Anaesthetic Management in a Patient of Osteogenesis Imperfecta with Basilar Invagination

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Citation

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Abstract

Osteogenesis imperfecta (OI) is a rare, autosomal inherited disorder due to a defect in collagen I synthesis leading to skeletal deformities with a characteristic tendency to fracture bones easily, and ocular, otologic, cutaneous and dental abnormalities1. Basilar invagination (BI) is one of its rare complications, occurring as the weight of the cranium progressively deforms the unusually soft skull base, producing invagination of the foramen magnum and translocation of the upper cervical vertebrae into the posterior cranial fossa2. This causes brainstem compression leading to severe neurological disability, respiratory compromise and even sudden death; anaesthetizing such a patient thus is quite a challenge. We report here a similar patient who underwent transoral odontoidectomy for relief of neuraxis compression.

CASE REPORT

A 55 year old woman, diagnosed since birth as OI, came with increasing breathing difficulty of two weeks' duration. She also had progressive difficulty in swallowing, nasal regurgitation of food, hoarseness and decreased volume of voice, reduced vision, squint in the right eye, headaches associated with vertigo, decreased hearing in the right ear and inability to walk. She was a short (1.25 m) and frail woman weighing 42 kg, with blue sclerae and multiple bony abnormalities including kyphoscoliosis and pectus excavatum. Her breathing was laboured and chest auscultation revealed bilateral basal crepts. She also had multiple cranial nerve palsies, grade III quadriparesis, and 50% sensory loss in her right foot. Remaining general physical and systemic examination including cardiovascular, were unremarkable. Airway assessment revealed an extremely short neck with little movement and an adequate mouth opening but a disproportionately large tongue.

Routine blood investigations, coagulation profile and an electrocardiogram were within normal limits. X-ray chest had bilateral patchy opacities and pulmonary function tests showed restrictive lung disease. CT scan and an MRI of the skull showed gross BI with platybasia and compression of the brain stem and lower cranial nerves by the dens.

Transoral odontoidectomy was planned. Preoperative cervical traction and lung physiotherapy greatly improved the patient's headaches, motor power and her respiration; she was taken up for surgery one week later.

Anaesthesia was initiated with IV morphine 3mg, IV midazolam 2mg, IV propofol 2 mg/kg, O2 and N2O (FiO2 0.5). Immediately thereafter the patient had upper airway obstruction with her SpO2 dropping to 60%. Insertion of an intubating laryngeal mask airway (ILMA) was attemped twice without success. Intubation was achieved with direct laryngoscopy under spontaneous respiration. Cormacke and Lehane score was Grade II. Anaesthesia was maintained with atracurium infusion 0.5 mg/kg/hr, propofol infusion 6 mg/kg/hr, O2 and N2O (FIO2 0.5) on controlled ventilation. Haemodynamics and arterial blood gases (ABG) were maintained within normal limits. Except for noticeably more surgical ooze requiring 2 units of whole blood., the operation was uneventful. Reversal was with IV 2.5 mg neostigmine and 0.4mg glycopyrrolate following which the patient was conscious but had inadequate respiratory efforts and airway reflexes, and significant oropharyngeal oedema. She was ventilated overnight and uneventfully extubated the next morning. She remained alert with satisfactory haemodynamics and ABG throughout the day.

Twelve hours later, she had a sudden excruciating midsternal pain radiating to the back and looked extremely distressed, followed soon by a cardiorespiratory arrest. Resuscitation was initiated immediately but she could not be revived despite continued efforts for over an hour. Consent for autopsy was refused.

DISCUSSION

Airway management in an OI patient coming for surgery is a major challenge for the anaesthetist. This is attributed to a short neck, large tongue, prominent occiput, fragile mandible & cervical spine and predisposition to mucosal bruising and broken teeth. Associated BI, with an upward translocation of the cervical spine, distorts the airway anatomy further. Besides, our patient also had an immobile cervical spine due to traction. Fibreoptic intubation, an ideal technique in such situations, was not available to us. ILMA has also been recommended for these patients as it facilitates intubation with minimal neck movement.

A greater predisposition to pulmonary disease in patients of OI with BI because of kyphoscoliosis and thoracic cage deformity, and recurrent aspirations requires aggressive preoperative optimisation of lung function. There may also be hypoxemia secondary to ventilation – perfusion mismatch. Delay in extubation is anticipated in these patients for the same reasons, with oropharyngeal oedema following transoral odontoidectomy being another factor. Proper positioning and adequate padding of all pressure points during surgery and transfer is required because of the extreme susceptibility to fractures in these patients. Upto 30% incidence of bleeding diathesis in patients of OI has been reported[3]. Increased capillary fragility, decreased levels of factor VIII and deficient collagen induced platelet–aggregation have been implicated as causes. Increased intraoperative bleeding may occur despite normal bleeding times and coagulation values. Intraoperative hyperthermia and malignant hyperpyrexia have been reported in patients with OI undergoing general anaesthesia[4]; avoidance of hyperthermia triggering agents and availability of rapid cooling methods are important.

Anticipating these problems helped us achieve a relatively uneventful intraoperative course in our patient. However, we were unprepared for the sudden and dramatic terminal events in her that we attributed to either an acute, extensive MI or a massive pulmonary embolism, neither of which could be confirmed since all our efforts were directed towards resuscitating the patient first. A third possibility of acute aortic dissection, perhaps more relevant because of the characteristic nature and location of pain, did not occur to us then. Later literature search showed 2 reports of patients of OI without a known cardiac disease presenting with acute aortic dissection[5,6]. In the absence of confirmatory tests or autopsy, our diagnosis remains speculative but we now feel that a preoperative echocardiogram and venous Doppler of the lower limbs (in a bedridden patient like ours) could have helped. We recommend a thorough multisystem work-up of OI patients to help anticipate any adverse outcome and start timely corrective measures.

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