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Anaesthesia in Hallerman-Steriff syndrome: A rare case report

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Abstract: *Anaesthetic management of 21 year old mentally retarded female diagnosed case of Hallerman-Steriff syndrome presented in this case. Anaesthesia was challenging because of difficult airway (mandibular hypoplasia and microstomia) and associated intellectual disability. Ventilation could be achieved using paediatric mask. Intubation was done in second attempt with help of bougie as there was lesser space in oral cavity for tube negotiation. Till now very few cases reported in medical world of this syndrome with unknown genetic inheritance.*

Key-words: *Hallerman steriff syndrome, Difficult airway, Rare syndrome*

Key Messages: Preoperative finding of Hallerman-Steriff syndrome should alert anaesthesiologist for possibility of difficult airway with intellectual disability of patient. Anaesthesiologist should be prepared for any event because of associated cardiac and neurological anomalies.

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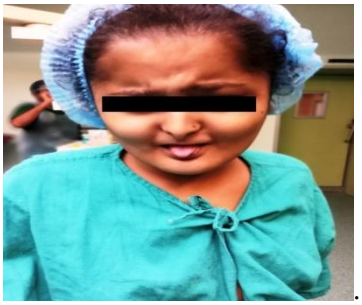
I. Introduction:

Hallermann-Steriff syndrome is a rare, congenital condition. Almost all reported cases appear to have occurred randomly.[1] In 1893, first described by Audry.[2] In 1953, Hallerman and Steriff described clinical features of syndrome. [3]

Till now only 150 cases have been reported in medical world. [4-7] Cardinal features may include a short, broad head, frontal bossing due to delayed ossification of sutures in calvarium, micrognathia due to hypoplastic mandible, highly arched palate, a thin, pinched, beaked nose due to hypoplastic cartilage, bird facies, congenital cataracts, microphthalmia, natal teeth, proportionate short stature.[1,8] .Hypotrichosis (sparse hair) is present in about 80 percent of affected individuals. Intellectual disability is present in some individuals. Cardiac abnormalities are present in 4.8% of patients which may be patent ductus arteriosus, tetralogy of fallot. In some cases, structural defect of brain like absence of corpus-callosum noted.

 . (Figure 1)

Case History: A 21 year old female was posted for laparoscopic cholecystectomy. She was having frontal bossing, microphthalmia, beaked nose, microstomia (figure 1). She was mentally retarded so not communicating with us. Her mother did not give any history of similar features in any family member. No history of snoring was present. She was operated for bilateral congenital cataract at age of 15 years. No significant history related to previous surgery was given. Weight of patient was 50 kg. Mouth opening was 2 and half finger, Mallampatti grading was 3. She was having complete artificial fixed denture. Last menstrual date was 15 days was back. Haematological investigations were normal. Bilateral air entry was equal, no added sounds were present. Heart sounds were normal. She was a diagnosed case of Hallerman-steriff syndrome. Bilateral basal ganglia calcification in CT brain was present so neurophysician advised her to start levetiracetam 250 mg BD 7 days prior to surgery. Echo was normal.

Patient was posted after taking informed written consent from parents. Patient was advised to continue levetiracetam on morning of surgery.

On day of surgery, patient was taken with adequate nil per oral status. In view of difficult airway, all accessories like various sizes of airways, laryngoscopes, endotracheal tubes, stylette, bougie, i-gel, intubating LMA were kept ready. Cannula 18G was taken after convincing patient. All standard monitors applied. Midazolam 1mg, fentanyl 80 µg, propofol 60 mg were given intravenously. As we checked for ventilation, we were not able to ventilate with normal adult size mask as it was too big. Ventilation was possible with paediatric no. one mask, after which succinylcholine was given. On laryngoscopy, Cormack-lehane grade was two. In first attempt, we were not able to intubate the patient. In second attempt, bougie was introduced and 7.5 mm endotracheal tube was railroaded over it. Because of microstomia, high arched palate, artificial denture, lesser space was available to insert the tube and visual axis was lost during first attempt. Anaesthesia was maintained with nitrous oxide, oxygen and sevoflurane. Muscle relaxation was obtained with vecuronium. Injection paracetamol 800 mg intravenously given for pain relief. Procedure lasted for 40 minutes. After return of spontaneous ventilation, patient reversed with injection neostigmine 2.5mg and glycopyrrolate 0.5mg. She was extubated fully awake.

II. Discussion:

Hallerman Steriff syndrome is a rare genetic disorder with unknown inheritance as in our case no positive family history was present. These patients are challenging in view of difficult airway. [8] Microstomia, high arched palate, glossoptosis, history of sleep apnoea, tracheomalacia are factors for difficult airway. Ascending ramus may be absent in these patients resulting in small oral cavity. [9] In our case, no history of snoring and symptoms suggestive of tracheomalacia were present. It was a difficult airway because of microstomia with artificial fixed dentures which posed difficulty in negotiation of endotracheal tube. 15% of patients can have intellectual disability as present in our case. Rarely these patients can have neurological abnormalities as in our case patient was having bilateral basal ganglia calcification. No history of seizures was present however she was started on Tab levetiracetam 250mg BD by neurophysician. In some patients, cardiac abnormalities may be present either congenital or because of right ventricular strain due to sleep apnoea or tracheomalacia. In our case, no cardiac abnormality was present.

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