Hajdu-Cheney Syndrome and Anesthetic Management
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Abstract

Background
- Hajdu-Cheney syndrome is a rare disorder affecting less than 1/2000 live births. A congenital disorder in connective tissue that leads to severe osteoporosis and bone resorption, demonstrates autosomal dominant inheritance.

Objective
- We report on a patient with mild expression of Hajdu-Cheney. We encountered extremely distorted airway anatomy despite mild expression that could potentially go unforeseen in similarly mild cases.
- To address potential pitfalls in Anesthetic management associated with patients with Hajdu-Cheney syndrome

Case Report
- 11 yr old boy with Hajdu-Cheney syndrome with mild to moderate expression, including short broad neck, low set ears, broad and flattened forehead and coarse thick hair, and noticeable retrognathia, with a very difficult airway to intubate.

Conclusion
- Meticulous preoperative evaluation and workup of all systems is necessary.
- Use of intraoperative adjuncts to narcotic therapy may decrease post-operative complications.
- Severity of syndrome expression may not correlate with potential complications in Anesthetic management

Background
- Hajdu-Cheney syndrome is an extremely rare congenital disorder of connective tissue leading to acro-osteolysis, skull deformities, characteristic facial abnormalities, secondary to significant osteoporosis and bone resorption [1].

Clinical Presentation
This patient was born to non-cosanguinous parents without family history of Hajdu-Cheney. The 40 week pregnancy was uneventful – and mother denied any smoking, alcohol intake or illness – and the patient was delivered without complications during anesthetic delivery. The patient required intubation in the NICU for respiratory distress but was extubated the next day and shortly thereafter discharged home with the family. The patient’s interim medical history is essentially uncomplicated – and the patient was delivered without complications and the awareness of the diagnosis and associated pitfalls is extremely important

References:
- 3) Khan, ZH, et al., A Comparison of the Upper lip bite test with Mallampati Classification in predicting difficulty in endotracheal intubation: A prospective blinded study. Anesthesia and Analgesia. February 2003 vol. 96 no. 2 393-399

Conclusions
- Patients with Hajdu-Cheney present a wide range of phenotypic expression but even a patient with mild expression can present major potential complications during anesthetic delivery
- Baseline cardiac and renal function, and bony and ligamentous instability. Airway anatomy including dentition, aberrant uvular anatomy, macroglossia, and retrognathia, as well as postoperative airway complications among the most common considerations.
- Access to previous operative records are extremely helpful
- Use of dexmedetomidine adjunct intraoperatively may reduce post-operative airway complications and decrease narcotic requirement
- Even mild expression of the syndrome presents major potential complications and the awareness of the diagnosis and associated pitfalls is extremely important