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ANAESTHETIC MANAGEMENT OF A PATIENT WITH OSTEOPETROSIS POSTED FOR DYNAMIC HIP SCREW FIXATION FOR FRACTURE FEMUR

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ABSTRACT Osteopetrosis is a rare genetic disorder of osteoclast dysfunction leading to anatomical and physiological disorders. We present the anaesthesia management for neck of femur fracture of a 19-year-old girl with malignant infantile type of osteopetrosis now presented with fracture femur posted for dynamic hip screw fixation. She had visual disturbance, growth failure, facial deformity, transfusion dependant anemia, and with splenomegaly

KEYWORDS : Airway management, bone fracture, osteopetrosis

INTRODUCTION

The term osteopetrosis is derived from the Greek word "osteo" meaning bone and "petros" meaning stone.[1] Osteopetrosis is also known as "marble bone disease," "osteosclerosis,"[2] and "Albers-Schonberg disease."[3]

The anesthesia literature contains little discussion and few recommendations for the management of these patients. The main problem in this disease is inability in effective bone resorption and regeneration, leading to anatomical and physiological disorders.[4,5] We present a case report of an osteopetrosis child who needed a general anaesthesia management for a femur fracture which illustrated the possibility of handling this type of patient's airway less invasively.

Case Report

A 19-year-old girl with 32kg weight and 130 cm height admitted for femur fracture. The child developed inability to read letters on the board at 13 yrs of age which was diagnosed as optic neuritis and subsequent evaluation done and osteopetrosis was diagnosed. Now she is admitted for fracture femur fixation. Physical examination revealed short stature, growth delay, facial deformity, dolicocephaly optimal mouth opening and neck extension [Figure 1] and splenomegaly. Heart rate and respiratory rate of the patient were 106/min and 20/min, respectively, with a 130/80 mmHg blood pressure. Laboratory results of the patients at the baseline were as follows – hemoglobin: 7.8 g/dL (anisocytosis, hypochromia, ovalocytosis, with tear drop cells ,target cells) and platelet: 46000/mm^3, which reached to 10.2 g/dL and 70,000/mm^3 after receiving two units of blood and 4 units platelet, respectively. White blood cell: 10.4 × 10^3/μL, red blood cell: 3.9 × 10^6/μL, alkaline phosphatase: 103 U/L, lactate dehydrogenase: 832 U/L, phosphorus 4 mg/dL, Na: 134 mEq/L, K: 4.6 mEq/L, and calcium 9 mg/dL, Magnesium :1.8 mEq/L, ECG – NSR, HR-108/min, No ST-T changes, CXR- WNL

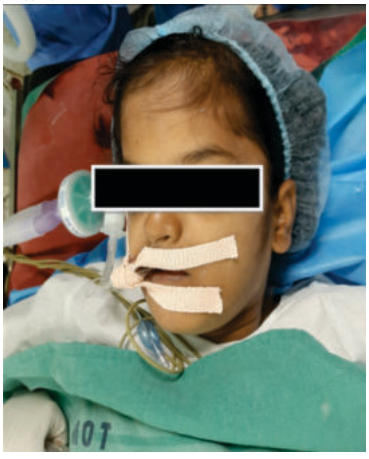

Figure 1

Because of the femur fracture, the patient was candidate for applying Dynamic Hip Screw under general anaesthesia.

Due to the probability of difficult airway management (regarding facial abnormality), necessary precautions were made in the setting of anaesthesia before induction .

PLAN- General Anaesthesia with Controlled Ventilation with Endotracheal tube intubation

The equipment for difficult airway management was included into the preparation setting which were laryngoscopes with Mackintosh and Miller types blades (sizes 1-3), videolaryngoscope, ventilation face masks sizes 1-3, stylets with endotracheal tubes sizes 5.5-6.5 mm ID (un-cuffed and cuffed) and supraglottic airway devices.

Before anaesthesia induction, the patient was monitored with electrocardiography, noninvasive blood pressure, oxygen saturation (SpO_2), and end-tidal carbon dioxide ($ETCO_2$). Balanced isotonic solution was administered perioperatively during the anesthesia management.

Midazolam 0.15 mg/kg and fentanyl 1 μg/kg with 0.1 mg/kg of Glycopyrrolate were administered as premedication. General anaesthesia was induced with propofol 2 mg/kg. The patient was intubated with videolaryngoscope with 6 mm COETT and maintained with O_2 +N_20 + Isoflurane. During the anesthesia, the heart rate was 110–120/min, blood pressure 100/70 mmHg, SPO_2 = 99%, and $ETCO_2$ 28–30 mmHg. Intraoperatively transfused 2 units SDP and the course of anaesthesia and recovery was uneventful.

DISCUSSION

Osteopetrosis has three clinical types: The malignant infantile type of osteopetrosis has poor prognosis and is inherited as an autosomal recessive genetic trait and the intermediate type and benign adult type of osteopetrosis have similar but milder clinical manifestations [Table 1]

Table 1 : Classification and anesthesia implications for autosomal dominant and autosomal recessive (intermediate type) osteopetrosis disease

Genomic sub-types	Osteoclastic changes	Phenotype severity	Age	General implications for accompanying pathologies	Important accompanying pathologies	Accompanying pathologies concerning anesthesia	Related anesthesia managements
Autosomal dominant							
Type I autosomal dominant osteopetrosis	Reduced number and size of osteoclasts	Mild severity	Adulthood	Remarkable coexisting pathologic conditions that may need a different expertise approach	Occasionally, diagnosis may be based on dental radiographs that show a diffuse increased radiopacity of medullary bone	Cranial nerve compression is common but fractures are rare[8]	Anesthesia implications for related cranial nerve pathologies
Type II autosomal dominant osteopetrosis	Large, highly multinucleated osteoclasts	Mild severity	Adulthood	Remarkable coexisting pathologic conditions that may need a different expertise approach		Nerve compression is uncommon and fractures frequent[8]	Anesthesia implications to prevent fractures
Benign autosomal recessive		Intermediate severity	Childhood	Some signs and symptoms of malignant osteopetrosis Fractures by the end of their first decade of life	Bony sclerosis renal tubular acidosis cerebral calcifications macrocephaly, mild or moderately severe anemia and ankylosed teeth that may predispose them to osteomyelitis of the jaws	Preoperative evaluation and treatment of anemia[8]	Oral and airway manipulation with precautions of dental problems General perioperative anesthesia implications[8]

Classic malignant autosomal type which occurs in infants and younger children is accompanied by multiple significant pathologies which are common and maybe sever. Bone resorption prevents the enlargement of bone pores, resulting in bone marrow damage that is associated with hematological disorders. Bone foramina put pressure on cranial nerves and lead to visual and hearing impairments and cranial nerve palsies. Among the other signs of this disease are failure to thrive, anemia, thrombocytopenia, hypokalemia, hepatosplenomegaly, esotropia, amblyopia, extramedullary hematopoiesis, and pathological fractures. A number of patients present with neurological symptoms such as seizures, hypotonia, retinal atrophy with absent evoked visual potentials, neural sensory-based deafness, significantly delayed myelination, diffuse progressive cortical and subcortical atrophy.

Four prior reports related to structural brain malformations in autosomal recessive osteopetrosis (ARO) patients have been published.

Reports of ARO variants (neuropathic ARO) with structural brain anomalies included macrocephaly, hypertelorism, agenesis of the corpus callosum, hydrocephalus, and Dandy–Walker malformation. Classical ARO is caused by TCIRG1 mutations.

Pathologic bone remodeling, which causes greater mineral density and compressive strength, lacks the needed elasticity and as a result pathologic bone fracture develops which is the hall mark of osteopetrosis.

Hematopoietic stem cell transplantation may reduce the difficult airway-related problems in subsequent anaesthesia procedures, but this has not been confirmed yet.

Patients with severe pancytopenia may develop heart failure or sepsis.A forward flow theory due to splenomegaly is considered to explain the etiology of portal hypertension in osteopetrosis patients Although controversial captopril has been used for the treatment of portal hypertension in patients with low portal variceal velocity to prevent complications of variceal bleeding.Osteopetrotic patients may develop hypertension and left ventricular heart failure.

Difficult airway management and failed intubation have been reported in 17.7% and 14.5% of cases, respectively.[6] Moreover, the chance of intubation failure has been reported to be higher in osteopetrosis children than other children.[3] In a report, the incidence of respiratory-related problems was 11% which included critical airway management and hypoxemia.[9] However, the rate of respiratory problems was reported to be only 2% of children younger than 8 years among pediatric surgical patients.

Factors that are related to difficult airway management in osteopetrosis patients can be due to mandibular abnormalities with hypoplasia. The space between the tongue and the posterior pharyngeal wall can also be narrow. A high arched, narrow hard palate, an obliterated nasal passage due to congestion, wide lower hyperplasic jaw, restricted temporomandibular joint movement, and cervical spine fractures may cause difficult endotracheal intubation.Hepatosplenomegaly-induced abdominal distention may also lead to ventilation difficulty.

Equipment and methods (e.g., awake intubation) required for a difficult intubation and possibility of tracheal aspiration must be taken into account.[2] Our particular patient did not pose any difficulty or adverse event during the anesthesia. short duration of the procedure, and no electrolyte abnormalities permitted a smooth course of anesthesia with no impending need for a more advanced airway management and the patient's oxygenation and end tidal carbon dioxide levels were in normal range.

CONCLUSION

Despite the possibility of critical airway management in osteopetrosis patients can be handled easily with proper planning and management.Multisystem evaluation and proper perioperative optimisation can lead to smooth conduction of anaesthesia in osteopetrosis.

Declaration Of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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