Single Stage Management of Hypopharyngeal, Esophageal, and Tracheal Stenosis Complicating Toxic Epidermal Necrolysis

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INTRODUCTION

Toxic Epidermal Necrolysis Syndrome (TENS) is a rare, severe skin reaction precipitated by exposure to medications or viral pathogens. TENS exists at the most severe end of a spectrum of skin reactions which includes Steven’s-Johnson Syndrome (SJS) and erythema multiforme (EM). The spectrum of organ manifestations is diffuse to include the cutaneous skin, gastrointestinal tract, respiratory tract, genitourinary tract, and patients may also have ophthalmic complications. We present the case of a patient referred to ENT clinic with esophageal, hypopharyngeal and tracheal stenosis following a severe case of TENS and the surgical management of these long term sequelae.

METHODS AND MATERIALS

Review of a single patient presenting to a laryngology practice at a tertiary medical center.

RESULTS

A 37 year old female was admitted to the burn unit at San Antonio Military Medical facility with eruption of a bullous rash over 80% of her body surface area (BSA) after ingesting Lamotrigine for migraine headaches. She subsequently required intubation. Her primary team placed tracheostomy tube due to support her respiratory status. Her cardiopulmonary status declined and she required extracorporeal membrane oxygenation (ECMO) for support. The Otolaryngology Service was consulted to address her severe mucosal involvement to include bullous disease of her oral cavity, pharynx and nasal cavity. To prevent nasal stenosis, the ENT service placed nasal stents which were removed prior to discharge. Her condition improved and she was discharged from the burn unit with minimal oxygen requirement. At follow up, she noted continued shortness of breath and progressive dysphagia to solids, with ability to tolerate only liquid diet. Flexible fiberoptic laryngoscopy, esophagoscopy and bronchoscopy along with Stroboscopic exam were performed in clinic. She was noted to have near complete hypopharyngeal stenosis with scar bands completely obliterating her left pyriform sinus and esophageal stenosis at 35cm with inability to pass a 5.5mm TNE scope. Lambdoid tracheal scar bands approximately 10cm above her carina were noted. The patient was taken to the operating theatre and direct laryngoscopy was performed with spontaneous ventilation allowing the tracheal stenosis to be dilated prior to intubation. Two additional dilations were performed with a Controlled Radial Expansion (CRE) balloon. The resultant lambda scar was ablated with microlaryngeal CO2 laser attached to a micromanipulator in a fashion described by Noureil et al. The same balloon was placed into the tight hypopharyngeal stenosis and resulted in no significant impact upon dilation. The surgical team lysed hypopharyngeal scar bands with laryngeal microscissors. The pyriform sinus scar band was considerably thicker than the others. Upon completion of the dissection, two balloon dilators were inserted and distal sections of the scar expanded well. The proximal and distal esophageal scar bands were dilated three times. The patient was placed on oral steroids, proton pump inhibitors, and sulcrate and discharged home the next day. At her one month follow up appointment, the patient noted 95% improvement in her dysphagia, ability to tolerate solids, and improvement in dyspnea.

CONCLUSIONS

TENS is a rare disease and upper aerodigestive tract stenosis in a single stage procedure.

DISCUSSION

TENS is an exceedingly rare condition with an incidence between 0.4–1.5 cases per million person years. In adults, it is more common among women by a ratio of 2:1 with increased incidence among persons over 60 years of age. This bullous skin reaction leads to detachment of the epithelium in over 20% of the body skin surface area and commonly affects mucosal surfaces as well. This spectrum of disease includes SJS which typically involves less than 10% BSA, as well as TENS-SJS overlap syndrome which involves between 10 and 30% BSA. TENS was first described in 1956 by Lyell as a cutaneous eruption similar to burn injury. The initial symptoms usually include fever and cutaneous reactions, dysphagia, or dysphonia. The main presenting symptoms are presented in Table 1.

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