Anesthesia Management for Blue Rubber Bleb Nevus Syndrome (BRBNS)
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Introduction: Blue rubber bleb nevus syndrome (BRBNS) was described by Bean (1) in 1958 and is a rare syndrome with only 150 cases reported in the world’s literature (2). The syndrome is characterized by cutaneous and gastro-intestinal hemangiomas, although lesions have been reported in the peritoneal cavity, heart, lungs, pleura, eyes and central nervous system (3). BRBNS is often present from birth or early childhood and complications of gastro-intestinal (GI) bleeding are frequent. The management of GI lesions varies depending on the degree of involvement and the severity of GI bleeding (4). We will discuss the anesthetic management of 12 patients undergoing 14 surgical procedures for the removal of gastrointestinal lesions. The design of the study was a retrospective review of medical records of 13 patients.

Methods: Following Institutional Review Board approval, the charts of 13 patients with BRBNS were identified and reviewed. Data collected included demographic data, the surgical procedure performed, surgical time, anesthetic time, anesthetic technique, fluid management, blood loss, urine output, monitoring used, and timing of extubation. Post-operative data included pain control measures and any complications.

Results: The patients ranged in age from 2 to 35 years of age of which 8 were female and 7 were male. The majority of patients were classified as ASA 2 (11 cases) while the 2 remaining patients were classified as ASA 3. Most patients had been conservatively treated initially with repeated blood transfusions. The induction technique was intravenous in all patients. Standard ASA monitors were used and in addition an arterial was placed in 12 of the 14 anesthetics. Anesthetic maintenance was most commonly inhaled isoflurane, fentanyl, and muscle relaxation. Epidural anesthesia combined with general was employed in three patients. The duration of anesthesia varied from 3 to nearly 28 hours. The intraoperative course was stable in most patients with only three cases requiring a blood transfusion despite pre-operative anemia in all patients. Most patients were managed with crystalloid and 5% albumin for intra-operative fluid replacement. One patient required fenoldapam for decreasing urine output despite adequate fluid replacement and one patient experienced a venous air embolism during endoscopy that was treated quickly and the patient recovered without sequelae. Eight patients were extubated in the operating room, while the rest were extubated in the ICU. Post-operative pain control was accomplished with an IV PCA Morphine in the majority of patients. Of the three patients with epidurals, one was removed immediately after extubation because it was ineffective. No other anesthetic complications were noted.

Discussion: While the majority of the cases we presented are without complication and use a standard anesthetic technique there are several issues that require attention. The lesions of BRBNS have been described in nearly every are of the body. Baiocco et al reported a case of BRBNS with lesion predominating in the head and neck (5) and endobronchial lesions were reported by Gilbey et al (6). Careful inspection of the oral cavity and gentle laryngoscopy during intubation are necessary to avoid bleeding from these lesions. Joint lesions may affect range of motion and pressure affects from adjacent lesions may cause bony deformity(2). The importance of this as well as the length of the procedures requires meticulous management of patient position with occasional movement of the patient’s limbs and head if possible. Finally, CNS lesions as well as vertebral body lesions may complicate the placement of an epidural catheter. Garen et al reported a case of spinal cord compression by a lesion on a vertebral body in a previously undiagnosed BRBNS patient(7). Our review showed no complication with epidural placement, but the possible complication of an epidural hematoma must be considered.

References:
4) Ertem D, Blue Rubber Bleb Nevus Syndrome, Pediatrics Vol 107(2), Feb 2001