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Case Report

Anaesthetic management in adolescent fraser syndrome

Kriti Nagar [1,*]

[1] *Dept. of Anesthesiology, Ram Manohar Lohia Institute of Medical Sciences, Lucknow, Uttar Pradesh, India*

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ABSTRACT

Fraser syndrome is a rare autosomal recessive genetic disorder characterized by cryptophthalmos, syndactyly, genital malformations, renal abnormalities, musculoskeletal abnormalities and mental retardation. We present a rare case of one such adolescent girl who presented with hematometra and was operated for the same. We hereby discuss the anaesthetic management of this patient with emphasis on points that may prevent any inadvertent complications due to associated co-morbidities.

1. Introduction

Fraser syndrome is a rare autosomal recessive genetic disorder characterized by cryptophthalmos, syndactyly, genital malformations, renal anomalies, musculoskeletal abnormalities and mental retardation. It is diagnosed on clinical examination and perinatal autopsy. It was described by Pliny The Elder, and first published in 1962 by a Canadian geneticist named George R. Fraser.[1] It has an incidence of less than 0.043 per 10,000 live births and 1.1 per 10,000 stillbirths.[2]

We report a case of Fraser syndrome with urogenital sinus with hematometra scheduled for hematometra drainage and sigmoid vaginoplasty surgery.

2. Case Report

A fourteen-year-old female presented to the hospital with chief complaint of cyclical abdominal pain for two months. Pain was spasmodic in nature, restricted to hypogastrium, non-progressive, subsided on taking pain medications and usually lasted for 2-3 days. She was born after full term normal vaginal delivery in a non consanguinous marriage, cried three hours after birth, no history of mechanical ventilation after birth, immunization complete, all milestones achieved as per age. No history of cyanotic spells, physiological jaundice, pneumonia or seizure disorder. She was diagnosed to have Fraser syndrome at birth based on clinical features and was operated on five different occasions for cryptophthalmos of one eye at the age of 8 years (no documents available). No history of any drug allergies.

On examination, she had cryptophthalmos of right eye, congenital cataract and ptosis in left eye, depressed nasal bridge, absent alar nasi, low set ears, hair growth extending from forehead to eyebrows, tongue tie present, uvula deviated to the right side, tongue depressed on the right side, and malocclusion of dentition was present. Mouth opening was adequate, Mallampati grade I, neck movement adequate, thyromental distance > 3 fingers. No pallor, icterus, clubbing, cyanosis, lymphadenopathy or oedema present. Her heart rate was 94 beats per minute,

* *Corresponding author.*
E-mail address: kriti.1188@gmail.com (K. Nagar).

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Blood pressure 114/68 mmHg, SpO2 on room air was 99%, body temperature 98.6ºF. Height was 145cms and weight as 25 kgs. On systemic examination, no abnormality was detected clinically. Lab investigations (complete blood picture, liver function tests, renal function tests, serum electrolytes, coagulation profile) were within normal limits. Electrocardiogram showed normal sinus rhythm and a 2D Echo done to look for any congenital anomalies of the heart revealed normal study. An ultrasound whole abdomen was done which showed normal size uterus with collection of fluid volume ~~ 7 ml seen in endometrial cavity, with collection of fluid volume ~ 50 ml seen in upper part of vagina (hematometra with hematocolpos [? Imperforate hymen]. A X-ray of lumbosacral spine was done to look for any vertebral anomalies and was found to be normal. Patient was then taken up for the surgery under American Society of Anaesthesiologists grade II.

3. Anaesthetic Management

Prior to the surgery, the risk associated with the surgery was explained and written and informed consent was obtained. She was instructed to stay nil per oral for 8 hours prior to the surgery. In the preoperative room, an intravenous access using 20G cannula was secured and balanced salt solution was started at 2ml/kg/hr. Preoperative vitals were recorded. Thereafter the patient was shifted to the operation theatre where the mode of anaesthesia to be given was chosen as combined spinal epidural anaesthesia. Difficult airway cart was kept ready in case of an emergency. Adequate blood products were arranged for replacement of intraoperative losses. Non-invasive blood pressure (NIBP), Electrocardiogram (ECG), and oxygen saturation SpO2 monitors were attached. Procedure was explained to the patient. Under aseptic precautions, with the patient in sitting position, 1.5 ml of 2% lignocaine was infiltrated in L2-L3 and L3-L4 interspace. At L2-L3 interspace, epidural space entered with 18G Tuohy's needle with loss of resistance technique and 20G epidural catheter was passed upto 3cm mark through the needle. The catheter was secured in place at 9 cm mark following negative aspiration for blood and cerebrospinal fluid. Test dose of lignocaine with adrenaline (1:200000) 1.5ml was given. A 27G spinal needle introduced in L3-L4 interspace. After free flow of CSF, 0.5% inj. Bupivacaine heavy 1.6ml along with injection fentanyl 12.5 μg was given intrathecally. The patient was moved to supine position and oxygen was administered via Hudson mask. Subarachnoid block was achieved till T6 level. Inj. Midazolam 1 mg given intravenously. The usual side effects of hypotension and bradycardia after subarachnoid block were not noticed. Heart rate was maintained between 80-92 bpm, Systolic blood pressure between 108-118 mmHg and Diastolic blood pressure between 66-76 mmHg. The surgery lasted for about 90 mins and the patient was given an epidural bolus of 0.25% Bupivacaine 6 ml for postoperative pain relief. Patient was monitored for 24 hrs in postoperative ward and epidural catheter was removed the next day as the patient remained pain free. Perioperative period was uneventful.

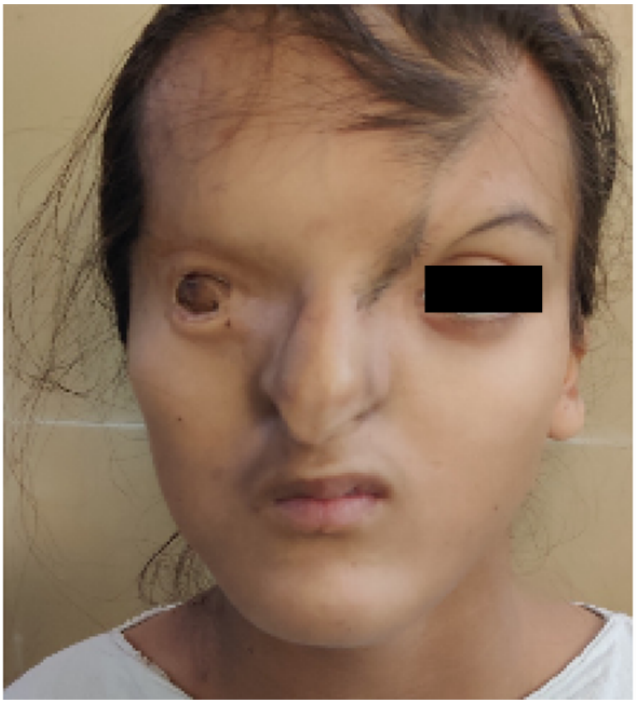

Fig. 1: Facial appearance showing right eye cryptophthalmos, left eye ptosis, low set ears, depressed nasal bridge, hairline extending to the eyebrows

4. Discussion

Slavotinek et al. reviewed 117 cases diagnosed as Fraser syndrome and found cryptophthalmos in 88%, syndactyly in 61.5%, ambiguous genitalia in 17.1%, ear malformations in 59% and renal agenesis in 45.3%.[3] These patients may present with atrial and ventricular septal defect, dextrocardia, univentricular heart, a variant of Ebstein anomaly, coarctation of the aorta, and transposition of the great vessels.[4,5] Meningoencephalocele and spina bifida occulta are some of the common neurological anomalies associated with Fraser syndrome. Other less commonly reported abnormalities are hydrocephalous, encephaloceles, mild cerebellar hypoplasia, and periventricular leukomalacia. Renal agenesis with or without ureter agenesis or cystic dysplasia of the kidney and abnormalities of the urinary tract may present with cryptophthalmos.[3,4]

Although various aspects of this syndrome have been reviewed, there is a paucity of literature on anaesthetic management in Fraser syndrome. In our patient, owing to anticipated difficult mask ventilation due to facial dysmorphism and to avoid unnecessary manipulation of

an anticipated difficult airway, a choice of regional anaesthesia was decided upon. Jagtap et al[6] in their case report described management of abdominal hysterectomy in an adolescent with Fraser syndrome under general anesthesia. They attempted retrograde intubation due failure in visualization of vocal cords on indirect laryngoscopy, and predisposing cardiac defect posing a risk for sympathetic stimulation following direct laryngoscopy. Garg et al.[7] suggested that a check laryngoscopy and airway assessment after induction allowed the safe use of supraglottic airway device without need for tracheal intubation in their patient with Fraser syndrome.

Due to insufficient data available on anesthetic management in a child with Fraser syndrome, all preparation for a difficult airway management and professionals skilled in surgical airway access are required prior to any attempt at airway handling owing to a high incidence of laryngeal anomalies.

5. Conclusion

Patients with Fraser syndrome may pose a challenge for the anaesthesiologists due to its associated co-morbidities. Hence, a thorough assessment of the airway and investigations to rule out any systemic involvement must be carried out before proceeding for any anaesthetic intervention.

6. Source of Funding

None.

7. Conflict of Interest

None.

8. Acknowledgement

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Author biography

Kriti Nagar, Assistant Professor https://orcid.org/0000-0002-2227-3144

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