Anesthetic considerations in a parturient with Freeman–Sheldon syndrome

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ABSTRACT
Freeman–Sheldon syndrome is a rare genetic disorder characterized by malformations of the face, oral cavity and musculoskeletal system. This case report describes the anesthetic management of a parturient with Freeman–Sheldon syndrome, kyphoscoliosis and a cardiac pacemaker for a cesarean delivery and tubal ligation. With a predicted difficult airway, our team decided to provide a combined spinal–epidural anesthetic. Problems encountered included difficult intravenous access, failure to identify the subarachnoid space and patient discomfort during surgery.

Keywords: Freeman–Sheldon syndrome; Cesarean delivery; Anesthesia; Combined spinal–epidural

Introduction

Freeman–Sheldon syndrome (FSS), also known as cranio-carpal-tarsal dysplasia, Windmill-Vane-Hand syndrome, distal arthrogryposis type 2 and whistling face syndrome, is a rare congenital myopathy disorder.[1] First reported in 1938, it is described by facial and skeletal abnormalities, the most common being microstomia with pouting lips, camptodactyly, ulnar deviation of the fingers and talipes equinovarus.[2] Facial characteristics include micrognathia, microglossia, high arched palate, vertical skin folds in the jaw and 'mask-like' facies.[3] Other features include dental crowding, strabismus and hearing loss. Kyphoscoliosis presents later in life. General health, mental intelligence and life expectancy are usually unaffected.[4]

Complications during anesthesia have been reported in individuals with FSS. The most common complication is difficult intubation.[5,6] Malignant hyperthermia (MH), muscle rigidity and pyrexia have been reported in patients receiving MH-triggering anesthesia.[4,5,7–9] We present the anesthetic management of a pregnant patient with FSS undergoing an elective cesarean delivery and tubal ligation. We were unable to find any reports of anesthetic management of parturients with FSS.

Case report

A 36-year-old pregnant woman was seen in consultation at 36 weeks of gestation for elective cesarean delivery and tubal ligation. Her past medical history was significant for FSS, bipolar disease and cannabis use. She described numerous orthopedic surgeries to her distal lower limbs as a child but was unsure of the anesthetic technique. Bilateral hip arthroplasties were performed 5–10 years previously at another institution. Records obtained after the cesarean delivery showed they were performed under spinal anesthesia. Oral maxillofacial surgery to the patient's upper palate and jaw had been completed as a child. Facial trauma when she was 18 years old necessitated wiring of her jaw for stabilization. The patient was unable to recall the type of anesthesia delivered, and had no recollection of any anesthetic complications.

The patient was scheduled for an elective cesarean delivery due to concerns about hip dislocation if a vaginal birth were attempted. The patient also requested tubal ligation. A previous pregnancy ended in a spontaneous abortion that did not require surgical intervention. Gestational diabetes was diagnosed during pregnancy and was managed by diet. Prenatal ultrasound did not detect any fetal abnormalities but the fetus was small for gestational age and had a two-vessel umbilical cord.

A ventricular pacemaker had been inserted 10 years previously for arrhythmias from chronic methamphetamine use. Pacemaker function and battery life had recently been checked. A recent echocardiogram showed no obvious abnormalities with a left ventricular ejection fraction of 60–65%. Current medications were prenatal vitamins, ranitidine, pyridoxine and doxylamine. Acetaminophen was used as needed. She did not require medications for bipolar disease.

At the time of assessment her weight was 55 kg, height 142 cm (body mass index 27.3 kg/m^2) and vital

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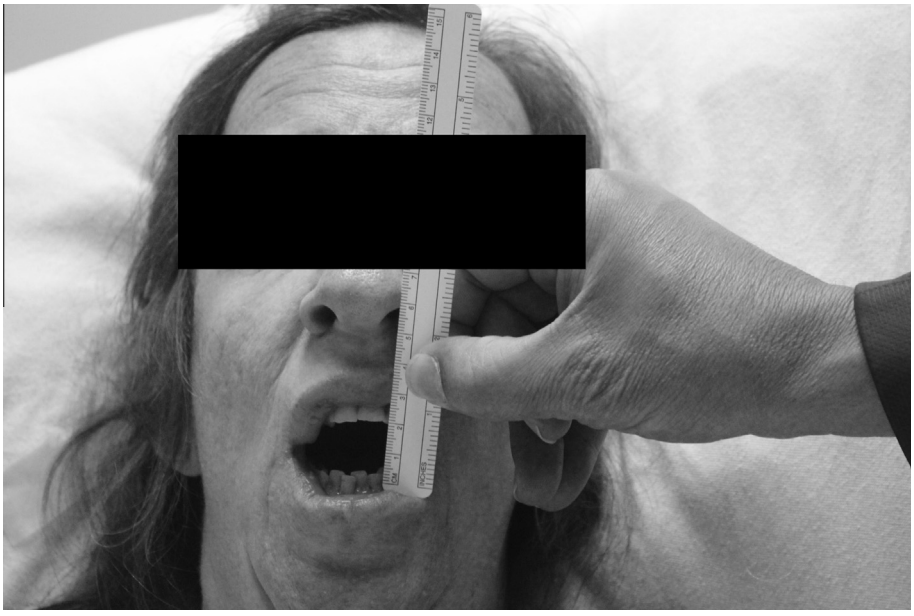

Fig. 1 Photograph demonstrating patient's limited mouth opening

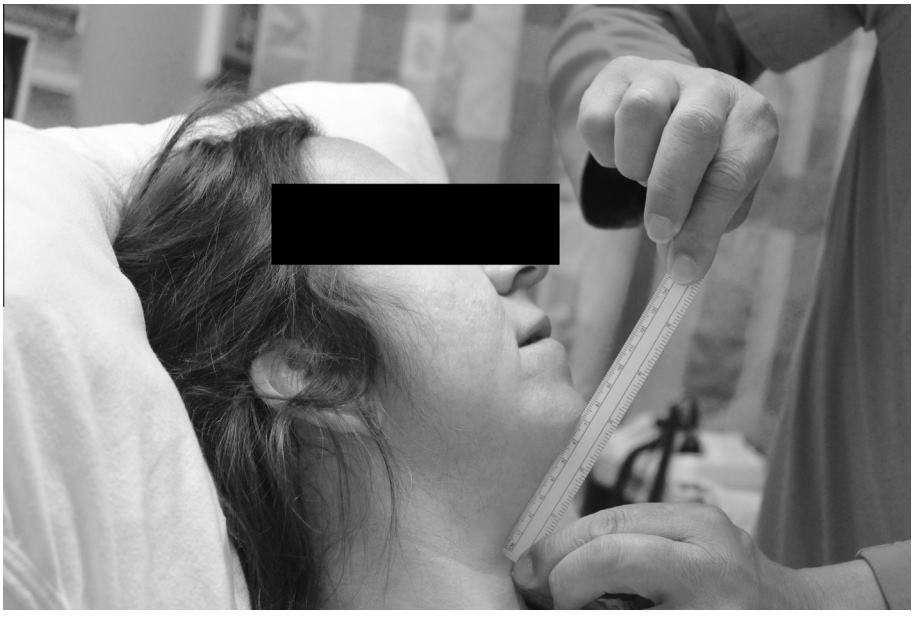

Fig. 2 Photograph demonstrating shortened thyromental distance

signs were normal. Physical findings related to FSS were camptodactyly, ulnar deviation of the fingers, contractures in distal extremities, small nose, lengthened philtrum and masked facies. The characteristic microstomia with pouting lips had been repaired, and she had severely limited mouth opening of approximately 2 cm (Fig. 1). She had a Mallampati III score, poor dentition, shortened thyromental distance of 3 cm (Fig. 2) and could not protrude her jaw. Cervical spine movement was adequate. We concluded that intubation would be difficult, but that bag mask ventilation would be possible.

Contractures in the patient's upper and lower limbs, in conjunction with decreased muscle mass, suggested difficult intravenous access. Cardiac and respiratory examination was unremarkable. Examination of her back showed a mild lumbar kyphoscoliosis with curvature to her left side. The spinous processes were palpable and she was able to maintain a position for placement of neuraxial anesthesia.