

Dexmedetomidine: A Saviour in a Child with Hand Schuller Christian Disease for MRI

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Abstract

Hand Schuller Christian disease (HSC) primarily affects infants and children. Its classical presentation includes triad of exophthalmos, diabetes insipidus, and calvarialytic lesions. Systemic involvement may include hepatomegaly, lymphadenopathy, dermatological, gastrointestinal tract, renal, pulmonary and CNS involvement. CNS involvement includes convulsions, increased intracranial pressure, focal neurological deficits, mental retardation, hearing disturbance, and tremors. We encountered a 4 year old female child with HSC disease who was posted for elective MRI scan under anaesthesia. Patient's history, physical examination, and lab reports revealed multitude of problems enumerated in case report. Such patients when posted for anaesthesia pose a major challenge in view of multiple organ involvement. Polydipsia made difficult to keep the patient fasting and giving general anaesthesia as per standard protocol became risky. The loose teeth with bleeding gums and protruded mandible would have compounded the mask ventilation, LMA placement or laryngoscopy and intubation, manifolds. We managed this case successfully with intravenous sedation with inj. dexmedetomidine at 1 µg/kg/min over 10 min and then maintenance infusion at 0.1 µg/kg/min. Spontaneous ventilation was supplemented with oxygen (FiO₂ 0.60). MRI scanning was completed in 45 minutes comfortably. Hence, dexmedetomidine offers advantage in such patients if anaesthesia has to be given in remote locations like MRI suite.

Keywords: Dexmedetomidine; Hand Schuller Christian disease; MRI

Introduction

Hand Schuller Christian disease (HSC) is one of the three components included in the histiocytosis X, the other two being eosinophilic granuloma and Letterer-Siwe disease. This disease is primarily seen in infants and children and rarely seen in adults. The classical triad of HSC disease-exophthalmos, diabetes insipidus, and calvarialytic lesions seen only in one third of patients [1]. CT scan is particularly helpful in detecting and defining the bony lesions and soft tissue involvement. The bones involved are skull, ribs, pelvis, scapula and mandible. Vertebrae and the appendages are less frequently involved. Optic atrophy, otitis media, and extrusion of teeth may occur as a result of adjacent bony involvement [2]. Systemic involvement of this disease includes hepatomegaly, lymphadenopathy, and dermatological, gastrointestinal tract, renal and pulmonary involvement. CNS involvement includes convulsions, increased intracranial pressure, focal neurological deficits, mental retardation, hearing disturbance, and tremors. Involvement of the pituitary and hypothalamus results in delay in sexual maturity and bone development, which is often seen in the pediatric age group [3]. Perioperative risks in such patient depend upon the organ system involved and the extent of dysfunction. We report a case wherein a child suffering from HSC disease required anaesthesia for getting MRI done.

Case Report

A 4-year-old female child, weighing 15 kg, diagnosed as a case of HSC disease was posted for elective MRI scan under anaesthesia.

History of the patient revealed chief complaints of bleeding gums since 2-3 months and polydipsia since 15 days. Child was undergoing regular follow up by endocrinologist at our institute who advised an MRI scan. On examination, her lower jaw was protruded (Figure 1).



Figure 1: Female child with protruded lower jaw.

Oral examination revealed inflamed gums with foetid breath, irregular dentition; with almost all the teeth loose (Figures 2 and 3).



Figure 2: Showing inflamed gum with loose, irregular dentition.



Figure 3: Showing inflamed gum with loose, irregular dentition.

Her investigations included hemoglobin 5.2% g, with MCV 52.80, MCH 12.60, MCHC 23.80. Serum chloride level was increased. Urine osmolality was 17.23. Hepatic and renal functions and serum electrolytes were within normal limits. Urine specific gravity, serum ADH levels, serum calcium and phosphates were awaited. USG abdomen and X-ray skull were normal. X ray STN showed lytic lesion in the mandible. Bone marrow examination showed normoblastic erythropoiesis with non-specific myeloid reaction. Past and family history was unremarkable. Patient had been advised 6 hours for

fasting. On the day of MRI under anaesthesia, child had water an hour before as she could not resist due to polydipsia. No premedication was given. Thorough clinical examination was done. All standard monitor like ECG, SpO₂ were attached. Intravenous access was established with 24 G cannula. Intravenous sedation was started with inj. dexmedetomidine at 1 µg/kg/min over 10 min and then maintenance continued with slow infusion at 0.1 µg/kg/min. Spontaneous ventilation was supplemented with oxygen (FiO₂ 0.60) via venturi mask. MRI scanning was allowed to commence. The procedure was completed in 45 minutes comfortably. Once the scanning was done, infusion was stopped and the patient responded in the recovery room after 15-20 minutes.

Discussion

Langerhans cell histiocytosis (LCH), is a rare disease characterized by proliferation of Langerhans cells or their precursors [4,5]. It is estimated that the disease occurs at the rate of 0.2 to 0.5 cases per 100,000 children per year. The skull, mandible, ribs, vertebrae, and long bones are involved. Head and neck lesions are common and 10% of all patients have oral lesions [6,7]. The etiology and pathogenesis of LCH remain obscure.

The disease has been classified by Lichtenstein depending on the patient's age at onset and distribution of lesions as follows: eosinophilic granuloma (chronic focal LCH), which is refer to solitary or multiple bone lesions with no extra skeletal involvement; chronic disseminated LCH (Hand Schuller Christian disease), which is a specific clinical traid of lytic bone lesions, exophthalmos, and diabetes insipidus; Letterer Siwe disease (acute disseminated LCH), which is a malignant form of LCH [8,9].

The oral changes are often the first clinical signs in all the form of LCH [10]. The diagnosis is confirmed by the histopathological examination supported by clinical and radiological examination. Although there is no specific laboratory test for LCH, the blood and urine tests reveal the extent and seriousness of the disease [8].

These patients when posted for anaesthesia pose a major challenge due to following concerns:

- Specific organ dysfunction caused by infiltration of histiocytes as in liver, lungs, hematopoietic system, pituitary, spleen, and bone.
- Treatment with steroids and chemotherapy, which add the risk for adrenal insufficiency and may require stress steroids.
- Diabetes insipidus due to posterior pituitary involvement.
- Cervical instability, if lesion present in cervical vertebrae.
- Severe pulmonary dysfunction possible, pulmonary hypertension without right heart failure.

In our patient, polydipsia made it difficult to keep the patient fasting and hence giving general anaesthesia as per standard protocol would have been risky. The loose teeth with bleeding gums and protruded mandible would have compounded the mask ventilation, LMA placement or laryngoscopy and intubation, manifolds.

Sedation is often required in young children in order to obtain quality diagnostic imaging like MRI. It can be challenging to obtain the deep sedation level required to prevent the patient's movement while maintaining respiratory and hemodynamic stability. In addition, limited access to the patient may pose a safety risk during MRI [11]. Therefore, it is very important to select the appropriate drugs and dosage that allow optimal imaging, while maintaining hemodynamic

and respiratory stability. Dexmedetomidine, a selective alpha-2 adrenoceptor agonist, has a very safe therapeutic window with respect to respiratory depression. This quality offers a distinct advantage in procedural sedations, where the patient is not immediately accessible to the medical team. There are various studies demonstrating that dexmedetomidine is a good option for procedural sedation [12,13]. Dexmedetomidine is a well-tolerated and effective drug for successful quality imaging in the majority of pediatric patients.

In our scenario, even though we were fully prepared for an anticipated difficult airway, use of dexmedetomidine helped us avoid all the above problems and proved to be an appropriate choice in preventing given situations.

Conclusion

Children with HSC disease poses a great challenge for general anaesthesia and should be managed with careful history taking, thorough general examination, and well planned anaesthesia strategy with full preparation for anticipated problems like difficult airway under adequate monitoring. Dexmedetomidine offers advantage in such patients if anaesthesia has to be given in remote locations like MRI suite.

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