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Research Article

ANAESTHETIC MANAGEMENT IN A PATIENT WITH OSTEOGENESIS IMPERFECTA UNDERGOING CRANIOTOMY FOR SUBDURAL HAEMORRHAGE

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A B S T R A C T

Osteogenesis imperfecta (OI) is a genetic disorder of connective tissue in which bones fracture very easily, often for no apparent reason. The aetiology of the disease is a gene defect that produces very little or poor quality type 1 collagen, an important building block of bones.

Anaesthetic management is challenging because of the multi-organ system involvement in OI. Most notably, anaesthesiologists are challenged by a difficult airway, the fragile skeletal system during positioning, cardiovascular involvement, coagulation abnormalities and an association with malignant hyperthermia. We report successful anaesthetic management of an OI patient with a subdural haemorrhage associated with a road traffic accident who underwent craniotomy.

INTRODUCTION

Osteogenesis imperfect a (OI) is a fibro-osseous disorder of the collagen tissue. Also known as brittle bone disease, it is a genetic disorder of connective tissue in which bones fracture very easily, often for no apparent reason. The aetiology of the disease is a gene defect that produces very little or poor quality type 1 collagen, an important building block of bones.[1] Usually, patients inherit the disease from a parent, but sometimes cases are sporadic and the result of new genetic mutation.[2] Other clinical features of the disease include blue sclerae, progressive deafness, brittle teeth (dentinogenesis imperfecta) and hypermobile joints. In the severe form of the disease, kyphoscoliosis may lead to significant chest wall deformity and restrictive lung disease. These two pathological conditions, together with recurrent pneumonia, can progress to cardiac failure.[3] The central nervous system may be involved, with spinal cord or brainstem pressure effects, like basilar invagination or impression and craniovertebral junction problems.[4] The incidence is 6-7 per 100 000 people.[5] It leads to defects in skeletal growth and these patients present with short stature.

Anaesthetic management is challenging because of the multi-system organ involvement in OI. Most notably, anaesthesiologists are challenged by a difficult airway, the fragile skeletal system during positioning, cardiovascular involvement, coagulation abnormalities and an association with malignant hyperthermia.[2] The diversity of presentation means that patients with severe forms may present with multiple fractures with minimal or no trauma, and persons with mild forms may only manifest with premature osteoporosis or severe postmenopausal bone mineral loss.[6] There is no cure for the disease, and currently medical management remains symptomatic. Intramedullary nailing of long bones remains the method of choice for correcting deformities and preventing fractures.[7] We report successful anaesthetic management of an OI patient with subdural haemorrhage associated with a road traffic accident who underwent craniotomy.

CASE REPORT

A 24-year-old male, weighing 70 kg, with a height of160 cm, and a history of repeated fractures since the age of 8 years, presented to our centre after sustaining a subdural haemorrh age associated with a road traffic accident. He was a known case of OI tarda. The patient's history revealed an uneventful birth history and no delay in milestones. There was no past surgical history and family history of the disease. Patient's GCS was E1V2M5 during preoperative assessment. A general physical examination revealed bowing and valgus deformity in upper and lower limbs. There was blue sclera present in both eyes. There was no pallor, icterus, cyanosis, clubbing, lymphadenopathy or pedal oedema. Her blood pressure was 140/80mmHg and her heart rate 60 beats/min. The O2 saturation was 95% on room air. Except for the musculoskeletal deformities, all other systemic examinations of the patient were normal including the cardiac status.

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The preoperative laboratory investigations were normal (Table I). The preoperative electrocardiography, chest X-ray and coagulation profile were also normal. Airway assessment could not be done because of the low GCS of our patient. The patient was accepted for surgery as an American Society of Anesthesiologists (ASA) grade IV E patient. In view of the anticipated difficult airway, emergency surgery and presence of a threatened airway as the GCS of the patient was very low, it was decided to intubate the patient and to proceed with a general anaesthetic technique. Before anaesthesia was conducted, a thorough preoperative examination and operating room preparation were completed, including the procurement of difficult airway equipment (a stylet, different sizes of face mask, different sizes of endotracheal tubes and laryngoscope blades, laryngeal mask airways (LMA), Proseal LMA, and a tracheostomy set). The patient was positioned very carefully on the operation table to prevent a new fracture from occurring. Two 16-Gintravenous cannula were inserted, one each on the dorsum of both the hands. Standard ASA monitors were placed on the patient including electrocardiography, noninvasive blood pressuremonitor, EtCO2, Temprature and pulse-oximeter. Baseline BP was 140/80 mmHg, HR 60 beats/minute, respiratory rate 24 breaths/minute and the SpO2 on room air, 95%. An intravenous normal saline was administered. Patient was premedicated with midazolam 1mg and ondansetron 4mg IV and preoxygenated with 100% oxygen. Induction was done with thiopentone 275mg IV and after checking for ease of bag mask ventilation, vecuronium 7mg IV was given. After 3min, laryngoscopy was done which revealed Cormack Lehane grade II visualization and trachea was intubated with 8.5 mm ID endotracheal tube in the first attempt. Maintenance was done with O2, N2O, isoflurane and intermittent doses of vecuronium.

Positioning was done cautiously and all the pressure points were well padded. The surgery lasted for three hours and 40 minutes and there was profuse bleeding during the surgery. Estimated blood loss was approximately 2 – 3 litres, which was adequately replaced by fluid, packed red blood cells, fresh frozen plasma and platelets. Vasopressor infusion IV had to be started during surgery to provide haemodynamic stability, which was tapered off successfully by the end of surgery. At the end of surgery, patient was shifted to ICU with ETT in-situ and kept on ventilator support postoperatively. Patient was weaned off from ventilator and extubated on day 4 of surgery and was shifted to ward on day 6 after some improvement in general condition. Further management was continued there.

Table I Laboratory Investigations of the patient with osteogenesis imperfect

Investigations	value
Haemoglobin	12.3 gm/dl
Total leukocyte count	10, 000/mm^3
Prothrombin time	13 second
INR	1.00
Blood sugar	80 mg/dl
Blood urea	35 mg/dl
Blood creatinine	1.00 mg/dl
Serum sodium	139 mEq/l
Serum pottassium	4.89 mEq/l
Platelet count	1.3 lac

DISCUSSION

OI is a rare autosomal dominant inherited disease of connective tissue that affects bone, sclera and the innerear.[8] The underlying cause is mutation in the gene coding for type 1 procollagen, i.e. collagen, type I, alpha 1 (COL1A1)and COL1A2. It affects 6-7 of 100 000 people, and occurs approximately in 1:20 000 births.[5] Males and females are affected equally. No racial difference has been noted.[5] Initially, OI was divided into two forms: OI congenita and OI tarda. Fractures occur in utero, and death usually occursin the perinatal period in the congenita form. Typically, the tarda form presents during childhood or early adolescence, and patients usually have a normal lifespan.[4] The most commonly used classification of OI by Sillence *et al*.[9] categorises it into four clinical types: I, II, III and IV; based on its phenotypic manifestations and the radiographic appearances of the bones. The latest classification divides OI into nine types.[6] The presentation of clinical severity depends upon the effect of mutation. Type I is considered to be the mildest form of the disease, and is compatible with long-term survival in adulthood. Types III-IX are moderate to severe forms of OI, depending on the defect in the structural proteins. Type II is lethal in the perinatal period and is not compatible with life.[6] Anaesthetic management is influenced by coexisting orthopaedic deformities, vulnerability of fracture during simple positioning, platelet dysfunction, difficult intubation, cardiovascular abnormalities like mitral valve prolapse, a tendency to develop malignant hyperthermia, and rarely, extra skeletal manifestations.[10,11] Because of abnormal skeletal growth, odontoaxial dislocation and hypermobilejoints, a difficult airway must always be anticipated in these patients.[12] Associated kyphoscoliosis with pectus carinatummay decrease vital capacity and chest wall compliance, with resulting arterial hypoxaemia due to ventilation perfusion mismatch.[2] This can pose an increased risk under general anaesthesia. Succinylcholine should be avoidedbecause of its potential to cause malignant hyperthermia. Fasciculations can also lead to fractures.[13] Use of halothane should be avoided as it may lead to malignant hyperthermia.[10,11] Bergstorm *et al*, and Rampton *et al* reported several cases of malignant hyperthermia in OI patients.[14,15] The general consensus is that many cases of hyperthermia in OI are not of the malignant type, but instead are the result of a hypermetabolic state. It has been suggested that hyperthermia is the result of either abnormal central nervous system temperature regulation or abnormal cellular energy metabolism.[13,16] At least 50% of patients with OI also have an elevated serum thyroxine level, leading to increased O2 consumption and heat production.[5] Avoidance of common malignant hyperthermia-triggering drugs, temperature monitoring and the provision of necessary drugs and cooling devices should be easily available.[15] The fragility of bones in these patients is well known. Malde *et al* reported a fracture shaft of the femur in a patient with OI which occurred during the transfer of the patient to the recovery room.[12] The best anaesthetic technique in these patients is regional anaesthesia as it avoids the necessity of tracheal intubation. The development of perioperative hyperthermia is also less likely to occur with regional anaesthesia than with general anaesthesia, and the former facilitates detection of a thyroid storm.[17] However, before administration of a regional anaesthetic, a coagulation profile must be undertaken because of the associated increase in bleeding time, despite the normal platelet count.[2] Chances of bone injury during positioning should be kept in mind. Automated blood pressure cuffs may be hazardous as over inflation may result in a fracture.[17] Pressure points should be

padded during prolonged surgery, and transportation must be gentle to prevent fracture occurrence.[12] There have been several successful case reports of surgery being conducted under general, as well as regional anaesthesia on patients with OI.[1,2,12] Karabiyik *et al*. recommended total intravenous anaesthesiatogether with an intubating LMA to manage cases electively, while Malde *et al*. successfully used balanced general anaesthesia in a case of OI with gross deformity of the pelvis for abdominal hysterectomy. As the airway was threatened due to low GCS and we anticipated difficult airway, we intubated the patient beforehand and got the surgery conducted under GA and it was uneventful.

CONCLUSION

Patients with OI pose a significant challenge to the anaesthesiologist owing to a difficult airway, problems with positioning, susceptibility to fractures, a tendency to develop perioperative hyperthermia, platelet functional abnormalities and difficult spinal anaesthesia. A detailed preoperative work-up including a careful history and examination, as well as prompt and adequate anaesthetic management, taking care of the various manifestations of the disease, can improve the outcome in these patients.

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