Title: 10 year old boy for dental surgery with fulminate Adrenoleukodystrophy: Considerations for Anesthesia management

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ABSTRACT BODY:

Case Report: A 10 year old, 28 kg boy reported for extensive dental restoration and multiple teeth extractions. His medical history is significant for an advanced case of Adrenoleukodystrophy. On pre-operative evaluation, the patient was wheelchair bound and had generalized spasticity. He was conscious of his surroundings and was non-verbal. His vitals were within normal limits. Airway exam also exhibited limited mouth opening. His medical regimen includes chronic steroid supplementation. Patient was induced with 8% sevoflurane with oxygen and nitrous oxide mixture and after loss of consciousness, an IV was started. Patient was administered vecuronium (1 mg/kg), fentanyl (1.5 mcg/kg) and 100 mg hydrocortisone. A naso-tracheal intubation was performed uneventfully. Total case duration was 4 hours and estimated estimated blood loss was 50 cc, and a total of 1 liter of Lactated Ringers containing 100 mg hydrocortisone was given. The trachea was extubated at the end of the case. The patient recovered uneventfully and was discharged home once the discharge criteria were met.

Case Discussion: Disease of white matter can be broadly distinguished as either defective formation of myelin (i.e. leukodystrophies) or demyelination (i.e. multiple sclerosis) (1). All of the leukodystrophies are characterized by progressive neurological deterioration. The primary defect lies in defective processing of Very Long Chain Fatty Acids (VLCFA) that accumulate in all tissues, but predominantly in the adrenal glands and central nervous system (4). The genetic marker was identified to be on the X chromosome, coding for a peroxisomal membrane protein called adrenoleukodystrophy protein (ALDP), responsible for transporting VLCFA’s into the peroxisome for degradation. The range of phenotypic expression is quite variable, and the distinction among subtypes is subtle. There are at least 6 different subtypes ranging from asymptomatic to severely advanced (i.e. Adrenomyeloneuropathy) progressive spastic paraparesis, polynucleopathy, adrenocortical insufficiency, psychosis and dementia. X linked Adrenoleukodystrophy or X-ALD is an X linked inherited disorder of peroxisomal transport protein for VLCFA’s. These VLCFA’s accumulate throughout the body, but most severely impact adrenal function and the central nervous system. The incidence is estimated to be closer to 1:17,000 (3). Due to the progressive nature of this disorder and its devastating effect on the central nervous system, these children frequently require anesthetic care during procedures such as MRI or during various surgical procedures.

Anesthesia considerations: Though several case reports exist, X-ALD is more common than previously thought and has important considerations for anesthetic management (4-7). Possible airway complications include the high incidence of gastroesophageal reflux, transient pulmonary aspiration secondary to poor pharyngeal muscle control and gastric dismotility. Pre-operative sedation should be carefully titrated. Rapid sequence intubation may be performed when appropriate. Incentive spirometry preoperatively may give a rough estimate of pulmonary dysfunction if needed. Behavioral manifestations of X-ALD and thrombocytopenia may prevent regional anesthesia. Anesthetics which lower seizure threshold should be avoided due to high incidence of seizure disorder. Succinylcholine should be avoided because of the possibility of hyperkalemia, especially in the bed bound patient (5). Generalized spasticity may necessitate the use of muscle relaxants. Hormone replacement and steroid therapy (HRST) is a key component to medical treatment of X-ALD. Stress dose supplementation must be given before the procedure to avoid adrenal crisis. Electrolyte imbalances from chronic HRST also require special attention. Finally, if the patient has undergone hemopoetic stem cell transplantation for treatment of X-ALD, they may be relatively immunosuppressed. Malnutrition impacts positioning of these patients, special padding may prevent pressure necrosis even during brief procedures (3).

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