Anesthetic Management of a Patient with Shprintzen–Goldberg Syndrome

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INTRODUCTION

- Shprintzen-Goldberg syndrome (SGS) is a rare syndrome characterized by craniosynostosis, distinctive craniofacial features, skeletal and joint changes, neurologic abnormalities, cognitive deficits, and cardiac anomalies. Due to low prevalence of the syndrome, clinical encounters are limited and therefore, creating an anesthetic plan with expected outcomes may be difficult.

- To help address the lack of anesthesia related information in patients with SGS, we present the anesthetic management of a patient undergoing dental rehabilitation.

CASE DESCRIPTION

- A 22-year-old female with SGS presented for dental rehabilitation. Prior anesthetic history did not reveal any complications. She had moderate mental retardation, dysmorphic facial features, and was Mallampatti 3 with a small mouth opening. The patient was premedicated and subsequently transferred to the procedure room where standard ASA monitors were placed.

- A slow controlled mask induction using sevoflurane, and a mixture of nitrous oxide and oxygen was started. An IV was placed as soon as the patient was asleep. Mask ventilation was easy with an oral airway. Both nostrils were prepared for nasal intubation while the patient was kept spontaneously breathing.

- Before the fiberoptic scope was introduced, a “quick look” with direct laryngoscopy was performed. A grade 3 view was noted but this changed to a grade 2 with cricoid pressure. Propofol and fentanyl were then administered intravenously and the patient was intubated successfully.

- Spontaneous breathing was maintained throughout the procedure using a mixture of sevoflurane, oxygen and air.

- The patient was extubated awake at the end of the dental procedure with no issues and was discharged home a few hours later after meeting discharge criteria.

DISCUSSION

- To date, less than 50 cases of SGS have been reported worldwide. Diagnosing SGS is a clinical process that requires the recognition of a combination of characteristics (Table 1).

- For example, cardiac evaluation and acknowledgement of existing craniofacial and skeletal abnormalities are essential. Potential airway difficulties should be anticipated due to the presence of craniofacial anomalies.

Table 1: Systemic manifestation of SGS

<table>
<thead>
<tr>
<th>ORGAN SYSTEM</th>
<th>CLINICAL FEATURES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Craniofacial</td>
<td>Craniosynostosis, dolichocephaly, high prominent forehead, choanal atresia, maxillary hypoplasia, high narrow palate and micrognathia.</td>
</tr>
<tr>
<td>Cardiovascular</td>
<td>Mitral valve prolapse, mitral regurgitation, and aortic regurgitation</td>
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<tr>
<td>Gastroenterological</td>
<td>Intestinal malrotation, anteriorly placed anus and abdominal hernia.</td>
</tr>
<tr>
<td>Musculoskeletal</td>
<td>Dolichostenomelia, pectus excavatum or carinatum, scoliosis, joint hypermobility, contractures and hip subluxation.</td>
</tr>
<tr>
<td>Neurological</td>
<td>Delayed motor and cognitive milestones, intellectual disabilities, hydrocephalus, and Chiari 1 malformation and cerebral atrophy.</td>
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</tbody>
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CONCLUSION

- Although a rare syndrome, these patients undergo frequent surgical procedures and anesthetics. Thus, it’s good to know such insights as a benefit to provider and patient. As with all patients, a carefully thought out and tailored anesthetic plan, with contingencies, should be followed in patients with SGS.

REFERENCES

3. http://www.orpha.net/com