A Case of Costello Syndrome Presenting for Chiari I Malformation Decompression

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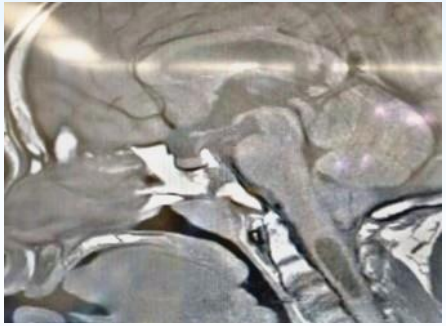
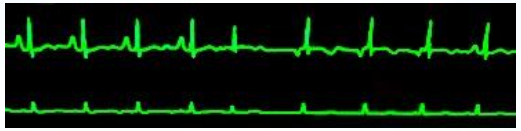
Background

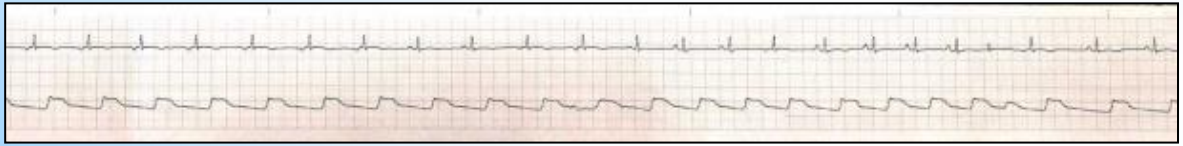
Costello syndrome is a rare genetic disorder with an estimated prevalence ranging from 1:300,000 to 1:1.25 million that can be caused by mutations in the HRAS gene on chromosome 11 which causes cell overgrowth and abnormal cell division. This syndrome has been described in the literature as a faciocutaneousskeletal syndrome that involves growth retardation, developmental delay, metabolic dysfunction, distinctive physical features, dermatologic findings, and cardiac involvement. Typical neurologic findings include seizures, cerebral atrophy, dilated ventricles, and postnatal cerebellar overgrowth which may lead to the development of a Chiara I malformation.

Case Report

We describe a case of a 10 year old 26.3kg female with a history of Costello syndrome who presents for suboccipital craniectomy and cervical laminectomy for Chiari Type 1 decompression and syringomyelia. Her past history includes GERD well controlled on raniditine and precocious puberty requiring supprelin implant. The chiari malformation was diagnosed during routine workup for precocious puberty. A screening echocardiogram had previously been performed that revealed trivial mitral regurgitation and qualitatively normal biventricular systolic function. The patient previously tolerated anesthetics utilizing LMA for MRI and Supprelin implantation. Preoperative airway evaluation revealed a child with limited neck mobility and a Mallampati class III. The patient's vital signs and cardiorespiratory evaluation were otherwise unremarkable. The patient was premedicated with oral midazolam (0.5mg/kg) before proceeding to the operating room. Following an inhalational induction, an IV was placed after multiple attempts and the patient was intubated via video laryngoscopy.







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Discussion

There are significant anesthetic implications to consider in patients with Costello syndrome due to its multisystem involvement. Patients present with a short neck, macroglossia, airway papillomata, and hypertrophied airway tissues. As a result, intubating conditions must be optimized which involves proper positioning, use of muscle relaxants, and appropriate airway equipment. Despite concern for airway obstruction, severe developmental delay often requires premedication. In our case, the decision was made to intubate using a Mcgrath after demonstrating successful mask ventilation. Cardiac abnormalities have also been described in this patient population. Although an old echo was within normal limits in our patient, she was lost to follow up and had not been re-evaluated by cardiology. Intraoperatively, she experienced variable P-wave morphology unrelated to the brainstem manipulation by the surgeon. Electrolyte abnormalities must be ruled out due to the potential for metabolic dysfunction. The perioperative use of TEE and precordial Doppler in addition to invasive monitoring must be considered depending on the surgical procedure. Dermatologic manifestations include excessive skin wrinkling with deep creases in the palms and soles, hyperpigmentation, and acanthosis nigricans. The increased skin laxity in our patient made obtaining IV access difficult. Neurologic findings include seizures, cerebral atrophy, dilated ventricles, and postnatal cerebellar overgrowth which may lead to development of a Chiari I malformation. Patients with Costello syndrome presenting for other surgical procedures may benefit from neurologic evaluation including brain MRI. Additionally, they often exhibit hypotonia that can impact the use of muscle relaxants or increase the risk of post-operative respiratory complications.

Conclusion

Successful anesthetic management of patients with Costello syndrome requires a comprehensive preoperative evaluation and intraoperative plan.

- Preoperative Cardiac Evaluation including ECG and echocardiogram
- Baseline Neurologic Exam
- Potential Difficult Airway: hypertrophied tissues, papilloma
- Difficult IV access due to dermatologic manifestations