, as already mentioned, endoscopic fenestration of the third ventricle was performed. During intubation, the patient's neck and head were immobilized by hand and Cormack grade was 1.

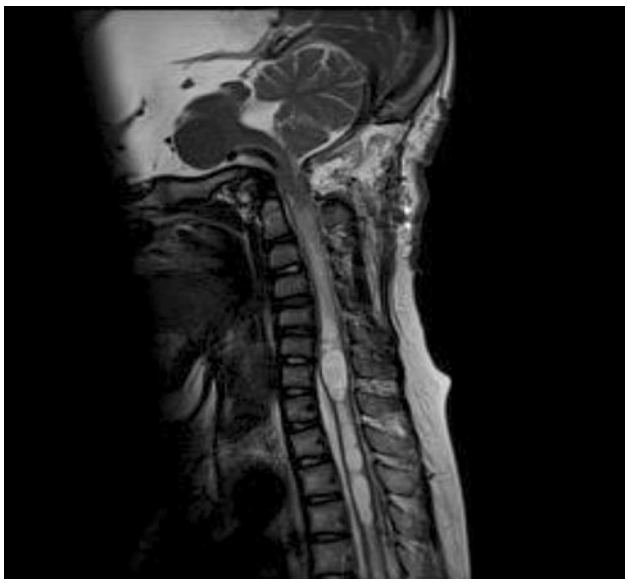

Fig. 1 Sagittal cervical T_2-weighted magnetic resonance imaging (MRI) of a 10-year-old girl with Hajdu–Cheney syndrome. Chiari malformation with basal invagination had recurred after endoscopic foramen magnum decompression

To perform FMD this time, anesthesia was started with 8 % sevoflurane. Under anesthesia we inserted a VBM bronchoscope airway without inducing a gag reflex, and ventilation was easily accomplished. We inserted a fiberscope through the side hole of the airway, and then could succeed in quickly achieving fiberoptic orotracheal intubation.

The patient was in a prone position while intubated, with her head placed in three-point fixation. There were no complications with the three-point fixation and the patient was extubated immediately after surgery. The patient showed no neurological complications associated with having undergone fiberoptic orotracheal intubation and three-point fixation.

Patients with Hajdu–Cheney syndrome, as in the present case, often need to undergo surgery during childhood because of progressive acro-osteolysis, which results in extremity fractures and deformity of the spine [1, 2]. These patients present a high risk of difficult airway during surgical treatment because of the complication of maxillofacial anomalies [1, 2]. The present patient had previously been intubated using a Macintosh laryngoscope before the first FMD at 9 years of age. Direct visualization of the larynx had been easy at that time. However, as deformity of the cervical spine and skull base were becoming worse, we expected her to have a difficult airway for the second FMD 1 year later.

Hajdu–Cheney syndrome is accompanied by syringomyelia, a basilar impression, and medullary compression symptoms [3, 4]. Also, as our patient had dysphagia and sleep apnea syndrome that had resulted in aspiration pneumonia, we considered that neck immobilization and fiberoptic intubation without neck extension would be appropriate.

The pediatric airway is prone to obstruction immediately after induction because of the short neck and large tongue. We must secure the airway during induction, and move on to operating fiberoptic orotracheal intubation quickly. The VBM bronchoscope bite block has a pediatric size that causes less discomfort to the patient. We could insert the VBM bronchoscope bite block during slow induction without gag reflex. We ensured the visual field via fiberscope soon with guiding with the VBM bronchoscope bite block side hole. There was no oxygen desaturation.

It was impossible for the patient to undergo awake intubation because of her young age, and fiberoptic orotracheal intubation was preferable to fiberoptic nasotracheal intubation because of the risk of fracture at the skull base. It was necessary that this patient's neck be immobilized. Orotracheal intubation was accomplished via the VBM bronchoscope airway.

There are few available reports on anesthesia for Hajdu–Cheney syndrome. We report here our experience with anesthesia for a child with Hajdu–Cheney syndrome.

Conflict of interest None.

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