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Anesthesia for a Patient with Gorham's Syndrome: "Disappearing Bone Disease"

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GORHAM'S monocentric massive osteolysis is a rare chronic disease associated with vascular abnormality, angiomatosis, or hemangiomatosis.¹⁻³ The syndrome is

characterized by progressive regional loss of bone with resultant deformity.⁴ We describe the anesthetic management of a patient with Gorham's disease for revision of left pleurosubclavian and placement of right pleurosubclavian shunts to drain chylous pleural effusion during general anesthesia.

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Case Report

The patient was a 15-yr-old, 35-kg, 166-cm boy with a 2-yr history of Gorham's disease diagnosed by bone biopsy and a history of multiple surgeries including placement of Harrington rods for deformed thoracolumbar spine, complicated by T10 paraplegia. The patient had had a left pleurocephalic vein shunt placed 1 yr earlier for left chylous pleural effusion, and the patient manually pumps the shunt (50 times per day) when he becomes dyspneic, with good relief of symptoms. The patient presented with increasing bilateral pleural effusion and respiratory distress due to a nonfunctional shunt. On the morning of surgery, the patient had a respiratory rate of 40 breaths/min and diffuse rhonchi in both lung fields. Arterial blood

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gas analysis revealed pH 7.24, carbon dioxide tension 83 mmHg, and oxygen tension 64 mmHg, with oxygen saturation of 70% during breathing of 2 l/min oxygen *via* nasal cannula. A chest roentgenogram showed several missing ribs on the left and bilateral pleural effusions. Remaining laboratory studies were normal except for a slightly increased hematocrit (50%).

In the operating room, while he was breathing room air, the patient's oxygen saturation was 61% and his end-tidal carbon dioxide greater than 65 mmHg. After preoxygenation with a fraction of inspired oxygen of 1.0, his oxygen saturation increased to 98%. General anesthesia was induced with 225 mg intravenous thiopental, and 20 mg intravenous atracurium was given for muscle relaxation. The trachea was intubated with a 7.0 mm ID cuffed tracheal tube. Anesthesia was maintained with 0.5–1% isoflurane. Ventilation was mechanically controlled. From the right chest, 1,400 ml and, from the left chest, 900 ml bloody chylous fluid was removed. At the end of the operation 30 mg intravenous ketorolac was administered.

The patient's trachea remained intubated, and he was taken to the pediatric intensive care unit. Approximately 4 h after the surgery, the patient was separated from ventilator support. After 20 min of spontaneous breathing with 5 mmHg of continuous positive airway pressure, his end-tidal carbon dioxide was 59 mmHg, and oxygen saturation was 88% with a fraction of inspired oxygen of 0.4. It was decided to remove the tracheal tube. Because of hypoventilation, however, the trachea was reintubated 6 h later. Mechanical ventilation was required for 11 days, after which the trachea was extubated with a resulting arterial carbon dioxide tension of 64 mmHg and arterial oxygen tension of 87 mmHg during breathing of 40% oxygen *via* a face mask.

Discussion

Gorham's monocentric massive osteolysis is a rare and specific type of bone destruction associated with benign, noncystic osseous angiomatosis and characterized by a progressive localized resorption of bone.^{1,5-7} The disease occurs most commonly in adolescence and in the 2nd and 3rd decades, although the age range of those affected is 1.5–72 yr.^{7,8} It occurs with equal sex distribution; with no racial predilection; and with no associated endocrine, metabolic, neoplastic, infectious, or neurologic disturbance.⁷ In comparison to hemangioma, disappearing bone disease will completely replace the bone with fibrous tissue. The disease sometimes is self-limiting but often results in severe deformities⁹ and may be fatal.^{2,3,10-14} Diagnosis is by exclusion based on combined clinical, roentgenographic, and histopathologic findings.^{9,15-20}

Patients usually present with mild aching pain and a pathologic fracture, but deformity may be the only complaint. Pain may be completely absent. Chylous pleural effusion may be present secondary to compression or obstruction of the thoracic duct.^{2,14,21} Cystic lymphangioma may be present.²²

Anesthetic considerations for this disease include adherence to precautions related to cervical spine involvement and the implications of tracheal intubation. Therefore, cervical spine roentgenograms are warranted to rule out pathologic processes such as subluxation and to determine whether extension of the neck is feasible. If there are signs and symptoms of cervical spinal cord compression, awake fiberoptic intubation should be considered.

The disease is not associated with kidney or liver dysfunction. Hence, the doses and timing of administration of medications are not routinely modified. Succinylcholine was avoided in our case because of T10 paraplegia. Moreover, because of unpredictable fasciculation after succinylcholine, the possibility of fractures in weakened and osteoporotic areas of the bone remains a concern. Hence, succinylcholine may be relatively contraindicated.

As illustrated by this case, postoperative ventilatory problems may arise. Unfortunately, in our case, preoperative pulmonary function testing was not available. As the disease progresses, however, patients may retain carbon dioxide, and the need for postoperative ventilation should be anticipated.

In conclusion, we present the first report of the anesthetic management for a patient with Gorham's syndrome. Surgical correction of deformities requiring procedures such as placement of Harrington rods may be necessary in patients with Gorham's syndrome, and principal concern should be directed to possible spinal cord compromise and pulmonary dysfunction.

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Muscular Spasm in the Lower Limbs of Laboring Patients after Intrathecal Administration of Epinephrine and Sufentanil

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THE use of intrathecal opioids for the management of labor pain has the advantage of providing analgesia for patients in labor without producing motor or autonomic blockade. In addition, intrathecal opioids produce no demonstrable deleterious fetal or neonatal ef-

fects.¹ Combinations of opioids such as fentanyl and morphine have been reported to give rapid-onset and prolonged analgesia for patients in labor.² Sufentanil provides an extremely rapid onset of analgesia and a duration comparable to that of the morphine-fentanyl combination.³ Additional work has demonstrated that intrathecal sufentanil produces more profound analgesia than does intrathecal fentanyl in the management of labor pain.⁴

Epinephrine has a long history of use as an adjuvant to intrathecal anesthesia in obstetrics. Recently, in our initial management of patients in labor, we have used 10 μg intrathecal sufentanil in a double-needle technique similar to that used for cesarean delivery.⁵ When the parturient patient requests additional analgesia, epidural analgesia is administered. Because it has been demonstrated that the addition of 300 μg epinephrine to 50 μg sufentanil, epidurally administered, prolongs analgesia,⁶ we thought that the addition of 200 μg epinephrine to 10 μg spinally administered sufentanil would prolong the action of the sufentanil and also

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Key words: Analgesia; intrathecal; obstetric. Analgesics, intrathecal: side effects; sufentanil. Complications: muscular spasm. Sympathetic nervous system, catecholamines: epinephrine.