

CASE REPORTS

RECURRENT RESPIRATORY PAPILLOMATOSIS: THE KORLE-BU EXPERIENCE

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SUMMARY

Background: Recurrent respiratory papillomatosis is a benign disease of the aero digestive tract which is caused by the human papilloma virus type 6 and 11 and mainly affects children.

Objective: The aim of this study is to describe the pattern of recurrent respiratory papillomatosis at the E.N.T Unit Korle Bu Teaching Hospital, Accra.

Method: This is a retrospective study of theatre records of patients managed for respiratory papillomata from January 1995 to December 2004 at the E.N.T Unit of Korle Bu Teaching Hospital, Accra which is a tertiary facility. These records were studied and we obtained information on gender, age, number of surgeries and the presence of tracheotomy.

Results: Sixty-nine patients were identified over the period. Thirty-three males and 36 females with a M: F ratio 1:1.1, and ages ranging from 2 to 54 years. The mean age was 12.3 years and a median age of 8.5 years. Children less than 10 years accounted for 69% and 46.4% had repeated surgery with overall tracheotomy rate of 14.5%.

Conclusions: Recurrent Respiratory Papillomatosis is primarily a paediatric disease. Locally, its distribution is comparable to others in the sub region and worldwide and surgery is the mainstay of treatment.

Keywords: Recurrent respiratory papillomatosis, Tracheotomy, ENT, Paediatrics

INTRODUCTION

Recurrent respiratory papillomatosis (RRP) is a benign disease associated with exophytic lesions in the airway. It is caused by the Human Papilloma virus types 6 and 11 (HPV 6 and HPV 11)¹ which is also implicated in genital condylomata. There is an association between cervical HPV in the female and respiratory papillomatosis. It has a worldwide distribution and malignant transformation has been reported in adults.^{1,2}

It is the commonest benign neoplasm of the larynx in children causing persistent hoarseness and progressive upper airway obstruction.^{1,3-6}

It may be misdiagnosed as asthma or croup leading to late diagnosis of RRP with resultant acute airway obstruction.^{1,7} Some of these patients may require tracheotomy for immediate breathing and further airway management^{3,6,8}.

A good history of persistent hoarseness with progressive difficulty in breathing especially in a child is pathognomonic. This can be complimented with indirect laryngoscope and if it cannot be tolerated then flexible fiberoptic nasolaryngoscopy can be used. Surgical management of this condition is by direct laryngoscopy under anaesthesia and removal of the papillomata by cold steel method, laser or with a microdebrider and subsequent confirmation by histopathology. This disease has a high rate of recurrence leading to multiple surgeries especially when diagnosed before 3 years of age.⁸

Adjuvant therapy with mitomycin C⁹, cidofovir, and alpha-interferon¹ has been used with some favourable results. There is currently no local data on RRP and it has become necessary to study this disease condition. The aim of this study is to describe the pattern of RRP at the E.N.T Unit, Korle Bu Teaching Hospital, Korle Bu Accra, Ghana.

METHOD

This is a retrospective study of theatre records of patients managed surgically for laryngeal or recurrent respiratory papillomata at the ENT Unit of the Korle Bu Teaching Hospital, Accra from January 1995 to December 2004. Information gathered included age, gender, number of surgeries per patient and surgical modalities of management. Data analysis was done using descriptive method.

RESULTS

Over the 10-year period 69 patients with respiratory papillomatosis were identified. They had been managed surgically by cold steel de-bulking and specimens sent for histopathology which confirmed the diagnosis. There were 33 males and 36 females indicating a male:female ratio of 1:1.1. A mean age of 12.3 years (range 2-54 years) was calculated with a standard deviation of 12.2 and a median of 8.5 years. Forty-eight (69.7%) of the 69 patients were children up to 10 years old.

There were no recurrences requiring further surgery in 37 (53.6%) of all the patients, while 32 (46.4%) required more than one surgical intervention for the respiratory papillomatosis. These include 20 who had up to 5 surgeries, 8 who had between 5 and 10 surgeries and 4 who had more than 10 surgeries. The cumulative tracheotomy rate was 14.5% (10) of the 69 patients seen. These include 4 out of 4 who had more than 10 surgeries, 2 out of the 8 who had 5-10 surgeries and 4 out of the 20 who had up to 5 surgeries.

DISCUSSION

Recurrent respiratory papillomatosis (RRP) has a worldwide distribution with almost equal distribution between the sexes. In our study the male to female ratio of 1:1.1 is comparable to other reported cases of 1:1.3¹⁰ in the West African sub-region. Onset of this disease can be in childhood where it is called juvenile onset RRP or in adulthood (adult-onset RRP). We noted that 69.7% of the patients in our series were 10 years or younger and this is similar to the findings of Mgbor et al of 51.8%¹⁰ and Nwaorgu et al 74.4%⁷ and thus confirms the paediatric nature of the disease.

The natural history of RRP is variable with some patients having spontaneous regression and others suffering aggressive papillomatous growth requiring multiple surgical procedures over many years.¹ No recurrence was reported in 23% of patients by Nwaorgu et al⁷ and we noticed that in 53.6% of our patients, but we are limited in our follow up systems such that some of these who did not report back may have died.

The issue of tracheotomy arises when alternate airway is needed when patients come in stridor. We are reporting an overall tracheotomy rate of 14.5 % which compares to 18.5 % by Mgbor et al.¹⁰ but lower than 25.64% by Kpemessi et al.⁶ and 70 % by Nwaorgu et al.⁷ This we attribute to our patients coming a little earlier than the reported series who acknowledge the late presentation as the reason for urgent tracheotomy.

In our series it is noted that the tracheotomy rate increased with increased surgical interventions and this can be due to the aggressive nature of the papillomata⁸ as well as parent fatigue.

It is a huge financial strain on parents for their children to have frequent admissions for surgery and sometimes they try to buy time by delaying the inevitable with the consequences of upper airway obstruction leading to a tracheotomy being done. Tracheotomy should not be encouraged but may be unavoidable where there are limitations to accessing medical care, since it can lead to seeding of the papilloma into the trachea.⁸ However, tracheal disease has not been reported in our local series. If tracheotomy is done decannulation should be considered as soon as the disease is managed effectively with endoscopic techniques.¹

Current research is into the genes that govern susceptibility and the possible use of vaccination in preventing this disease.¹ We acknowledge that more work needs to be done especially in the typing of the local HPV to access the possible benefits from the quadrivalent (HPV type 6,11,16,18) vaccine available.¹ Surgery to maintain an adequate airway is the mainstay of treatment. Managing this disease is frustrating because of its natural history of recurrence leading to multiple surgeries and its consequent financial implications.

CONCLUSIONS

Recurrent respiratory papillomatosis is primarily a paediatric disease. Its local distribution compares with others in the sub-region and worldwide. Surgery to maintain the airway is the mainstay of treatment.

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