CASE REPORTS

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The Anesthetic Implications of a Patient with Farber's Lipogranulomatosis

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FARBER'S lipogranulomatosis, inherited as an autosomal recessive trait, was first reported in 1952.[1] There is widespread involvement of the pleura, pericardium, synovial lining of the joints, liver, spleen, lymph nodes, alveoli, and the nerve cells of the central nervous system by this disease. Patients have a deficiency of ceramidase resulting in the accumulation of ceramide in tissues.[2] They present with progressive arthropathy, subcutaneous nodules, psychomotor retardation, and nutritional failure.[1,2] They usually die by 2 yr of age, principally because of problems associated with the airway and ventilation. We report the anesthetic management of a patient with Farber's lipogranulomatosis with a large oral cavity granuloma interfering with her feeding and causing upper airway obstruction.

Case Report

An 8-yr-old girl weighing 15 kg, and 74 cm in height, with the diagnosis of Farber's disease was scheduled for surgical excision of granulomas of the oral cavity and pharynx. The patient had clinical manifestations of the disease, including scoliosis, arthropathy of the neck and wrists, and protruding mouth. Farber's disease was confirmed with light microscopy demonstrating many foam cells in the dermis, and electron microscopy demonstrating large cells with round, cytoplasmic lamellar and microtubular bodies in the lesions on the neck, back, and perianal area.[3] The upper airway had been narrowed by invasion of granulomas into the oral cavity and the pharynx (fig. 1). The granulomas were fragile, and painful to palpation.

The patient was premedicated with 0.2 mg intramuscular atropine. An intravenous catheter for fluid and drug administration had been inserted. Monitoring included electrocardiography, pulse oximetry, capnography, and noninvasive measuring of blood pressure. A precordial stethoscope was used to listen to breath and heart sounds, and a rectal probe was used to monitor body temperature. The technique of awake nasotracheal intubation was explained to the patient, and she was receptive and cooperative during the awake intubation. She could speak and move her hands. The patient received no sedative or opioid. Topical anesthesia of the nasal passage was achieved by applying 1 ml 4% lidocaine using Jackson's spray. Topical anesthesia of the larynx and trachea was achieved by spraying of 1 ml 4% lidocaine through the suction channel of the fiberscope.

A Portex (Kent, England) 4.5-mm ID endotracheal tube was passed through the prepared nostril and into the nasopharynx. After suctioning secretions, a pediatric bronchofiberscope (Olympus 3C20, Tokyo, Japan) was passed through the endotracheal tube into the trachea, followed by the tracheal tube. Intubation was achieved in minutes, with ease, and with no discomfort to the patient. After securing the airway, anesthesia was induced and maintained with 50% nitrous oxide in oxygen and 0.2–1.5% isoflurane. The concentrations of fluoride ion in the plasma and urine were measured during and after anesthesia to monitor renal clearance of fluoride ion. A 20-watt YAG laser was used to excise the granulomas. Although the patient recovered quickly from the anesthesia, the nasotracheal tube was left in place overnight in anticipation of possible pharyngeal and soft tissue edema or postoperative bleeding. The patient was alert the next morning, and was breathing spontaneously. A Portex 8-Fr suction catheter with a Cook J-shaped wire inside was placed through the endotracheal tube into the trachea before extubation. This tube was used for oxygen administration and monitoring of end-tidal carbon dioxide tension after extubation, and would be used as a guide for reintubation if necessary. No wheezing or hoarseness was observed after extubation. The catheter was removed from the trachea after 30 min of monitoring adequate spontaneous ventilation. The patient returned to the ward 3 h after removal of the tracheal catheter.

Discussion

Ceramide can not be converted to sphingosine in patients with Farber's disease because of a ceramidase

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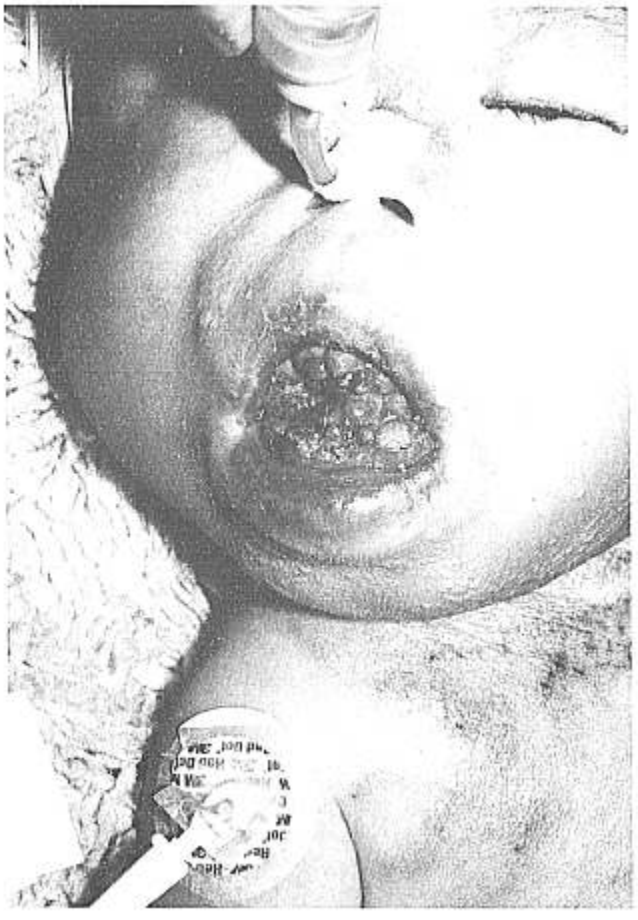

Fig. 1. Patient with Farber's lipogranulomatosis showing granulomas of oral cavity. The patient's neck was fixed, because of the disease, with the face turned toward the right side. Fiberoptic nasotracheal intubation was performed with ease (see text).

deficiency (fig. 2). In patients with Gaucher's or Niemann-Pick's disease, ceramide cannot be produced because of deficiencies of glucocerebrosidase and sphingomyelinase, respectively, resulting in accumulation of glucocerebroside and sphingomyelin. Ceramidase deficiency is typically associated with certain histopathologic and ultrastructural findings[3,4] in the dermal nodules, which were evident in our patient. Tissue cultures[5,6] of fibroblasts or of amniotic cells have also been used for diagnosis of Farber's disease.

A patient with Farber's disease is, potentially, at risk for postoperative acute renal and hepatic failure because of the accumulation of ceramide in the kidney and liver.[1,2,7] Intravenous anesthetic agents were avoided, because the nature of their metabolism and of the pharmacodynamic response to them is not known in patients with Farber's disease. Isoflurane is metabolized less than any inhalation anesthetic (minimal production of free fluoride). Hepatic blood flow and hepatic venous oxygen saturation can be maintained satisfactorily during an isoflurane-N_2O anesthetic.[8] There was no significant difference in plasma fluoride concentration in this patient before anesthesia (1.11 μmol/l) and during 1.67 MAC-h of anesthesia (1.50 μmol/l). The urine fluoride concentration, on the other hand, increased to 25.5 μmol/l at 6 h, and decreased gradually to 14.8 μmol/l 24 h after starting the anesthetic. There is no specific kidney or liver function test to indicate which drug or anesthetic can be appropriately used for a patient with this disease. An inhalational anesthetic was chosen because it was eliminated through the lungs and the dose was controllable. Although blood urea nitrogen and serum creatinine levels were within normal limits in our patient, the renal clearance of fluoride ion on the following day showed about 30% of that in the adult patients with normal kidney and liver functions.[9] Inhalational anesthetics metabolized to a greater extent should be avoided because of the possibility of reduced clearance of fluoride ion in a patient with this disease. We did not use halothane, because epinephrine was used on the surgical field to reduce blood loss in this patient, and because hepatic blood flow may be decreased more during halothane-N_2O anesthesia than during isoflurane-N_2O anesthesia.[8]

Tracheal intubation is best avoided in patients presenting with hoarseness and with laryngopharyngeal involvement of Farber's disease, because postoperative laryngeal edema or bleeding from the granuloma of the larynx is possible. In the patient considered here, however, the use of an endotracheal tube was unavoidable.

Various techniques have been applied in the management of difficult pediatric airways. These techniques include blind nasal tracheal intubation, retrograde wire technique, light wand devices, Bullard laryngoscope, and fiberscopic-aided tracheal intubation.[10–13] Blind nasal intubation is easy to perform in adults; however, it is difficult to perform in small children and, with repeated attempts, the possibility of trauma and bleeding increases, further compromising the airway. A retrograde approach in this patient was inadvisable, because it would be difficult to retrieve the catheter through the mouth or nose.

The light wand technique, which is best achieved using an oral approach, and rigid laryngoscopy, in-

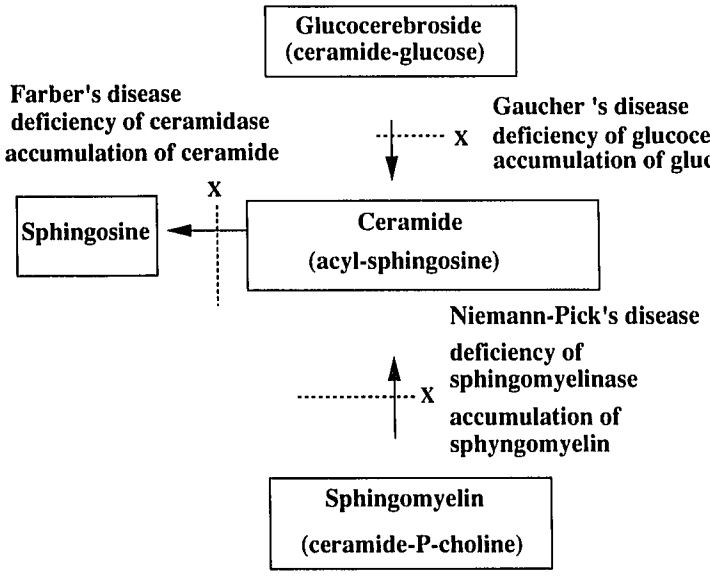

Fig. 2. Ceramide metabolism and the metabolic defect in Farber's disease. Ceramide is accumulated in various tissues because of a deficiency of ceramidase.

cluding the Bullard laryngoscope, were not suitable in this case, because of the painful oral mass and limited mouth opening.[11,12] Fiberscopic-aided tracheal intubation was chosen because it could be used nasally with the least trauma and has a high success rate.[13] In addition to tracheal intubation, the fiberscope provides upper and lower airway evaluation before intubation. This is important from three points of view: 1) any laryngeal or tracheal involvement will be identified; 2) the information will help in planning extubation strategy; and 3) postextubation airway difficulties, if encountered, may be dealt more appropriately with better knowledge and understanding of the airway status. The availability of small-size fiberscopes enables us to apply routine fiberoptic intubation techniques described for adults, in children and neonates. However, if a small-size fiberscope is not available, a two-stage intubation technique using an adult-size fiberscope may be applied.[14] We agree with previous reports that, if anesthesiologists learn and master fiberoptic intubation techniques, there will rarely be a need to use other, often less successful, techniques.[15] The only drawback to the fiberscope is its use in the presence of large amounts of blood or secretions. When intubation is planned and trauma is not the cause of the difficult airway, secretions or blood are not usually a major problem, and are easily managed.

When dealing with a difficult airway, we should also be prepared for reintubation in the event that the patient develops airway obstruction after extubation. Leaving a small-size catheter with J-shaped wire inside the trachea after extubation is an useful approach. The catheter, depending on its size and whether it has an open lumen, can be used for oxygenation, for jet ventilation,[16] and as a guide for reintubation. In our patient, an 8-Fr catheter was used for that purpose. The patient tolerated the catheter well until its removal. If, for any reason, she would need tracheal reintubation, we could have used the catheter and its wire as a guide. Placement of an 8-Fr catheter in the trachea, and leaving it there after extubation, disturbed neither spontaneous ventilation nor phonation by the patient. It proved helpful in providing oxygen to the patient and monitoring her ventilatory status after extubation. Tracheostomy would be the last choice in patients with Farber's disease, because of technical difficulties associated with anatomic deformity in the neck, a high incidence of postoperative infection because of malnutrition, and the possibility of tracheal stenosis after tracheostomy.

In summary, we have described anesthetic management of the first patient with Farber's lipogranulomatosis with a compromised upper airway caused by granulomas invading the oral cavity and pharynx. Associated problems with anesthesia, including management of the difficult airway, are discussed.

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Immediate Detection of Carotid Arterial Thrombosis by Transcranial Doppler Monitoring

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SINCE the introduction of Doppler ultrasound for intracranial applications by Aaslid in 1982,[1] the transcranial Doppler (TCD) has been used by several investigators to monitor cerebral blood flow velocity during surgery for carotid endarterectomy (CEA). Transcranial Doppler velocity as a monitoring technique has been compared to regional cerebral blood flow (rCBF),[2,3] EEG,[4–7] and carotid artery stump pressure.[8] Cerebral autoregulation with respect to changes in blood pressure has been demonstrated in humans using the TCD.[9] In addition to noninvasive, continuous monitoring of middle cerebral artery blood flow velocity (MCAV) during CEA, TCD monitoring also permits the detection of embolic events, shunt malfunction, vasospasm, and the assessment of collateral cerebral perfusion during cross clamping of the carotid.[2,4–8] The following case report serves as an example of immediate detection of internal carotid artery thrombosis by the TCD in the absence of EEG changes.

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