Case Report

Management of severe tracheal stenosis caused by repeated endotracheal intubation in a patient of myasthenia gravis

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Introduction

Myasthenia gravis (MG) is an autoimmune disorder of the neuromuscular junction. The disease is characterized by exacerbation and remission of the symptoms from time to time. Myasthenic crisis is a serious complication of MG and is defined as weakness from acquired MG that is severe enough to require intubations[1]. Myasthenic crisis may complicate in 15-20% of patient with MG[2, 3]. Most patients require ventilation for a brief period, usually less than two weeks[4]. Repeated intubations may cause tracheal stenosis. The potential risk factors for post-intubation subglottic stenosis include, the underlying disease requiring endotracheal intubations(EI), the age and body weight, the duration and number of EIs, absence of sedation and the occurance of infections, hypotensive or hypoxic events during the period of EI and traumatic EI[5, 6].

Key words: Tracheal stenosis, Repeated intubations, Myasthenia gravis.

The Case

A fourteen year old girl was admitted in I.C.U with upper respiratory tract obstruction. She was a known patient of myasthenia gravis diagnosed 3-4 yrs back. She also had Diabetes Mellitus (DM) and Cushing Syndrome.

In ER, she was unconscious with a GCS of 8/15. Respiration was laboured. Tracheal intubation was performed, her SpO_2 was maintained at 87-90% with Bain circuit and $EtCO_2$ was not recordable in capnograph. Respiration was maintained by Bain circuit against very high pressure registance. Initial management included replacement of ETT with under diameter tube but that did not produce reduction of peak inspiratory pressure (PIP). Her SpO_2 was maintained by ventilation with very low Vt (80-90 ml) and higher RR 40-50 (AS HFJV was not available).

It was found out that previously she had been intubated for several times and ventilated for periods of different duration in various hospitals in the city. But the duration of requirement of ventilation increased gradually. This time she was transferred from a different hospital as it was impossible to maintain effective ventilation due to tremendous increase in peak pressure.

The patient was transfered to ICU, reassessed and again her endotracheal tube (ETT) was changed to ventilate her properly. She was put on ventilator and ventilation was found to be very difficult for a very high peak pressure (50-60). A bronchoscopy was done to assess the trachea . A severe stenosis was found at 3-4 cm below the vocal cord. She was planned for tracheostomy to bypass the stenosed area and tracheostomy was done. Still ventilation was difficult, peak pressure was still high. A repeat bronchoscopy showed that the stenosis was below the level of tracheostomy. The site of stenosis is formed by irregular fibrous bands probably from previous intubations. At that time, to save her life, it was decided to dilate the trachea by cutting bands by multi teeth biopsy forceps under the guidance of bronchoscopy. The fibrous bands were removed by repeatedly cutting small pieces and thereby made the tracheal opening larger.

Her SpO_2 was maintained with 100% O_2 but $EtCO_2$ was >80 mmhg. Her blood pressure was maintained with high dose of Inj. Dopamine and she received Inj. Hydrocortisone for her Cushing Syndrome. Her Blood sugar was controlled with IV insulin in syringe pump.

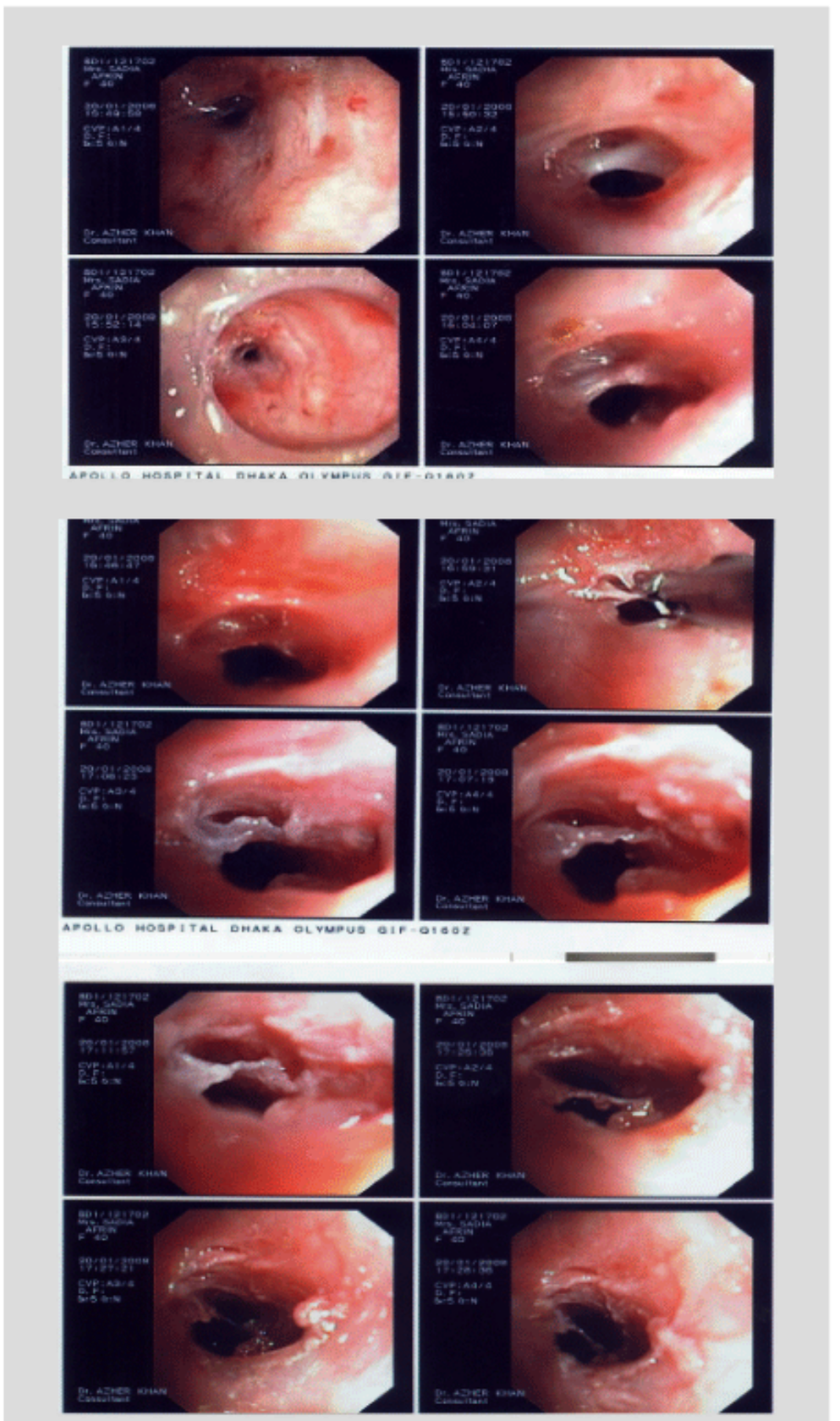

Figure: Serial photographs

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Later she was put into ventilator in Assist Control mode without muscle relaxant & sedation as she was already MG. Gradually the peak pressure of the ventilator came down but it was still higher than normal. With high concentration of O_2 (FiO_2 60-70%) and high pressure support (40 cm of water) her SpO_2 was maintained, at the same time her $PaCO_2$ started to wash out gradually. After 1 day she regained her level of consciousness. DM was controlled with insulin as required. Blood pressure was maintained with Inj. Dopamin & Nor - adrenaline.

On the 3rd day of tracheostomy she was put into SIMV mode and gradually weaning from ventilator as well as from inotrops started. On the 4th day her peak pressure became normal, tidal volume increased then she was put into spontaneous mode with some pressure support.

After 4 days she was weaned out of ventilator and she maintained her SpO_2 in room air through tracheostomy tube with HME filter and blood pressure maintained without inotrops.Then she was shifted to her cabin for care of tracheostomy and specific management of the disease.

Discussion: Repeated endotracheal intubation (EI) may result in significant injury to the larynx and trachea. The potential risk factors for post-intubation subglottic stenosis include, the underlying disease requiring endotracheal intubations (EI), the age and body weight, the duration and number of EIs, absence of sedation and the occurance of infections, hypotensive or hypoxic events during the period of EI and traumatic EI[5, 6]. The severity of stenosis depends upon the degree of involvement of the tracheal wall[7]. Cutting the stenosed part in tiny pieces during bronchoscopy, and repeating the procedure until the total part is cut, keeping the SpO_2 at 97%-100%, is quite safe. Toty L et al reported that if this is not corrective, then a laser resection of the stenosed portion of trachea is performed. Tracheal dilation, stenting and laser resection are only palliative therapies[8]. In this case, with this non-conventional method tracheal stenosis was overcome successfully.

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