



Pathology/Biology Section - 2013

G137 Fatal Hemorrhage From a Tracheostomy Site in a Patient With Blue Rubber Bleb Nevus Syndrome: A Case Presentation and Review of Literature

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After attending this presentation, attendees will understand the importance of a thorough examination of the viscera and soft tissues, especially in the head and neck region, and gain insight on potential complications including sudden death from the rupture of soft tissue vascular malformations due to emergency tracheostomies.

This presentation will impact the forensic science community by increasing the awareness of the rare Blue Rubber Bleb Nevus Syndrome (BRBNS) and the risk it presents for fatal hemorrhages depending on its location. A review of literature on the topic of fatalities linked with tracheostomies will be presented.

This case involves a 31-year-old Caucasian female with a medical history of multiple arteriovenous malformations on the right side of her body and a diagnosis of BRBNS. She went to the emergency room because of an infected tooth. Several years prior to her visit to the emergency room, she had had a single episode of bleeding-from-the-tongue-based AVM. Prior to extracting the tooth, she was injected with lidocaine and developed acute anaphylaxis with severe facial and airway swelling resulting in an obstructed airway. An emergency tracheostomy was performed, which was complicated by profuse hemorrhage from the adjacent ruptured soft tissue vascular malformation. Hemostasis could not be achieved by the Intensive Care Unit staff and the patient exsanguinated. The body was brought to the Office of the Cook County Medical Examiner for postmortem examination. An autopsy determined that the immediate cause of death was anaphylaxis due to a reaction to the anesthetic medication that was given prior to the tooth extraction; however, the condition, that significantly contributed to her death was hemorrhage due to a ruptured vascular malformation during the tracheostomy. The uncommon location of an exsanguinating vascular malformation in the neck region in a patient with BRBNS deserves a generous discussion.

Approximately 153 cases of BRBNS have been reported in the literature. What was possibly the first description of BRBNS appeared in 1860 in a paper by Dr. Gascoyne. Dr. Gascoyne described a nevus involving the parotid gland, which caused death from suffocation in a patient who displayed numerous nevi of the viscera. Dr. William Bean coined the name BRBNS in 1958 because of the rubbery, nipple-like texture of the cutaneous hemangiomas that tend to camouflage vascular malformations found in the viscera, and in this case, the neck region. BRBNS has since been reported in patients with numerous and varied types of cutaneous vascular nevi frequently associated with visceral malformations. Most commonly, these visceral malformations are located in the small and large intestines and cause death from hemorrhage. Reports of BRBNS associated with findings of hemorrhaging vascular malformations in the head and neck causing suffocation have also been reported. The association of BRBNS and hemorrhaging vascular malformations of the head and neck has led anesthesiologists to develop various techniques to manage the airway in these patients without disrupting the lesions. Although BRBNS is usually diagnosed in infancy, there are, unfortunately, cases of BRBNS which remain undetected until adulthood. A possible genetic inheritance pattern is currently being studied.

Cases describing prominent head and neck findings in patients with BRBNS emphasize that they often first present to an otolaryngologist or oral surgeon with the incidence of oral hemangiomas as high as 59% to 64%. None of these cases has discussed the risks and potential complications of emergency tracheostomy placement in these patients. This case is the first to discuss a patient with BRBNS who underwent a tracheostomy and developed a fatal hemorrhage due to this rare presentation.

Nevus, Tracheostomy, Hemangioma