Journal of Anesthesia & Clinical Research

Nabatame et al., J Anesth Clin Res 2013, 4:11
DOI: 10.4172/2155-6148.1000368

Case Report Open Access

Anesthesia for Emergency Cesarean Section in a Parturient with Noonan Syndrome

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Abstract

Patients with Noonan syndrome are characterized by many anesthetic problems, primarily related to potential difficult airway and technical issues with regional anesthesia. We describe the anesthetic management of a difficult case of a parturient with Noonan syndrome requiring emergency Cesarean section under spinal anesthesia. We had to evaluate the patient within a limited amount of time because of fetal distress. Although spinal injection was easily accomplished, it resulted in inadequate sensory block requiring supplemental analgesics and sedatives.

Keywords: Noonan syndrome; Emergency cesarean section; Spinal anesthesia

Introduction

Noonan syndrome is an autosomal dominant, multisystem disorder caused by missense mutations in genes involved in the Ras/MAPK signaling pathway and its prevalence is 1 in 1000-2500 individuals [1,2]. Noonan syndrome is diagnosed on the basis of the clinical presentation and genetic testing to confirm the presence of a mutation found in 63% cases [1]. The primary clinical features include facial abnormalities, short stature, congenital cardiac disease, and musculoskeletal malformations [3]. These abnormalities present with many anesthetic problems including difficult airway. The most common cardiac disease in Noonan syndrome is pulmonary stenosis (50-60%), hypertrophic cardiomyopathy (20%), and secundum atrial septal defect (6-10%) [3]. If patient has severe cardiac disease, anesthesiologist should be aware of their impaired cardiac function. Regional anesthesia may be technically difficult with the kyphoscoliosis and lumbar lodosis [4].The epidural and subarachnoid spaces are difficult to detect because of the abnormal curvature of the spine. Six cases of anesthetic management in a parturient with Noonan syndrome under both general and regional anesthesia have been reported [4-9]. Although previous reports have suggested that anesthesiologists should evaluate a patient early during the prenatal period we had no opportunity to assess the patient until emergency Cesarean section was selected, and we had to deal with issues associated with anesthesia as they arose [4-8]. This report describes the management of spinal anesthesia for emergency Cesarean section in a parturient with Noonan syndrome and reviews the existing literature.

Case Report

A 30-year-old primigravida (height-147 cm, weight-42 kg) presented to the obstetric department of our hospital for perinatal care at 10 weeks' gestation. She was known to have Noonan syndrome, the characteristics of which were as follows: typical flattened face, short neck, kyphoscoliosis, history of congenital cardiac defects, ptosis, nystagmus, dental extractions (frontal teeth), and short stature. Genetic testing was not performed. Her family history was unremarkable, and her case was considered sporadic. There was no bleeding disorder and her mental development was normal. She was diagnosed with atrial septal defect and pulmonary valve stenosis at 3 and 9 years of age, respectively, and the course of both conditions was unremarkable during adolescence. Five years ago, she had low back pain due to kyphoscoliosis for which she wore a lumbar corset (Figure 1). She was referred to a cardiologist for evaluation of the cardiovascular risks during pregnancy. Her electrocardiogram showed transient ectopic P-waves and bradycardia (heart rate 49 beats/min). Chest X-ray was normal (Figure 2). Transthoracic echocardiography showed no abnormal shunting and normal left ventricular function. After consulting the cardiologist, the obstetricians planned a normal vaginal delivery. The pregnancy course was uneventful. At 40 weeks and 3 days' gestation, the parturient was admitted due to early labor. Laboratory tests were within the normal limits, and no bleeding tendency was detected. Her body weight was 48.8 kg. The next day, because of non-progress of labor and fetal bradycardia, emergency Cesarean section was decided.

Approximately 25 min after the decision was made; the patient arrived at the operating room. On arrival, her arterial blood pressure was 116/57 mmHg and heart rate was 81 beats/min. Under standard non-invasive monitoring, a 25-gauge Quinke needle was inserted in the L3-L4 interspace by a midline approach with the patient in the left lateral position. Spinal anesthesia was induced with 0.5% hyperbaric

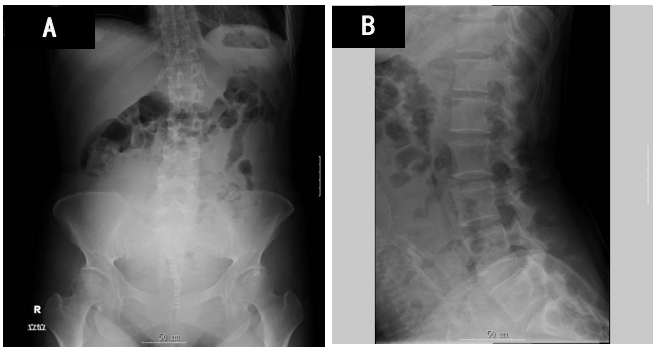

Figure 1: Standing lumbar spine anteroposterior (A) and lateral (B) X-ray images in the nonpregnant state showing kyphoscoliosis.

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Received October 17, 2013; **Accepted** November 27, 2013; **Published** November 29, 2013

Citation: Nabatame M, Hamada T, Tanaka K, Mori T, Nishikawa K (2013) Anesthesia for Emergency Cesarean Section in a Parturient with Noonan Syndrome. J Anesth Clin Res 4: 368. doi:10.4172/2155-6148.1000368

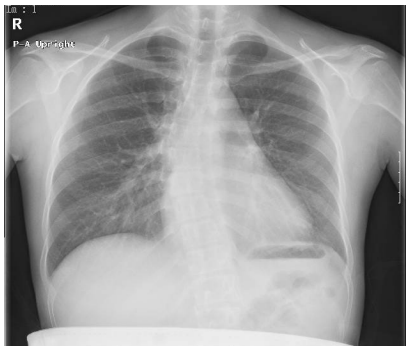

Figure 2: Preoperative posteroanterior chest X-ray showing no abnormal findings in the lung field.

bupivacaine 2.35 ml with morphine 75 μg (total 2.5 ml). Although the sensory block level of cold sensation was unclear, it appeared to have at least reached T4 bilaterally. When the operation was started, the patient complained of pain and discomfort, and nitrous oxide and oxygen (ratio 2:1) were then administered via a facemask. Furthermore, a total of 3 mg of intravenous morphine and 5 mg of midazolam were required to provide adequate analgesia and sedation. A healthy male infant weighing 2432 g was delivered 17 min after administration of the spinal injection. Apgar score was 8 and 8 at 1 and 5 min, respectively. The surgery was uncomplicated and lasted for 47 min, and hemodynamic stability was maintained throughout the procedure. The estimated blood loss volume, including that of the amniotic fluid, was 1410 ml. Although there was no significant adverse effect of midazolam such as respiratory depression at the end of the surgery, we concerned about gastric aspiration and 0.5 mg of flumazenil was intravenously administered for the purpose of safety. The sensory block level was T3 on the right and T4 on the left. The patient had an uneventful recovery without neurological complications, and she and the infant were discharged on postoperative day 6. Neonatal follow-up after 1 month suggested that the infant was not affected by Noonan syndrome.

Discussion

Patients with Noonan syndrome present with various kinds of anesthetic problems including difficult airway, limited cardiovascular reserve, technical difficulties with regional anesthesia and increased risk of neuraxial hematoma [3]. In patients with mental retardation, it is difficult to perform awake procedures as well as obtain their consent whereas in those with lymphedema and redundant skin, intravenous access is difficult [3]. These problems particularly the risk of difficult airway, may exacerbate in pregnant patients.

Detecting the epidural and subarachnoid spaces in regional anesthesia may be difficult because of kyphoscoliosis and lumbar lordosis. Even though the epidural space can be identified, catheter insertion would be complicated. Three cases of regional anesthesia in parturients with Noonan syndrome were reported [4,6,7]. In the first report, epidural anesthesia was unsuccessful because of technical difficulty in locating the epidural space, but spinal anesthesia was successful [4]. The second reported uneventful spinal anesthetic management while the third described that epidural labor analgesia was easily performed without any complications [6,7]. Despite the existence of technical difficulties, regional anesthesia has the advantage of avoiding the risk of difficult intubation and allows patients to remain awake during delivery. However, facilities for emergency and fiberoptic intubation should be available in all cases. When patients have severe pulmonary stenosis or other cardiac impairments, general anesthesia with invasive monitoring will be beneficial.

A history of abnormal bleeding or bruising are often found in patients with Noonan syndrome (56-74%), related to factor XI or XII deficiency, and Von Willebrand's factor or platelet dysfunction [5,8]. Bleeding tendency also indicates general anesthesia to prevent epidural hematoma.

Many of typical facial features of Noonan syndrome suggest a difficult airway because of their high arched palate, short webbed neck and micrognathia [5]. When general anesthesia is chosen, awake oral fiberoptic intubation is the safest option in the case of potential airway difficulties, provided the patient's consent is obtained [8].

Chase et al. reported successful rapid-sequence induction and endotracheal intubation using videolaryngoscopy for a Mallampati class 2 parturient with Noonan syndrome at 23 weeks' gestation [9]. However if physical examination shows higher Mallampati score, rapid-sequence intubation will be more difficult, and awake intubation using direct laryngoscopy or video laryngoscopy with topical anesthesia would become alternative methods [5]. The nasal approach should be avoided in patients with bleeding tendencies or severe facial abnormalities.

In our patient, we had to address the problems of both potential difficult airway and technical difficulties with regional anesthesia. Furthermore, the time available for preoperative evaluation and preparation was limited because of fetal distress. Although lumbar spine X-rays showed mild kyphoscoliosis, her cardiac status was stable and she had no bleeding tendency. Thus, spinal anesthesia was chosen. When the surgery started, the patient complained of pain and discomfort with the sensory block level having reached T4. Possible reasons for failed spinal anesthesia in this case were inadequate intrathecal spread of local anesthetics due to anatomical abnormality or technical issues and error in testing the block level. Abnormal spinal curvature (i.e. kyphoscoliosis) is considered to have restricted the spread of the injected drug, resulting in an unexpected lower sensory block level, but this cannot explain the inadequate analgesic effect with the block level reaching T4. Fettes et al. used the term patchy block to describe a block that appears adequate in extent but where the sensory and motor effects are incomplete [11]. Although most likely reason for patchy block is misplaced local anesthetics or insufficient dosage, anatomical abnormality could be one potential cause [11].

There is little possibility of misplaced injection because we always rotate the needle in 360 degrees after the initial appearance of Cerebrospinal Fluid (CSF) and check CSF flow in all directions to minimize the possibility of needle tip displacement. While using a higher dose of bupivacaine could guarantee an adequate effect in standard 'single shot' techniques it would be difficult to avoid adverse effects such as hypotention and patient discomfort [11]. Although no data regarding the appropriate dose of local anesthetic for spinal or epidural anesthesia in patients with spinal deformities is available our conventional dose of hyperbaric bupivacaine with morphine appears to be adequate [9].

Next, precise testing of the block level may be difficult in emergency situations. Our patient was uneasy and confused about the emergency surgery and her response to cold stimuli was unclear. There are two options in cases of failed block after skin incision, i.e., conversion to general anesthesia with tracheal intubation and continuation of spinal anesthesia using systemic supplementation we were able to complete the surgery using additional analgesics and sedatives [11]. If we had

enough time to discuss anesthetic procedures with the patient before the surgery, we could have considered alternative methods such as combined spinal and epidural anesthesia (CSE) or general anesthesia with awake intubation.

Developments in reproductive medicine and cardiac surgery may increase the number of parturients with Noonan syndrome requiring anesthesia for delivery. When such patients are presented to the hospital, even though vaginal delivery is planned, they should be referred to the pre-anesthesia clinic during early pregnancy for adequate preoperative evaluation in case emergency Cesarean section is required. Further cooperation between obstetricians and anesthesiologists is essential to obtain a successful outcome for both the parturient and fetus.

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