



## CHALLENGES IN THE AIRWAY MANAGEMENT AND MONITORING OF A PATIENT WITH DYSTROPHIC EPIDERMOLYSIS BULLOSA PRESENTING WITH SQUAMOUS CELL CARCINOMA FOREARM UNDERGOING BELOW ELBOW AMPUTATION -A CASE REPORT

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**KEYWORDS :** Dystrophic epidermolysis bullosa, squamous cell carcinoma, subclavian vein cannulation, Difficult airway.

### OBJECTIVE:

Dystrophic epidermolysis bullosa is a rare inherited disorder, caused by defect in type VII collagen leading to easy peeling of epidermis and dermis. There is widespread blistering and re-epithelisation. DEB is a pre-malignant condition, predisposing to squamous cell carcinoma. We are discussing a case of DEB developing to squamous cell carcinoma in forearm planned for forearm amputation.

### CASE REPORT:

A 32 year old male diagnosed with DEB since childhood had a chronic ulcer in his left forearm which developed to squamous cell carcinoma past 2 years –now planned for amputation. Patient's airway and nasal airway was assessed preoperatively with indirect laryngoscope in order to decide the plan of anaesthesia. Patient was a known case of seizure disorder seizure and was on tab.epoin past 2years. Loading dose of Inj.Fosolin was given preoperatively on the day of surgery. Adequate pre-operative blood transfusion done in view of hemoglobin 5.6g/dl and haemoglobin was improved to 9.4g/dl, other preop laboratory investigations were in normal range. Patient was hemodynamically stable and chest was clear. Mallampati grading was not able to be assessed since mouth opening permitted only one finger, neck movements were restricted. ANAESTHESIA CONCERNS: Fragile "butterfly skin" Securing monitors and life line is difficult as the skin peels off even with ECG leads /micropore Oral cavity is ulcerated and this repeated ulceration causes restricted mouth opening due to contractures at angle of mouth, denuded oral mucosa, degenerative teeth enamel, and bad oral hygiene General condition of patient is poor because of chronic ulceration, protein wasting, infection, sepsis.

### INTRA OPERATIVE MANAGEMENT:

Patient shifted to OR. All pressure areas were padded with cotton to avoid any friction and trauma to the skin. Lubricated gel part of electrocardiography (ECG) was placed on the healthy skin, SpO<sub>2</sub>, and noninvasive blood pressure cuff was applied on better arm padded with cotton. Non-adhesive tape was used to fix the tube. In view of difficult airway, fiberoptic bronchoscopy, video laryngoscope, emergency tracheostomy kit were kept in standby. Under aseptic precaution ultrasound guided right subclavian vein cannulated, triple lumen catheter inserted and secured. 10% lignocaine spray was sprayed in oral mucosa, 2.5 size laryngeal mask airway inserted slowly, and patient was asked to swallow simultaneously. LMA secured in place. Patient was induced with Inj. Propofol 2mg/kg and after checking bilateral equal chest rise and adequate ventilation, patient was paralysed with Inj. Atracurium 0.5mg/kg. Adequate plane maintained with N<sub>2</sub>O+O<sub>2</sub>+sevoflurane (MAC1). Vitals stable. Duration of surgery was 1 hour, blood loss was around 150 ml. Following procedure muscle relaxant was reversed, patient extubated after adequate muscle recovery, tidal volume and fully awake. Patient shifted to post op Ward for observation.

### DISCUSSION:

Epidermolysis bullosa<sup>2</sup> is autosomal inherited or acquired disease characterized by bullae formation in the skin or mucous membranes. The reported incidence is about 20 cases per one million inhabitants. DEB is caused by a defect<sup>7</sup> in type VII collagen. DEB produces severe

scarring<sup>4</sup> of the fingers and toes, ankylosis of the interphalangeal joints, and resorption of the metacarpal and metatarsal joints. Involvement of the esophagus and heart leading to dysphagia, esophageal strictures, dilated cardiomyopathy, and formation of intracardiac thrombi. Hypoalbuminemia, secondary to nephritis and protein loss can also occur. Anemia is due to poor nutrition and repeated infections. Hypoplasia of tooth enamel resulting in degeneration of the teeth. DEB patients rarely survive beyond the third decade. Medical therapy for DEB has not been very successful.

Anesthetic management of DEB patients are always a challenge. In order to avoid skin trauma and mucous membranes, ECG gel pads can be used. Blood pressure cuff should be padded with cotton dressing, intra vascular catheter can be fixed with sutures. Face mask trauma can be minimized with appropriate size face mask and lubrication of face mask. Airway devices<sup>5</sup> can be avoided to minimize oral trauma. If tracheal intubation or supraglottic airway insertion required then adequate lubrication must be applied on laryngoscope, endotracheal tube or LMA. In our case all precautions were taken for successful intubation and extubation with LMA atraumatically.

### CONCLUSION:

Despite of all potential complications careful intraoperative management is associated with few adverse effects. Avoiding trauma to the fragile skin and mucus membranes is the key to success for providing atraumatic anesthetic care to DEB patients. Also atraumatic airway management is very essential for a successful outcome of the surgery. We suggest that DEB patient care should be provided in centers where adequate facilities and expertise are available for taking care of these types of patients.

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