Anaesthesia management of a known case of Progeria for Functional Endoscopic Sinus Surgery

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Citation

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Abstract

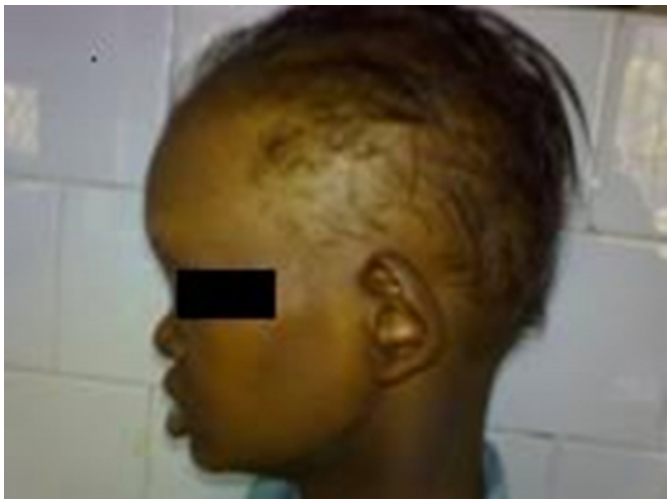
Progeria or premature ageing is a very rare congenital abnormality in children. It is a challenge for an anesthesiologist due to the difficult airway and ischemic heart disease. Here we report the successful anaesthesia management using general anaesthesia of a known case of progeria. The patient underwent Functional Endoscopic Sinus Surgery (FESS). Intubation was uneventful and perioperative vitals remained stable. Postoperative observation was uneventful.

INTRODUCTION

The incidence of progeria is reported to be 1 in 4 million to 1 in 8 million[1]. Patients have characteristic features like scanty hair and teeth, wrinkled skin, early atherosclerosis of blood vessels and joint deformities. Prognosis is poor with survival beyond second decade very rare[2]. The management of a case of progeria is always a challenge to the anesthesiologist as it is complicated by difficult airway. Several cases of repeatedly failed intubations even with fiber optic bronchoscope have been reported [6,7,8]. These patients have ischemic heart disease, which is reported to be the most common cause of death in these patients[2]. We report a case of successful airway and intraoperative anaesthetic management of a case of progeria using general anaesthesia.

Figure 1

Figure 1: Facial features of the patient of progeria



CASE REPORT

A thirteen year old male child presented with swelling over the right eye, diagnosed as mucocele of frontal sinus and was posted for FESS. Progeria was diagnosed during the preoperative work-up. He was a full term normal delivery, with normal attainment of milestones, adequate immunization till date and no history suggestive of any mental retardation. There was a history of delayed eruption of teeth, till date only 9 teeth had erupted. On examination the child had scanty hair on the head, wrinkled skin, flattened nasal bridge and protruding ears with thin limbs. He was well oriented and cooperative. All vital parameters, respiratory and cardiovascular examination were normal.

Airway assessment showed adequate mouth opening, MPC-I, widely spaced teeth (total-9) and micrognathia. However, neck movements, thyromental and sternomental distances were normal. IDL showed bilateral mobile vocal cords. All investigations were normal except for ECG changes showing T wave inversion in V1-V4 and Q wave in II and III. 2-D echo showed ejection fraction of 65% and mild MS with MR and no other abnormality. Stress test was negative for any inducible ischemia.

After securing i.v. line, monitoring was done with cardio scope, pulse-oxymeter and NIBP. Appropriate size endotracheal tubes, laryngeal mask airways and fiber optic bronchoscope were kept ready. Anaesthesia was induced with Inj. Propofol 2mg/kg and Vecuronium 0.1m/kg after confirming adequate mask ventilation. The patient was intubated with a 6.5 no. portex cuffed endotracheal tube and

anesthesia was maintained on N2O:O2::50:50, Isoflurane 0.4-0.6% and Vecuronium 0.1mg/kg/hr. Surgery was uneventful. Anaesthesia was reversed with Inj. Neostigmine 0.05mg/kg and Glycopyrrolate 8mcg/kg and the patient was extubated. The patient was observed for 24 hrs postoperatively and discharged from anaesthesia care.

DISCUSSION

Progeria refers to Hutchinson-Gilford Progeria syndrome. The disease affects between 1 in 4 million (actual) and 1 in 8 million (reported) newborns.[1] A 2006 report in Nature said progeria may be a de novo dominant trait[1]. It develops[1,2] during cell division in a newly conceived child or in the gametes of one of the parents. It is caused by mutations in a LMNA (Lamin A protein) gene on chromosome 1. Researchers now believe that the defective Lamin A (which is the structural scaffolding that holds the nucleus of a cell together) protein makes the nucleus unstable. That cellular instability appears to lead to the process of premature aging in Progeria. The condition is distinguished[1,2,3,4,5] by limited growth, alopecia and a characteristic appearance with large head for size of face (macrocephaly), small jaw (micrognathia) and pinched nose. Delayed and abnormal tooth eruption and morphology are commonly present. Later the condition causes dry, scaly and wrinkled skin. There may be early atherosclerosis of blood vessels leading to abnormal stress test. Patients usually have thin limbs with prominent joints, short stature, joint stiffness, hip dislocations. Insulin-resistant diabetes has been reported. Mental development is not affected. Stroke and myocardial infarction are two major causes of death. There is no known cure. Survival beyond second decade is very rare.[2]

Anaesthetic problems encountered in a case of progeria include difficult airway and myocardial infarction. Review of literature[6,7,8,9] shows repeated failed attempts of intubation even with fiber optic bronchoscope. Some anaesthesiologists have used laryngeal mask airways as life saving devices in such cases before intubating with the help of fiber optic bronchoscopes. A case of propofol infusion syndrome with lipaemia has been reported in a child with progeria[10]. Myocardial infarction has been reported as one of the most important causes of death in progeria[2]. However intraoperative myocardial infarction in case of progeria has not been reported till date.

Keeping all these cases in mind we kept all equipments of difficult intubation ready. IDL was done to rule out any airway abnormality. We induced with propofol and after checking adequate mask ventilation vecuronium was given. Scoline was avoided regarding concern of any occult myopathy and also ventilation not being a problem. Sevoflurane induction and intubation while the child being still on spontaneous breathing might be a safe alternative for induction of anaesthesia. FESS being a nasal procedure having risk of post nasal bleed, we did endotracheal intubation with throat pack and avoided laryngeal mask airway. Preoperative stress test and 2-D echo were done to rule out any evidence of myocardial ischemia. However, in our patient it showed mild MS with MR. Intraoperative ECG didn't show any signs of ischemia. Blood pressure, pulse rate also remained stable intraoperatively. After 24 hrs of uneventful post operative observation child was discharged from anaesthesia care unit.

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