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Laryngeal Lesion of Epidermolysis Bullosa: Topical Use of Mitomycin-C

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1. Abstract

Epidermolysis bullosa (EB) is a mucocutaneous genetic disease characterized by fragility of the dermoepidermal junction. Laryngeal lesions frequently occur in junctional epidermolysis bullosa (JEB), a subtype of EB.

Laryngeal involvement can be life-threatening with a risk of upper airway obstruction. Current management includes repeated endoscopic procedures due to frequent recurrences, and may even require tracheotomy. These invasive procedures are associated with high morbidity and high risk of iatrogenic injuries.

We advocate intraoperative application of topical Mitomycin-C to prevent formation and recurrence of granulation tissue.

We describe two cases of children with laryngeal involvement of JEB. Laryngeal endoscopic surgery was completed by local application of Mitomycin-C. We did not deplore any recurrence or adverse secondary effect for both children.

Endoscopic laryngeal surgery completed with topical application of Mitomycin-C seems to be a safe and effective procedure for JEB laryngeal involvement. It may limit recurrences and prevent iterative and invasive therapeutic procedures.

2. Introduction

Epidermolysis bullosa is a rare genetic disorder characterized by skin and mucosal fragility of the dermoepidermal junction leading to blister formation and erosions.

Several subtypes have been defined according to the histological involvement. Junctional epidermolysis bullosa (JEB) is characterized by a dermo-epidermal fragility with blister formation in the lamina lucida. This form involves severe cutaneous and mucosal lesions [1].

Laryngeal injuries can appear as blisters, erosions or granulation tissue with chronic evolution leading to upper airway stenosis. The clinical course goes from moderate forms with inspiratory stridor, dysphonia and dysphagia to severe forms with dyspnea and respiratory distress [2].

Management of laryngeal lesions requires repeated endoscopic surgeries because of recurrence, and sometimes needs tracheotomy.

These treatments are associated with a significant risk of secondary lesions due to the mucocutaneous fragility of these patients. The investigation of therapeutic alternatives led us to evaluate the use of Mitomycin-C.

Topical Mitomycin-C can be used in ENT endoscopic procedures to prevent scar tissue formation after laryngeal surgery. Topical application of Mitomycin-C inhibits fibroblast proliferation during the wound healing process and thus reduces the risk of granulation tissue formation and stenosis [4, 5].

We describe two cases of children with laryngeal involvement of JEB, managed by endoscopic resection and adjuvant local application of Mitomycin-C.

3. Case Report

3.1. Case no. 1

An 11 months patient presenting with severe laryngeal involvement of Junctionnal epidermolysis bullosa. He was referred to the ENT department for laryngeal dyspnea and feeding difficulties. Nasofibroscopy revealed a voluminous blister inserted on the left ary-epiglottic fold and ventricular region with 50% obstruction of the laryngeal lumen. Despite medical treatment with nebulized and oral corticosteroids, the symptoms progressively worsened, requiring surgical management. Rigid laryngoscopy was performed with specific installation precautions in order to limit iatrogenic injury. The examination revealed a blister of the left ary-epiglottic fold on granulation tissue obstructing 80% of the glottic lumen. The blister was collapsed with cold instruments and we removed granulation tissue. Then topical Mitomycin-C was applied, 1 mg/ml, 2 applications during 3 minutes.

On day-1, the child recovered and was discharged at home.

Follow-up regular clinical examinations for 1 year did not reveal any recurrence of laryngeal lesions.

3.2. Case no. 2

An 8 years-old patient with moderate form of JEB. The child developed laryngeal symptoms, characterized by dysphonia and nocturnal stridor with snoring and symptoms of obstructive sleep apnea syndrome (OASA). The nasofibroscopy revealed granulation tissue inserted on the left vocal cord obstructing half of the glottis lumen (Figure 1). Surgical rigid laryngeal endoscopy was performed with cold instrument excision of the granulation tissue, which resolved the symptomatology. Three months after surgery, dysphonia and OSAS symptoms appeared again. Nasofibroscopic examination revealed recurrence of the granulation tissue on the left vocal cord. We performed a new surgical management completed with local application of mitomycin C, 1mg/ml, 2 applications during 3 minutes.

No recurrence occurred during the 2-years follow-up following surgery.

Figure 1:

4. Discussion

Current management of JEB laryngeal involvement requires iterative endoscopic procedures and may sometimes lead to tracheostomy. Liu et al described the case of a JEB patient who required 20 endoscopic procedures over a 4-year period [7]. The significant morbidity of the treatment is explained by the cutaneo-mucosal fragility of these patients.Special precautions must be applied during the procedure to limit the risk of secondary lesions [6].

A study estimates a 10 to 23% death rate related to upper airway obstruction in JEB. Some authors justify the need to perform a preventive tracheotomy for patients with laryngeal involvement [8]. Performing tracheotomy is not without any risk: cannula rubbing, cutaneous fixation and repeated aspirations are important sources of secondary injuries. Tracheostomy must be considered as the last resort.

Usually, endoscopic laryngeal procedure are realized with cold instruments management and postoperative inhaled and systemic corticosteroids therapy [9].

For our two patients, we used Mitomycin-C with the intention to improve postoperative healing and to reduce recurrences.

Mitomycin-C is an antibiotic produced by Streptinomyces caespitosus that shows antiproliferative and antineoplastic properties.The efficacy of topical use of Mitomycin-C to prevent granulation tissue formation and laryngeal stenosis still remains debated. However, positive results of some studies, its simplicity of use, and the lack of complications of topical application contributed to its frequent use in laryngeal management [10]. Roh et al and Simpson et al described a decrease of laryngeal stenosis cases following topical application of Mitomycin-C 0.4 mg/ml for 5 minutes after endoscopic management of laryngeal lesions. They did not report any complication related to Mitomycin-C [11,12].

Some complications were described by Hueman et al after topical application of Mitomycin-C at a high concentration of 10 mg/ml: 2 patients out of 14 developed an accumulation of fibrinous tissue requiring a new procedure [13].

In our cases, we used a concentration of 1 mg/ml which provided a successful outcome with no recurrence and no secondary complications. There were no adverse effects.

5. Conclusion

The management of JEB laryngeal lesions is complex. These lesions are potentially life-threatening. Multiple managements may induce severe morbidities.

We advocate that topical application of Mitomycin-C combined with endoscopic surgery for laryngeal blisters is a safe and effective procedure.

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