KRABBE'S DISEASE: A CASE OF COMPLEX PAIN AND ANESTHETIC MANAGEMENT

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INTRODUCTION / AIM

Krabbe’s disease, also known as globoid cell leukodystrophy, is a rare progressive neurodegenerative disease that often presents early in infancy. Medical management of these children poses several challenges to the clinician due to medically refractory irritability, dystonia and pain. Children affected by Krabbe’s disease often require repeated diagnostic and invasive procedures under conscious sedation or general anesthesia. Due to the rarity of the disease and its rapidly progressive nature, there is a paucity of literature dealing with the anesthetic and pain management in children suffering from this condition.

METHODS

The multidisciplinary complex pain and anesthetic management of a child affected by Krabbe’s disease was reviewed. Health care provider notes and anesthetic records were examined.

RESULTS

The case of a 3-year-old male diagnosed with late infantile onset Krabbe’s disease was reviewed. Therapy was directed towards management of severe irritability, dystonia, spasticity and pain. Management of these symptoms was complex and effectively provided from a multidisciplinary team and included pharmacological and non-pharmacological interventions. Agents used in pain management in this case included morphine, indomethacin, acetaminophen and gabapentin. Irritability, dystonia, and spasticity were treated with a variety of agents including baclofen, benzodiazepines and clonidine. Physical medicine and rehabilitation management including the use of ankle foot arthoses, standing frames, splints, and stretching were also used to improve this patient’s overall condition. Anesthetic concerns highlighted in this case included treatment of acute on chronic pain, perioperative management of medications, and seizure control and chronic lung disease. Regional anesthesia was successfully utilized in this patient.

DISCUSSION / CONCLUSIONS

Management of pain and anesthesia in patients with Krabbe’s disease is complex. Multidisciplinary involvement is key in the care of these patients. There is overall a paucity of literature addressing specific strategies for management of pain and anesthesia in Krabbe’s disease.

OTHER AUTHORS

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