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Anesthetic Management of a Syndromic Child with Difficult Airway for Debulking of Fibrous Dysplasia: A Case Report

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KEYWORDS

McCune-Albright syndrome, fibrous dysplasia, difficult airway, pediatric anesthesia, craniofacial anomalies

ABSTRACT:

Background: Children with craniofacial syndromes present unique anesthetic challenges due to abnormal facial anatomy, airway involvement, and systemic associations. We report the anesthetic management of a 14-year-old boy with McCune-Albright syndrome and polyostotic fibrous dysplasia involving the mandible and facial bones, who presented for debulking surgery. His history included prior transnasal endoscopic excision and optic nerve decompression. Airway examination revealed limited visualization, flat facial features, and increased intercanthal distance, suggesting possible intubation difficulty. Despite the challenges, careful preoperative assessment, optimization, and preparation of airway adjuncts allowed successful induction and airway management. This case highlights the importance of vigilance, anticipation of airway compromise, and multidisciplinary planning in managing syndromic children undergoing craniofacial surgery.

INTRODUCTION

McCune-Albright syndrome (MAS) is a rare genetic disorder characterized by the triad of polyostotic fibrous dysplasia, endocrine dysfunction, and café-au-lait skin macules. Craniofacial involvement, particularly fibrous dysplasia of the mandible and facial bones, can cause disfigurement, functional impairment, and airway challenges during anesthesia. Children with MAS often require multiple surgeries, and anesthesiologists must anticipate difficult airway scenarios due to altered craniofacial anatomy. This case report describes the anesthetic considerations in managing a syndromic child with fibrous dysplasia for debulking surgery, emphasizing perioperative preparedness and individualized planning.

CASE PRESENTATION

A 14-year-old boy with a known diagnosis of McCune-Albright syndrome presented with progressive left-sided facial swelling and blurred vision in the left eye. He had been previously diagnosed with polyostotic fibrous dysplasia of the mandible and facial bones. One year earlier, he had undergone transnasal endoscopic excision and optic nerve decompression under general anesthesia. On preoperative evaluation, he was clinically stable, with no additional comorbidities. Airway examination revealed mouth opening of more than two finger breadths, Mallampati grade III, and normal neck mobility. His facial features were flat, with increased intercanthal distance, raising concern for a potentially difficult airway. Systemic examination was unremarkable.

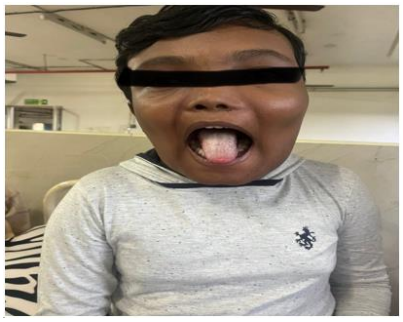

Figure 1

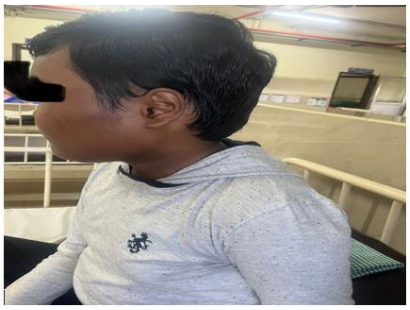

Figure 2

Laboratory investigations showed hemoglobin of 12.6 g/dL, normal coagulation profile, and thyroid-

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stimulating hormone of 5.21 μIU/mL. Electrolytes were within normal range. Echocardiography demonstrated structurally normal heart with good biventricular function and mild mitral regurgitation. CT imaging revealed massive bone expansion with ground-glass appearance involving the calvarium, mandible, and facial bones, consistent with polyostotic fibrous dysplasia. Narrowing of the bilateral orbital fissures and optic canals was also noted.

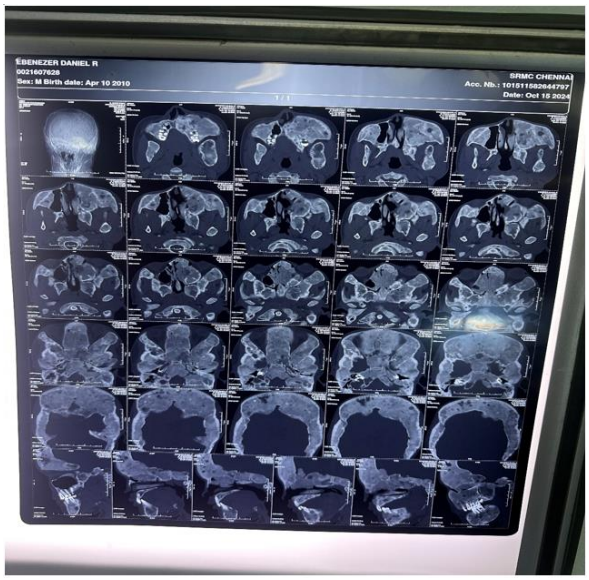

Figure 3

In the operating room, standard monitoring was applied, and the patient was preoxygenated with 100% oxygen for three minutes. Anesthesia was induced with intravenous fentanyl (2 μg/kg), lignocaine (1.5 mg/kg), propofol (2 mg/kg), and vecuronium (0.1 mg/kg) to facilitate intubation. Using a video laryngoscope, the trachea was secured with a 6.5 mm cuffed endotracheal tube orally without difficulty. Anesthesia was maintained with an air–oxygen mixture and sevoflurane as the inhalational agent. The intraoperative course was uneventful, with stable hemodynamics throughout. At the end of the procedure, the patient was extubated smoothly and monitored in the post-anesthesia care unit (PACU) for two hours before being shifted to the ward.

DISCUSSION

Anesthetic management of children with McCune-Albright syndrome requires a multidisciplinary approach due to craniofacial deformities and associated systemic manifestations. The presence of fibrous dysplasia in the mandible and facial bones predisposes to airway obstruction, difficulty in mask ventilation, and challenging intubation. In our case, despite restricted oropharyngeal visualization, the child had adequate mouth opening and neck mobility, which facilitated airway access. However, preparation of adjuncts such as fiberoptic bronchoscope, video laryngoscope, and surgical airway equipment was crucial to ensure safety. Endocrine abnormalities, particularly thyroid dysfunction, should be anticipated in MAS patients. Our patient demonstrated a mildly elevated TSH, emphasizing the need for careful preoperative metabolic assessment. Cardiac involvement is rare but should be excluded through echocardiography, as was done in this case. This case underscores the importance of individualized planning, meticulous airway evaluation, and readiness for alternative airway strategies when managing syndromic children with craniofacial dysplasia.

CONCLUSION

McCune-Albright syndrome with craniofacial fibrous dysplasia presents significant anesthetic challenges. Preoperative airway assessment, anticipation of potential difficulties, and preparation of multiple airway strategies are essential. A multidisciplinary approach ensures safe perioperative care and favorable outcomes in such complex cases.

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