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CASE REPORT

A child with Russell–Silver syndrome (RSS) undergoing surgery for congenital cataract: Case report

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ABSTRACT

Background: Russell–Silver syndrome (RSS) is an uncommon but well-known imprinting condition primarily characterized by postnatal development failure and idiopathic intrauterine growth retardation (IUGR) and an inverted triangular face and a prominent forehead with relative macrocephaly that distinguish it from idiopathic IUGR and other causes of postnatal growth failure. Few case reports of RSS with cleft palate have been published and those who have perioperative issue such difficult intubation owing to trismus and difficulty to use a mouthpiece due to mandibular development failure.

Case presentation: Female child with RSS was subjected to cataract surgery performed under general anesthesia. Despite limited mouth opening and short thyro-mental distance, the intubation was relatively easy. The patient was extubated and moved to the postoperative care area. Postoperative interval passed uneventful.

Conclusion: In RSS patients, anesthetic considerations of difficult airway and pharyngeal edema must be avoided as a perioperative complication using pediatric fibro-optic device.

SUMMARY OF THE CASE

The rare condition Russell–Silver syndrome (RSS) is characterized by intrauterine growth restriction, poor postnatal growth, a big head, triangular-shaped face, and a prominent forehead (seeing the face from the side), body asymmetry, and major issues in feeding. It can be diagnosed clinically and by genetic mapping in about 60% of cases. The severity and frequency of the wide range of symptoms vary from one affected person to another. Although most individuals with RSS have normal IQs, common symptoms include movement and/or speech difficulties. This condition affects the anesthesia in many ways including difficult in airway and difficult cannulation.

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Russel–Silver syndrome; congenital cataract surgery

1. Introduction

Silver and Russell originally identified the Russell–Silver syndrome (RSS) in 1953 and 1954, respectively [1]. It took doctors over 20 years to realize that they had seen several facets of the same ailment; first, it was believed that they were discussing distinct disorders. In America and Europe, the illness was, respectively, known as Silver–Russell syndrome and RSS [2].

2. Case report

A 7-year-old female child with RSS was presented to ophthalmology clinic with loss of vision and inability to follow objects Figure 1. On examination, this patient was diagnosed as congenital cataract. On inspection, there was underdevelopment (hemi-hypotrophy) of the left side. Pre-operative evaluation: airway assessment showed underdevelopment of left side of jaw (micrognathia), and there was less than one finger in thyromental distance, so we have had the pediatric fibrooptic device ready for intubation [3]. Chest examination: on inspection showed that there was underdevelopment of left side of the chest. On auscultation, heart sounds and lung fields were normal. Her blood tests came back within the expected ranges, Hb 12.6 g/dl, Hct 42.6%, platelet count 430 × 109/l, and WBC 11.1 × 109/l. Good systolic function was detected by echocardiography with no other anatomical abnormalities. Preoperative monitoring: HR Heart Rate 130 beats/min, RR Respiratory Rate 35 breaths/min, oxygen saturation 98% and there was no signs of respiratory distress and close monitoring to blood glucose level to avoid and manage any hypoglycemia which is common in such patients. No other limb abnormalities except for discrepancy in size between both lower limbs.

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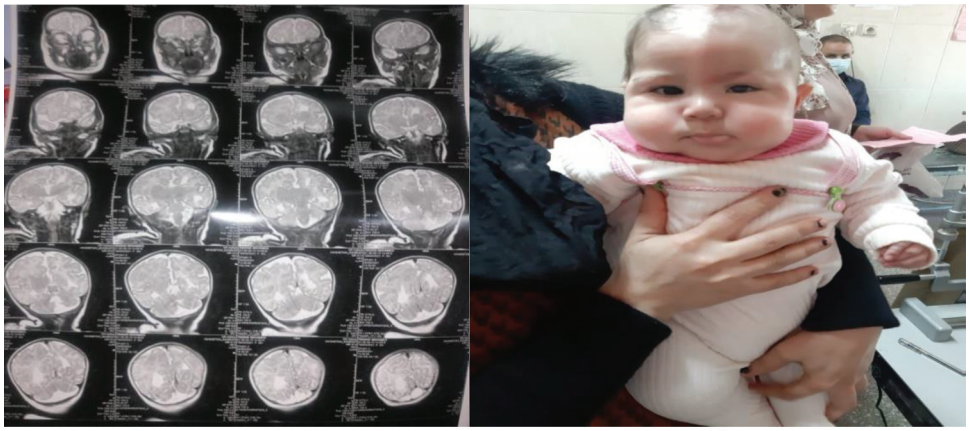

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Figure 1. (a) MRI of the brain showing different sizes of both hemispheres; (b), a photo of the child showing asymmetry of the face including eyes (not obscured) after consenting of the mother of the child.

3. Intraoperative management

Intravenous cannula was inserted inhalational induction with sevoflurane, which was done using Ayer's T piece, and laryngeal mask size 1 was inserted, which was the proper size for that patient depending on the previous airway evaluation and the patient received 1/4 normal saline about 50 ml and 20 ml of paracetamol calculated according to her weight. Maintenance was done by sevoflurane MAC Minimum Alveolar Concentration 3 duration of operation was 30 min. Recovery was smooth without complication.

4. Postoperative management

Patient was sent to the postoperative ward, where she was under constant observation. Her recovery from surgery went without a hitch. Patient received 20 ml of paracetamol as analgesic for her postoperative pain according to her weight.

5. Discussion

RSS patients must have skilled anesthesia treatment for improved surgical outcomes [4]. In our case, because the head circumference is nearly always much greater on the development curve as opposed to either weight or length (a condition known as head sparing/relative macrocephaly), we opted to move through with general anesthesia with a laryngeal mask. This results in the classic triangle face shape seen in children with RSS, in addition to the propensity for a small jaw (micrognathia). Induction was by sevoflurane; it is drug of choice for induction of anesthesia in infant. Asymmetry often just affects arm or leg length, but in certain instances, the entire body may be impacted. As a result, people could have trouble keeping their balance and walking. Craniofacial features: it is known as head sparing or relative macrocephaly when the head circumference is often always substantially bigger than either weight or length in the development curve. This is in addition to the jaw's propensity for being tiny (micrognathia).

6. Conclusion

These patients show a variety of issues; thus, we should administer a secure anesthetic method. Careful preparation of the patient, careful selection of the patient, and discussion of risks and technical issues and extra caution should be exercised during intraoperative and postoperative treatment [5,6]. A lot of these patients may have anesthetic issues in the future.

Disclosure statement

No potential conflict of interest was reported by the author(s).

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