

PBLD – Table #30

Complications during Sclerotherapy for Blue Rubber Bleb Nevus Syndrome

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OBJECTIVES:

- Review presentations of patients with arterio-venous malformation
- Discuss management of children undergoing ethanol sclerotherapy in the remote anesthesia setting
- Explore potential complication associated with ethanol sclerotherapy
- Discuss systems-based approaches to decrease the risk of such complications

A Case Study: Assessment and Management of multiple Complications in a Child with Blue Blubber Nevus Syndrome

On a Tuesday morning you are the supervising attending for the pediatric room in the interventional radiology suite. The assignment does not look too difficult: some routine urologic and GI procedures and 2 kids with AV malformation for sclerotherapy.

One of them is a 6-year-old boy with Blue Rubber Bleb Nevus Syndrome. That is a new one for you. You are not sure you have heard of this diagnosis before. He is an adorable young Asian boy with multiple small bluish lesions on his hands and arms. After he changes into a gown you notice that he has a larger lesion involving the lateral aspects of his left leg from the hip to the knee. While his left leg seems to be slightly larger overall this does not seem to affect the young man as he is bouncing around the room while you are trying to talk to his parents

- What is Blue Rubber Bleb Nevus Syndrome?
- How do you assess this child?
- Any particular concerns?

Clearly this child has more than one arterio-venous malformations. In addition he has had several recent episodes of bronchitis but otherwise the parents cannot provide too many details of the medical history: he has been adopted from China just over a year ago. His laboratory values are remarkable for an elevated D-Dimer and fibrinogen levels

- Why does this child have an elevated D-Dimer and fibrinogen levels?
- What are your recommendations for induction and intraop monitoring? Do you ever use more than routine monitoring for these patients?

- Do you think sedation is adequate? Do you need general anesthesia? Any regional techniques?

You decide that general anesthesia is most appropriate in this case as the interventional radiologists may have to turn the child from a supine to a lateral or prone position to obtain better access to the pelvic lesions.

- Are you familiar with the sclerotherapy techniques the interventional radiologists employed at your center?
- Percutaneous/transvascular?
- Agents?

After ultrasonography of the lesion the best site for ethanol injections is identified. When contrast medium is injected there are two problems: the flow through the lesion is pretty rapid and there seems to be a larger draining vein. The needle is adjusted. When four needles are in place and approximately 0.3 mL/kg have been injected you notice that the child's blood pressure has increased from 78/46 to 96/67, then to 108/78. You notice a number of PVC's. The ventilator alarms as you are hitting the upper limit of your PIP.

- What may be going on?
- Airway problem? Cardiac problem?
- How are you going to manage this?
- If this child develops complete cardiovascular collapse do you have adequate resources in the cardiovascular lab?

Fortunately the child stabilizes after your intervention. The radiologists are a bit rattled and decide to move on to another lesion slightly more distal on the same leg. There a multiple peripheral injections made until they decide their procedure is complete. You discontinue your inhalational anesthetic and wait for the patient's emergence...and wait ...and wait. Not only is the patient slow to recover but you notice an obstructive pattern on your ETCO₂ monitor, TV are small. The patient is wheezing.

- How would you have approached this situation?
- What is your differential?

Finally you sent an ABG which shows metabolic and respiratory acidosis. The serum glucose is 60.

- Why does the patient have a metabolic acidosis?
- Could this be a neurologic complication? A stroke?
- How long are you going to wait for the patient to wake up?

Finally, after spending (sufficient) time you are satisfied that the patient meets criteria for extubation. In the pacu the patient is combative. He tries to climb out of bed, needs to be restrained. When you explain to the parent that this may be alcohol intoxication they are flabbergasted. Nobody has ever told them that this could happen...

- Should you have included alcohol intoxication in your preop discussion?
- Is this the responsibility of the radiologist?
- What is your experience in making sure common and not so common complications at the surgery/procedure/anesthesia interface are covered when patients or families provide consent?

When you finally leave the hospital you are “beat”. Your nice, “routine” day has been a nightmare. “Something needs to be done”. In response to this case, you embark on a mission to improve teamwork and communication between the interventional radiology staff and the pediatric anesthesia division..

- Do you have recommendations for preventing similar events in the future?
- Research projects? Communication projects?

DISCUSSION:

What are Vascular Malformations?

Vascular malformations can develop within any section of the vascular tree from the aorta to the capillaries as well as within the lymphatic system. They are developmental abnormalities that often represent a cease in normal angiogenesis or vascular remodeling. Other causative mechanisms include persistent embryonic vessels, aberrant signaling at the molecular level resulting in abnormal proliferation, differentiation, maturation and apoptosis of vascular cells. The incidence of vascular malformation is approximately 1.5% of the population with the majority 90% of these abnormalities present at birth. Venous malformations are the most common with an incidence of 1 in 5-10,000 childbirths. Thus the Hamburg classification was developed in 1988 to classify and subdivide the many different types of vascular malformations based on their vascular origin and further divided by their embryonic stage of development arrest.

Most venous malformations are benign in nature but can cause significant cosmetic deformities. Treatment of extratruncular venous malformations is surgical excision or endothermal ablation for localized lesions, but sclerotherapy has emerged for treatment of both localized and diffuse lesions

Modified Hamburg Classification	
Primary Classification	<ol style="list-style-type: none"> 1. Arterial 2. Venous 3. Arteriovenous 4. Capillary 5. Lymphatic 6. Combined
Embryological Sub classification	<ol style="list-style-type: none"> 1. Extratruncular forms (Diffuse/infiltrating or Limited/localized) 2. Truncular forms
Obstruction or Narrowing	<ol style="list-style-type: none"> 1. Aplasia, atresia or membranous occlusion 2. Stenosis caused by hypoplasia, coarctation, spur or membrane
Dilatation	<ol style="list-style-type: none"> 1. Localized (aneurysm) 2. Diffuse (ectasia)

What is Blue Rubber Bleb Nevus Syndrome?

Blue Rubber Bleb Nevus Syndrome, BRBNS is a rare congenital disorder with only 150 cases reported in the world's literature. The disorder is characterized by multiple cutaneous venous malformations of the skin and soft tissues. These lesions preferentially involve the trunk and upper extremities, can be painful, and can range from a few millimeters to several centimeters in diameter. In addition there can be visceral lesions especially within the gastrointestinal (GI) tract. While these may associated with minor bleeding and iron deficiency anemia there is the potential for life threatening hemorrhage. Most cases are sporadic in nature but some are of autosomal dominant inheritance. It equally affects male and females and has no increased prevalence within any race. Cutaneous lesions are often identified at birth or in early childhood. In early adulthood extracutaneous lesions present usually with GI involvement and present with evidence of GI bleeding. There are no reported cases of malignant transformation of cutaneous lesions. BRBNS has been associated with the development of certain tumors, including medulloblastoma, chronic lymphocytic leukemia, renal cell carcinoma, and squamous cell carcinoma. Because BRBNS is quite rare, the exact risk of development of each of these neoplasms is unknown. There are no reported cases of malignant transformation of cutaneous lesions. While cutaneous lesions are benign they can be cosmetically devastating therefore a guideline was devised to determine treatment strategies for patients with venous malformations.

Sclerotherapy

The most common type of sclerosing agent used in treatment of vascular malformations is ethanol. Ethanol has been used for intra-arterial and percutaneous embolization of AVMs. This can be associated with a variety of complications.

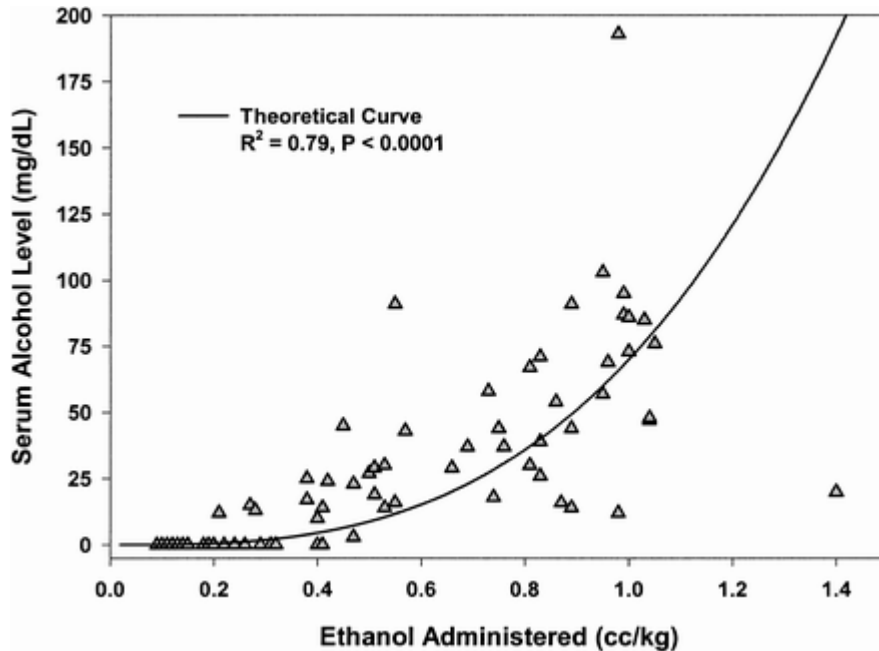
Complications of Sclerotherapy

Complications of sclerotherapy with alcohol are reported in the range of 7.5 to 48%. Local side effects at the site of injection are most common. Systemic effects can reach from mild sedation due to alcohol intoxication to cardiovascular collapse. High blood alcohol ethanol concentrations are associated with cardiac arrhythmias. Serious complications have been observed with relatively minor alcohol injections. Some authors thought that a mild rise in PA pressure is due to systemic stimulation and pain from the ethanol injection as it correlates with a similar rise in the systemic blood pressure. This would not explain more severe pulmonary hypertension. Other authors observed that the rise in pulmonary artery pressure correlates with the alcohol level in the pulmonary circulation which may rise rapidly in AVMs with high flow. Systemic alcohol levels obtained at the conclusion of the case may not be representative of sudden changes in the pulmonary circulation during the case. There is speculation that alcohol in the pulmonary circulation leads to vasospasm via dose-dependent smooth muscle contraction but the exact mechanism is unclear.

Complications

Neurologic	<ol style="list-style-type: none">1. Alcohol intoxication (>1mL/kg)2. Delayed awakening3. PACU agitation4. Transient peripheral nerve injury5. Seizures
Pulmonary	<ol style="list-style-type: none">1. Respiratory Depression2. Bronchospasms3. Pulmonary emboli
Cardiovascular	<ol style="list-style-type: none">1. Increase pulmonary artery pressures2. Cardiac arrhythmias3. Cardiovascular collapse
Other	<ol style="list-style-type: none">1. Skin necrosis2. Pyogenic granuloma3. Rhabdomyolysis4. Hypoglycemia5. Metabolic acidosis

Alcohol intoxication



“Serum Ethanol Levels in Children and Adults after Ethanol Embolization or Sclerotherapy for Vascular Anomalies” Mason KP et al., *Radiology* 2000;217, 127-132.

Alcohol serum levels have not been found to be related to morphology of the AVM, the venous drainage, or the injection technique but instead they correlate with the total amount of alcohol injected.

Communication between Providers: Improving Teamwork

Good communication between medical teams has been identified as being essential in avoiding complications in patient care. Recognizing a patient’s presentation and needs, sharing this information, being aware of perceptions, strength, and limitations of all members of the medical team can lead to a faster diagnosis of a problem, better workload distribution, more respect for each other’s contribution - and ultimately better patient care.

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