

A Case Report of Anesthetic Management in an Eight-Week-Old Infant with Trisomy 15 Presenting for Resection of Hemangiopericytoma of the Orbit

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Abstract

A difficult airway management situation was presented by an eight-week-old infant who presented with a bleeding orbital tumor. The infant also had a not yet diagnosed inherited trisomy 15 with facial abnormalities and an upper respiratory tract infection that made the face-mask ventilation and tracheal intubation more challenging. The urgent need for the surgery precluded any further work-up and optimization. Anesthetic challenges included difficult mask ventilation, difficult endotracheal intubation, extremely reactive airway and very labile hemodynamics during induction. An Air-Q laryngeal mask airway was used as a rescue airway device and as a portal for endotracheal tube placement during the anesthetic management with good outcome.

Keywords: Infant; Trisomy 15; Hemangiopericytoma; Difficult airway; Hemodynamic instability

Introduction

Hemangiopericytoma was described as a mesenchymal neoplasm in 1942 [1]. It can be benign or malignant; however, involvement of the head and neck has been a rare occurrence. The diagnosis is usually confirmed by imaging studies which typically show a tumor with indistinct margins invading surrounding tissues. The standard initial treatment is complete surgical excision.

General anesthesia care, including tracheal intubation, is challenging in an eight-week-old infant in view of peculiar anatomic and physiologic variations compared with an older child or an adult. This case report presents those challenges, made even more difficult by the infant having an expanding and bleeding orbital hemangiopericytoma, and an undiagnosed chromosomal aberration, namely trisomy 15, with the only suspicious presenting features of micrognathia and low set ears.

Parental consent for publication was obtained as per journal guidelines.

Case

This case involves an eight-week-old female infant scheduled for resection of an orbital mass. Infant was examined in office which revealed friable protruding mass and so surgery was scheduled. Preanesthetic evaluation revealed birth of the infant occurred at 36-weeks of gestation with a low birth weight and a weeklong admission to neonatal intensive care unit soon after birth as a result of feeding difficulties. The Pediatrician's evaluation was negative for any congenital cardiac or other organ anomalies. During initial interview, the mother disclosed that the infant had a runny nose and congestion for the past few days. Physical examination revealed a pale and lethargic looking infant with low set ears and somewhat small chin and no wheezing on auscultation. The infant weighed 3.4 kg with a heart

rate of 150/min, respiratory rate of 35/min and oxygen saturation of 94% on room air. Also present on exam was a protruding and very friable left orbital mass with bloody serous discharge (Figure 1). No preoperative laboratory values were available. In spite of upper respiratory tract infection and lack of baseline laboratory values, the bleeding nature of the tumor warranted immediate attention. An intravenous line was placed and blood samples were sent for hematocrit estimation, blood typing and cross-matching in anticipation of the need for intraoperative blood transfusion.



Figure 1: A protruding and very friable left orbital mass.

In the operating room, American Society of Anesthesiologists (ASA) standard monitors were placed and the infant was preoxygenated for several minutes. Our concern was difficult mask ventilation as a result of the protruding orbital mass and possible difficult tracheal intubation in view of the observed micrognathia. Atropine 0.1 mg was administered intravenously (IV) before induction of anesthesia. General anesthesia was induced with low inspired concentration of sevoflurane about 4% in oxygen followed by slow titration of propofol (up to 2 mg/kg) and fentanyl (up to 1 mcg/kg).

Goal was to maintain spontaneous ventilation until airway was secured. Mask ventilation was established with some difficulty. Direct laryngoscopic examination revealed a Cormack-Lehane grade III view of the larynx and tracheal intubation was not attempted. Thereafter, A Glide Scope was utilized that revealed a grade II view and the trachea was intubated with an uncuffed 3.0 endotracheal tube (ETT). We were not able to confirm tracheal intubation since end-tidal CO₂ was not detected and wheezing and conducted sounds on auscultation precluded appreciation of breath sounds over the lung fields. Concurrently, the infant became bradycardic and oxygen saturation declined to 40%. In view of the uncertain nature of the ETT placement and our prior experience with adequate mask ventilation, the ETT was removed and the infant was treated with positive pressure mask ventilation, epinephrine IV and albuterol inhalation. The infant responded promptly and a decision was made not to attempt another tracheal intubation at this time. A number one Air-Q laryngeal mask airway (LMA) was attempted and was able to establish with an adequate seal. Also we were able to stay away from the friable left eye mass after LMA placement. Anesthesia was deepened with LMA in situ. Decision was made to attempt passing ETT through the LMA as the device was meant for the purpose. An uncuffed 3.0 ETT was placed through the LMA blindly and we were able to confirm its correct placement with end-tidal CO₂ detection and bilateral breath sounds on auscultation. The air Q LMA was a relatively new device in our institution and a "pusher" or stabilizer was not available. Therefore, we carefully wedged another same size ETT to the end of the first ETT and then slowly removed the LMA over both of the tubes. Once again, ETT position was confirmed and it was secured. During surgery, the hematocrit (Hct) laboratory value was estimated to be 19.5; however, blood transfusion could not be initiated for lack of cross-matched blood. Instead, hemodynamic stability was achieved with crystalloids and albumin in a dose of about 15 ml/kg. Following surgery, the infant was transported to the pediatric intensive care unit (PICU), still intubated and with stable vital signs. In the PICU, 50 ml of packed red cells was transfused after cross-matched blood became available. The infant was extubated in the evening after surgery and post-operative course was uneventful.

A recurrence of the tumor was noted by the surgeon within three weeks and again at one year of age, following the initial resection. In both cases, resection of the recurring orbital mass was performed under general anesthesia, and the trachea was uneventfully intubated using a Glide Scope. For the subsequent resections, the infant lacked the challenges of an active upper respiratory tract infection and bleeding-induced hypovolemia. Confirmation of ETT placement was easily achieved.

The infant also underwent genetic testing after the first resection, revealing the diagnosis of trisomy 15.

Discussion

Most pregnancies with trisomy 15 end in early miscarriage. In pregnancies that have progressed, intrauterine growth retardation

(IUGR) and abnormal facial and cranial features are commonly seen. It is not easy to diagnose trisomy 15 in an infant as young as eight weeks since many clinical signs and symptoms are still evolving after birth. In general, physical features of trisomy 15 can include flat nasal bridge, deep-set eyes, low-set ears, high-arched palate, cleft lip and/or palate, hypotonic, variable skin pigmentations and growth retardation. The physical features evolve over a period of time after birth, and the diagnosis is difficult to make as early as eight-weeks after delivery.

The features listed most pertinent to the anesthesiologist are the craniofacial defects, which have been shown to be predictors of potentially difficult intubation [2]. In our case, micrognathia and low set ears were the only presenting features. Trisomy 15 has also been linked to Prader Willi syndrome and Angelman syndrome. This case report shows a rare association of orbital hemangiopericytoma with trisomy 15 as seen in our patient [3]. The aggressive and potentially lethal behavior of this type of tumor resulted in the need for urgent perioperative fluid resuscitation and surgical resection. Its recurrence within a month of the procedure and again a year later is suggestive of very aggressive nature of the disease. Air Q LMAs have had a reported blind intubation success rate of 77%. Though not used at our facility, Fastrach LMAs have a reported success rate of 99% [4]. In the case of intubating through an LMA, removal of the LMA over the endotracheal tube (ETT) can be a difficult task with potential complications, including pilot balloon damage or dislodging the endotracheal tube. Air Q intubating LMAs provide pushers or stabilizing rods to help facilitate LMA removal. In our case, the device was just introduced and the stabilizer was not available. Use of an equal size endotracheal tube to stabilize the placed ETT while removing the LMA is a potentially effective method that is readily available in the OR setting. However, rupture of pilot balloon has been reported. Thus, caution is advised when removing the LMA [5].

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