Recurrent Respiratory Papillomatosis: our Experience at Queen Rania Al-Abdullah Hospital for Children at the Royal Medical Services

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Abstract

Objective is to evaluate our clinical experience in dealing with recurrent respiratory papillomatosis (RRP) at Queen Rania Al-Abdullah Hospital for children. A total number of 9 patients (8 males, one female) were diagnosed by means of history; physical examination and flexible nasopharyngolaryngoscope. Patients age at time of presentation were 2-10 years. The presenting symptom was just hoarseness of voice in 5 patients (group 1). Hoarseness and dyspnoea on exertion in 3 patients (group 2) and respiratory distress in only one patient (group 3). All 9 patients had been treated by microdebrider excision using apnoic technique. In 3 of patients (group 2) we injected MMR vaccine intralesional and we injected sometimes MMR vaccine and sometimes Cidofovir intralesional in only the most severe case (group 3). In group 1 and 2 the post operative symptom free period was 6-8 months. In group 3 the symptom free period was 4-5 weeks when MMR was injected and 10-12 weeks when Cidofovir was injected. Recurrent Respiratory Papillomatosis (RRP) is a benign neoplasm caused by HPV, it affects mainly children, it has high recurrence rate with remissions. Best results are achieved using microdebrider and Cidofovir intralesional injection.

Key words: Recurrent respiratory papillomatosis, Microdebrider, Cidofovir.

Introduction

Recurrent respiratory papillomatosis was first described in the 17th century as “warts in the throat” [1]. It is caused by the different types of the human papilloma virus. The greater incidence of onset is before the age of 5 years which is called the Juvenile form or the Juvenile Recurrent Respiratory papillomatosis or childhood-onset RRP, while there is about one third of patients their first presenting symptom is during adulthood [2]. Recurrent respiratory papillomatosis (RRP) is caused primarily by human papillomavirus (HPV) types 6 and 11, with
all other HPV types causing less than 2% of disease [2]. Warty growths in the upper airway may cause significant airway obstruction or voice change. The interval between surgical interventions varies between patients, ranging from 3 weeks to several years. Differences in host immune responses to HPV infection may explain this variability.

The papillomas are histologically benign neoplasms, which often recur after surgical removal and can produce sudden respiratory obstruction and become life-threatening. In the most severe cases, the disease may fluctuate in severity and may enter remission after several years, or persist into adulthood. The disease is most common on the vocal folds but may extend to other sites (trachea, lung) in the respiratory tract. In rare instances, the papilloma may undergo malignant transformation.

There is an etiologic link between maternal condyloma at delivery and childhood-onset RRP in the infant and found that more than 50% of mothers of childhood-onset RRP cases gave a history of having condylomas during pregnancy and/or at delivery[1,2]. Patients with childhood-onset RRP are more likely to be firstborn and vaginally delivered than are control patients of similar age [3, 4]. One hypothesis is that primigravid mothers are more likely to have a long second stage of labor, and this results in prolonged exposure of the fetus to the virus. If newly acquired genital HPV lesions are more likely to shed virus than long-standing lesions, this would explain the higher incidence of papilloma disease among offspring of younger mothers [3, 4].

Material and Methods

A total number of 9 patients (8 males, one female) were diagnosed by means of history; physical examination and flexible nasopharyngolaryngoscopy. Patients age at time of presentation were 2-10 years. The presenting symptom was just hoarseness of voice in 5 patients (group 1). Hoarseness and dyspnoea on exertion in 3 patients (group 2) and respiratory distress in only one patient (group 3). In 8 children the site of lesion was only glottis (group 1 a& 2). In the most severe case (group 3), the papillomas were found in the nasal cavities, naso- and oro-pharynx, trachea, bronchi and oesophagus. In 8 children there was a positive family (parents) history of vaginal or penile warty lesions and only one child with history of maternal rectal warty lesion, but parenteral work-up was not analyzed. All 9 patients had been treated by skimmer laryngeal microdebrider excision using the zero degree telescope. During surgical procedure an initial examination of the whole aerodigestive tract (nasal cavities, naso- and oro-pharynx, trachea, bronchi and oesophagus) was always performed. Suspension laryngoscopy was used. At the first operation always biopsy material was obtained for histological proof of the diagnosis. During removal there was no hesitation in taking the surface mucosa adjacent to the base of the papilloma. In 5 of patients (group 1), no intralesional injection was used and papillomas were few and limited to the vocal cords. In 3 of patients (group 2) we injected 3 ml MMR vaccine intralesional in every operation and papillomas were multiple and aggressive but still limited to the glottic area. In the severe case (group 3) we injected sometimes 3 ml MMR vaccine and sometimes 75 mg Cidofovir intralesional after removing the multiple papillomas from the supra- and the glottic area, naso- and oropharynx. The nasal cavity and oesophageal papillomas were left untouched.

Anaesthetic techniques

All operations were performed using general anesthesia and spontaneous respiration without anesthetic tube insertion in the larynx. Apnoic technique was used with the need to insert a small diameter endotracheal tube frequently in order to raise the O2 saturation.

Results

In group 1 and 2 the post operative symptom free period was 6-8 months. In group 3 the symptom free period was 4-5 weeks when MMR was injected and 10-12 weeks when Cidofovir was injected. The maximum number of surgeries is 32 surgeries in 4 years in the most severe case (group 3).

Complications of surgery: Only one patient (from group 2) got a small anterior laryngeal web not causing any significant symptom.

Discussion and Conclusion

Recurrent Respiratory papillomatosis is a frustrating disease to treat. The goals of surgical therapy are to maintain a clear airway while avoiding excessive scarring and maintaining useful vocal cord function [5]. A disease confined to the larynx has a better outlook than that which is more widespread. Panendoscopy under general anaesthesia
at the initial examination is important [6]. No single type of therapy has been consistently effective in eradicating RRP [5]. The use of powered instrument is a viable alternative to laser excision [7] and clearly has become the preferred surgical method. The use of the zero degree telescope without anaesthetic tube gives excellent visualization of the papillomas and the laryngeal structures. With the use of powered instruments the postoperative vocal cord edema is minimal and the voice improvement is significant. The use of Cidofovir injection intralesional offers significant improvement and delay in recurrence despite the fact that we use it just in only one child with the severe form of RRP. The recurrence of symptoms and the need for surgical excision was 10-12 weeks when Cidofovir was used while it was just 4-5 weeks when MMR vaccine was used and not more than 3 weeks when no intrallesional injection was used.

Future research is needed regarding prevention of transmission of HPV from mother to child. Specifically, the roles of caesarean section [5]. The rarity of cesarean delivery in RRP cases is indirect evidence that the cases may be preventable by cesarean delivery. The data from literature identify being first-born, and birth to a teenage mother as additional risk factors for RRP [8]. The longer labor in first order birth could potentially increase the time of contact between the fetus and infected maternal secretions [9, 10, 11].

Finally, universal or near-universal use of an HPV vaccine that provides protection against HPV 6 and 11 may do for RRP what the Haemophilus influenzae type B (HiB) vaccine has done for H. influenzae type B epiglotitis, virtually eliminating new cases in less than a decade [5].

References
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