Anesthesia management in a 4 year old with merosin-deficient congenital muscular dystrophy undergoing a nephrectomy
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Introduction

We discuss a child with merosin-deficient congenital muscular dystrophy (MDCMD) who presented with abdominal pain and was found to have a ruptured renal mass and was scheduled for embolization of the right renal artery and nephrectomy. MDCMD is a rare heterogeneous subtype of CMD that can affect multiple body systems influencing the anesthetic plan and perioperative decision making.

Patient History

-4-year-old girl, 28.9kg, with MDCMD presents with a ruptured anaplastic Wilm’s tumor after a fall
-Born at 28 weeks GA by C-section 2/2 maternal pre-eclampsia
-Spent 2 weeks in the NICU for prematurity and feeding problems without intubation
-Diagnosed with MDCMD by muscle biopsy after multiple falls and failure to meet motor developmental milestones
-No family history of CMD
-Physical exam: Obese, interactive little girl. MP I airway with adequate mouth opening and normal neck range of motion. Normal CV exam. Poor muscle strength and tone in all 4 extremities but otherwise no focal neurologic deficits
-TTE showed EF of 44% and mildly depressed left ventricular wall motion

Perioperative Course

-Renal artery embolized in IR first then transport under GA to OR for nephrectomy
-Standard ASA monitors + arterial line and large bore PIV access
-MH precautions: induced and intubated successfully with propofol and rocuronium and maintained on propofol, remifentanil, and dexmedotomidine infusions
-ABGs Q1H, 1800ml of crystalloid for a metabolic acidosis and increased lactate with good effect
-EBL ~250ml
-Patient left intubated post-op and transported to the PICU.
-Monitored overnight, successfully extubated the following morning.

Intraoperative Record

Figure 2: Intraoperative record from compurecord

Color legend: Green - Heart rate, Red - Oxygen saturation, White – Blood Pressure, Blue - ETCO2

Nephrectomy specimen, anaplastic Wilm’s tumor
Cellular and extracellular proteins involved in pathogenesis of muscular dystrophy (cmd.org)

Discussion

-MDCMD is a heterogeneous subtype of CMD 2/2 mutations in the laminin alpha-2 chain of merosin, matrix protein linked w/ dystrophin
-Incidence is estimated to be 0.68-2.5 per 100,000
-Age at onset 6-12months old
-Merosin expressed in cardiac muscle, schwann cells, skin, skeletal muscle
-Still controversial if MH precautions needed
-No known treatment is available

Clinical Manifestations and Considerations of MDCMD

<table>
<thead>
<tr>
<th>Body System</th>
<th>Abnormalities</th>
<th>Anes. Concerns</th>
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<tbody>
<tr>
<td>Cardiovascular</td>
<td>Cardiomyopathy, LV dysfunction</td>
<td>Hemodynamic instability, prep CV eval (TTE)</td>
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<tr>
<td>Respiratory</td>
<td>Weak respiratory muscles, restrictive disease, jaw contractures</td>
<td>Higher risk for respiratory failure, aspiration, PFT’s, possible difficult airway</td>
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<tr>
<td>MSK</td>
<td>Generalized weakness, contractures, scoliosis</td>
<td>Sensitivity to depolarizing agents, ?MH risk, regional anesthetic difficulty</td>
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<tr>
<td>Neurologic</td>
<td>Hypotonia, MRI white matter changes</td>
<td>Normal/near normal intelligence, limited physical activity</td>
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References


Conclusion

MDCMD is a heterogeneous disease that effects multiple body systems. By understanding the characteristics of the disease one can better tailor the pre-operative evaluation and prepare for the intra-operative and post-operative management.