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Cervical Cord Decompression and Posterior Cervical Immobilization Following Anterior C_1-C_2 Subluxation in a Patient with MPS Type IV (Morquio Syndrome) : A Team Approach

Suwannee Suraseranivongse, M.D.*
Pornswan Wasant, M.D.**
Achara Sathienkijkanchai, M.D.**
Surin Thanapipatsiri, M.D.***

Abstract : **We report a 23 month old girl who presented with right hemiparesis after falling for 6 months. She was also diagnosed with mucopolysaccharidosis type IV (Morquio syndrome) with C_1-C_2 subluxation. Anesthesia was uneventful after inhalation induction, manual in-line stabilization, intubation, external fixation of the cervical spine, followed by prone position and manual assisted ventilation. Postoperative pain was relieved using a tramadol infusion.**

เรื่องย่อ : การผ่าตัด Cervical Cord Decompression และการทำ Posterior Cervical Immobilization ในผู้ป่วย MPS Type IV (Morquio Syndrome) ที่มีการเคลื่อนของกระดูกสันหลังคอที่ 1 และ 2 : การดูแลผู้ป่วยเป็นทีม

สุวรรณี สุรเศรณีวงศ์ พ.บ.*, พรสวรรค์ วสันต์ พ.บ.**, อัจฉรา เสถียรกิจการชัย พ.บ.**, สุรินทร์ ธนพิพัฒนศิริ พ.บ.***
**ภาควิชาวิสัญญีวิทยา, **ภาควิชากุมารเวชศาสตร์, ***ภาควิชาศัลยศาสตร์ออร์โธปิดิคส์, คณะแพทยศาสตร์ศิริราชพยาบาล, มหาวิทยาลัยมหิดล, กรุงเทพมหานคร 10700.*

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ผู้ป่วยเด็กหญิงอายุ 1 ปี 11 เดือน มาโรงพยาบาลด้วยอาการสำคัญคือแขนขาด้านขวาอ่อนแรงมานาน 6 เดือนภายหลังหกล้ม ผู้ป่วยได้รับการวินิจฉัยว่าเป็น mucopolysaccharidosis type IV (Morquio syndrome) และมีการเคลื่อนของกระดูกสันหลังคอที่ 1 และ 2 การระงับความรู้สึกเป็นไปอย่างราบรื่นประกอบด้วยการนำสลบด้วยวิธีสูดดม การตรึงคอและศีรษะโดยวิธี in-line stabilization ขณะใส่ท่อช่วยหายใจ การใส่ external fixation ของกระดูกสันหลังคอ แล้วจัดผู้ป่วยอยู่ในท่านอนคว่ำ จากนั้นช่วยการหายใจของผู้ป่วยโดยใช้มือควบคุม (manual assisted ventilation) หลังผ่าตัดผู้ป่วยได้รับ tramadol infusion เพื่อระงับปวด

**Department of Anesthesiology, **Department of Pediatrics, ***Department of Orthopedics, Factulty of Medicine Siriraj Hospital, Mahidol University, Bangkok 10700, Thailand.*
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INTRODUCTION

Morquio syndrome or mucopolysaccharidosis type IV is a congenital syndrome with anomalies in vital organs such as cardiac defect and cervical spine abnormalities leading to airway problems. The following case illustrates the perioperative management of this congenital syndrome using a team approach.

CASE REPORT

A 23-month old girl presented with right hemiparesis for 6 months prior to her arrival at Siriraj Hospital. Six months previously, she fell from a table (about 80 cm high). Her head and back hit the ground and she was unconscious for 10 minutes. The following morning, she developed right hemiparesis without alteration of consciousness, nausea, vomiting or fever. The first tertiary care hospital which she attended, diagnosed right hip dislocation from an x-ray. She did not improve on treatment. One month later she was moved to a tertiary care hospital for children where the doctor noted dysmorphic feature, a right foot drop and motor power grade IV/V at the right extremities. She was admitted for further investigations. A cervical spine x-ray showed flattening and gibbus deformity of the vertebral body. A brain CT showed a fracture of the right parietal bone without a mass or hemorrhage. Brain MRI did not show any detectable abnormality but urine mucopolysaccharide (MPS) was positive. Her corneas were mildly cloudy, therefore she was diagnosed as mucopolysaccharidosis with a right hemiparasis. She was followed up every month without any improvement. She then presented to Siriraj Hospital, and was admitted for further investigation and management to prevent further neurological sequelae. Her vital signs were within normal limits for age, body weight was 10.2 kg, height was 82 cm and head circumference was 47 cm. She had a short neck, broad mouth, prognathism and pectus carinatum. Heart and lungs were within normal limits. Abdomen was soft, not tender, the liver was 2 cm below right costal margin. Extremities showed bilateral genu valgus and kyphosis. Nervous system examination revealed E_4 V_5 M_6, motor power grade IV/V of right extremities, an extensor plantar response with clonus, and intact pain sensation. Cranial nerve function was normal.

Laboratory investigations showed normal hemoglobin (121.3 g/dl), normal white cell count (12,200/mm^3) and normal platelet count (425,000/mm^3). Her serum electrolytes were normal (sodium 138 mmol/L, potassium 4.3 mmol/L, chloride 104 mmol/L, bicarbonate 25 mmol/L). She had normal blood urea nitrogen (15 mg/dl) and creatinine (0.4 mg/dl). Her liver function was normal (LDH 480 units/L, SGOT 21 units/L, SGPT 15 units/L, alkaline phosphatase 45 units/L, total bilirubin 0.3 units/L, direct bilirubin 0.2 unit/L and albumin 3.4 g/L). Urine analysis was normal. She had a normal result on echocardiography. A lateral radiograph of cervical spine demonstrated anterior atlantoaxial (C_1 - C_2) subluxation, Magnetic resonance images of brain and spine showed hypoplasia of the odontoid process and ligamentous hypertrophy in the first and second cervical spine region with marked narrowing of the spinal canal and severe spinal cord compression at C_1 level. Her urine metabolic screening was positive for mucopolysaccharides on the third sample. Eye examination showed mild corneal opacity and her hearing test was normal. Her developmental assessment was within normal range (DQ 101). The identification of urine mucopolysaccharides demonstrated keratan sulfate and chondroitin 6-sulfate via thin-layer chromatography which is consistent with MPS-Morquio syndrome. The definitive diagnosis was Morquio syndrome with cervical cord compression due to anterior C_1 - C_2 subluxation.

An orthopedic surgeon and anesthesiologist were consulted for preoperative evaluation and preparation including a preoperative plan. Clinical assessment of the range of active neck movement during flexion and extension did not induce further neurological deficit. The severity and direction of instability and reproducibility by means of flexion and extension views of lateral radiographs were assessed. An intubating position with mild cervical extension was allowed. Her mouth opened to about 3 cm, and the Mallampati test was grade 2. There was no micrognathia or retrognathia or any sign of difficult intubation. However, difficult airway equipment such as stylet, laryngeal mask airway and fiberoptic bronchoscope were prepared. After

intubation, preoperative stabilization of the cervical spine was planned using a halo vest.

On the day of surgery, atropine 0.1 mg IV was given as an antisialogogue. Her blood pressure was 160/80 mmHg and pulse rate was 120/min. Deep inhalation induction was achieved using nitrous oxide, oxygen and sevoflurane, and the trachea was intubated by conventional methods with manual in line traction neck stabilization. Anesthesia was maintained with nitrous oxide and oxygen, intravenous fentanyl 10 mcg followed by 5 mcg/hr and an intravenous infusion of diprivan 5-10 mg/hr. Ventilation was manually controlled throughout the operation with adequate oxygenation (SaO_2 99-100) and ventilation (end tidal CO_2 35-45 mmHg). Monitoring consisted of blood pressure from left radial artery catheterization, heart rate, electrocardiography, temperature, pulse oximeter, end tidal capnography, arterial blood gas, fluid intake and urine output including blood loss. Intravenous cefazolin 200 mg was given before taking the bone graft and one hour later before cervical incision. Dexamethasone 2 mg was given intravenously for reduction of tissue edema. After tracheal intubation, the cervical spine was stabilized with a halo vest, followed by the iliac bone graft. Then patient was turned to the prone position, and the upper cervical cord was decompressed by removing the posterior arch of C_1 which was compressing the spinal cord. Posterior fusion of the occiput to C_3 was performed to provide permanent stability using an autologous iliac crest bone graft in combination with a clip freeze dried allograft. Vital signs were stable (blood pressure 100/50 to 120/60 mmHg, heart rate 95 - 120/min), except during posterior fusion of cervical spine, when the blood pressure fell to 80/40 mmHg and heart rate fell to 80/min. The surgeon was immediately informed. Blood pressure and heart rate rose to 120/60 mmHg and 100/min respectively after readjusting the fusion instruments. Dextrose 5% in normal saline was given for fluid maintenance and Ringer's lactated solution was given to replace the fluid deficit and ECF loss. Blood sugar was normal (102 mg/dl), urine output was 3-6 ml/kg/hr. Estimated blood loss was 250 ml and 200 ml packed red cell were given as replacement. Operation time was about 3 hours. Five minutes after cessation of the diprivan infusion, the patient was warm, fully awake and able to breath spontaneously with normal airway protective reflexes. The patient was then extubated and transferred to the intensive care unit for close monitoring. Postoperative pain was relieved by intravenous tramadol 10 mg bolus and 2 mg/hr infusion. Ondansetron 1 mg was given intravenously every 6 hr to prevent nausea and vomiting. The postoperative period was uneventful.

Ten weeks after surgery, the halo vest was removed. Lateral radiographs of the cervical spine in flexion and extension positions demonstrated a solid fusion mass at occiput - C_3 and no obvious movement of C_1 - C_2 vertebrae. The patient could walk without assistance and she was discharged from the hospital 2 weeks later. She was scheduled for serial follow - up and radiograph examination.

DISCUSSION

In 1929, Morquio and Brailsford independently reported cases characterized by short - trunk dwarfism, progressive spinal deformity, and normal intelligence. The incidence is approximately 1:76,000 - 216,400. Morquio syndrome is one of the mucopolysaccharidoses, which are a group of lysosomal storage disorders. This syndrome is caused by defective degradation of keratan sulfate resulting in marked excretion of keratan sulfate in the urine. This autosomal recessive mucopolysaccharidosis occurs in a severe form (type A) and a milder form (type B). MPS IV A caused by a deficiency of N-acetylgalactosamine - 6 - sulfatase has been mapped to 16q24.3 and MPS IV B caused by a deficiency of B-galactosidase has been mapped to 3p21.3.

Clinical abnormalities

Craniofacial : Mild coarse facies with broad mouth and prognathism.

Eyes : Cloudy cornea which usually appears after 5 - 10 years of age.

Musculoskeletal : Short - trunk dwarfism, pectus carinatum, genu valgus, kyphoscoliosis, waddling gait, ulnar deviation of the wrist, joint laxity, thick and enlarged knee joint. The stance is semi - crouching.

Cardiac : Late onset of aortic regurgitation.

Radiographic findings

General platyspondyly with vertebrae changing to an ovoid shape with an anterior projection. Odontoid process is hypoplastic. The distal ends of the radius and ulna are inclined towards each other. There may be generalized osteopenia.

Most patients have normal intelligence. A severe defect of the vertebrae and odontoid process may result in cord compression or respiratory insufficiency.[1-4]

Patients with mucopolysaccharidosis are likely to have difficult airway management due to significant changes in the shape of the vocal cords and trachea. The tracheal surface area is smaller than in normal patients.[5] Odontoid hyperplasia and atlantoaxial subluxation aggravated this difficulty. A skilled assistant and equipments for difficult airway management were prepared such as laryngoscope with various sizes of blade, an appropriate and small size endotracheal tube, stylets, laryngeal mask airway, fiberoptic intubation and end tidal CO_2 detector. An otorhinolaryngologist and tracheostomy set were available for tracheostomy in case of failed intubation and failed mask ventilation.[6] Precaution should be taken to avoid compression of the cervical spinal cord during positioning or laryngoscopy. Inhalation induction and intubation under deep inhalation was preferred to awake intubation in difficult airway at this age. Succinylcholine should be avoided in case of motor weakness.[7] Sevoflurane was recommended as it induces rapid sleep and awakening. In addition, deep sevoflurane does not depress the myocardium or cause bradycardia.[8,9] Titrated intravenous ketamine and midazolam after topical anesthesia may be an alternative.[6,10] Precautions were taken to avoid compression of the cervical cord during laryngoscopy and positioning by using manual in line stabilization and halo vest immobilization.

During posterior cervical immobilization, there were signs of spinal shock (bradycardia, hypotension) which resolved after surgical adjustment. After the operation had been finished, the decision to extubation depended on adequacy of alertness, ventilation and oxygenation, and presence of airway protective reflex; and none of surgical complications such as bleeding or tissue edema were present. This patient fulfilled the criteria for extubation. Tramadol infusion was used to relieve postoperative pain for 2 days because it is a weak opioid with minimal effects an respiration.[11,12] The common side effects of tramadol are nausea and vomiting which may be deleterious for stability of immobilization. Intravenous ondansetron 100 to 150 mcg/kg every 6 hr is effective in preventing nausea and vomiting.[13,14]

CONCLUSION

Morquio syndrome is a rare congenital syndrome with involvement of several systems. The most important problems include difficult airway management, cervical spine instability and valvular heart disease. Perioperative care by a team approach comprising pediatricians, orthopedic surgeons and anesthesiologists provided satisfactory outcome after posterior cervical immobilization of atlantoaxial subluxation.

REFERENCES

1. Baum VC, O'Flagerty JE, eds. Anesthesia for genetic, metabolic and dysmorphic syndromes of childhood. Philadelphia : Lippincott Williams & Wilkins, 1999: 204-6.
2. Jones KL. Smith's recognizable patterns of human malformation. 5th ed. Philadelphia : WB Saunders, 1997: 466-67.
3. Gorlin RJ, Cohen MM, Hennekam RCM, eds. Syndromes of the head and neck. 4th ed. New York: Oxford University Press, 2001: 131-35.
4. Neufeld EF, Muenzer J. The mucopolysaccharidosis. In: Scriver CR, Beaudet AL, Sly WS, Valle D, eds. The metabolic and molecular bases of inherited disease. 8th ed. New York:Mc Graw-Hill, 2001: 3421-52.
5. Shih SL, Lee YT , Lin SD, Shen CY, Blickman C. Airway changes in children with mucopolysaccharidosis. Acta Radiologica 2002; **43:** 40-43.
6. Wheeler M, Cote' CT, Todres ID , Goudsouzian NG. Pediatric airway. In : Cote' CJ, Ryan JF, Todres ID, Goudsouzian NG, eds. A practice of anesthesia for in-

fants and children, 3rd ed. Philadelphia : WB Saunders, 2001: 79-116.
7. Cooperman LH. Succinylcholine - induced hyperkalemia in neuromuscular disease. JAMA 1970; **213:** 1867-71.
8. Kern C, Erb T, Frei FJ. Hemodynamic responses to sevoflurane compared with halothane during inhalation induction in children. Pediatr Anaesth 1997; **7:** 439-44.
9. Cote' CJ, Lugo RA , Ward RA. Pharmacokinetics and pharmacology of drugs in children. In : Cote' CJ, Ryan JF, Todres ID, Goudsouzian NG, eds. A practice of anesthesia for infants and children, 3rd ed. Philadelphia: WB Saunders, 2001:121 -71.
10. Wheeler M, Ovassapian A. Pediatric fiberoptic intubation. In : Ovassapian A, ed. Fiberoptic endoscope and the difficult airway, 2nd ed. Philadelphia : Lippincott Raven, 1996: 105 -16.
11. Lehman KA. Tramadol for the management of acute pain. Drugs 1994; **47**: 19 -32.
12. Lee CR, McTavish D, Sorkin EM. Tramadol : a preliminary review of its pharmacodynamic and pharmacokinetic properties, and therapeutic potential in acute and chronic pain states. Drugs 1993; **46:** 313 -40.
13. Davis A, Krige S, Moyes D. A double -blind randomized prospective study comparing ondansetron with droperidol in the prevention of emesis following strabismus surgery. Anesth Intens Care 1995; **23:** 438-43.
14. Furst SR, Rodarte A. Prophylactic antiemetic treatment with ondansetron in children undergoing tonsillectomy. Anesthesiology 1995; **83:** 956 -60.