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Editorial

All About Juvenile-onset Recurrent Respiratory Papillomatosis (JORRP)

Ankit Mishra*

Consultant and Assistant Professor, Department of ENT-HNS, LN Medical College and Research Centre, Bhopal (MP), India

***Corresponding Author:** Ankit Mishra, Consultant and Assistant Professor, Department of ENT-HNS, LN Medical College and Research Centre, Bhopal (MP), India.

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Introduction

Recurrent respiratory papillomatosis (RRP) is caused by human papillomavirus (HPV). RRR is characterized by recurrent papillomas occurring in the respiratory tract, predominantly larynx. It has two types of clinical presentations – juvenile-onset RRP (JORRP) found to occurs under 12 years of age, and AORRP i.e. adult-onset RRP. JORRP predominates in region like sub-Saharan Africa, whereas in Europe and American continents AORRP predominate.

Although there is low incidence, but papillomas that are associated to Human Papilloma Virus (HPV) 6 and HPV11 caused most common benign tumor of larynx, almost 84% of cases. These are mainly papillomas, limited to larynx. The natural history of recurrent respiratory papillomatosis (RRP) is unpredictable, some patients present has a mild symptoms which resolves spontaneously and get cured in a single intervention, while some are quite an aggressive disease which has multiple recurrences for which multiple surgical interventions are needed. The latter is less commonly seen, it has incidence of only 1.8 cases per 100,000 in adults population and 4.3 per 100,000 in pediatric population. Very few numbers of patients present its malignant transformation as well.

It is believed that JORRP is vertically transmitted to the child, i.e through the mother's infected genital tract during birth. In a case-control study of 3033 Danish births the risk of JORRP is 231 times high in children whose mother had genital warts during gestational period, on the other hand urogenital dissemination and subsequent re-activation of HPV which was acquired during childhood is responsible for AORRP.

In 2008 a systemic review conducted by Gélinas., *et al*. it was found that 83% of cases were diagnosed during 0- 4 years with median being 2 years, ranging from 2 months to 19 years. Niyibizi., *et al*. in the year 2021 conducted a study in which he analysed 19 studies to look for the age of diagnosis in patients presenting with JORRP, according to which mean age of onset was 2-9 years. By looking at above mentioned studies it can be concluded that JORRP is more aggressive than AORRP, and it is more prone to a poorer clinical course when its onset is 0-3 years of age.

Diagnosis

All patients present with chronic symptoms like change in voice, stridor, respiratory distress. All such patients should undergo fibreoptic laryngoscopic examination or in case dependent manner direct laryngoscopy and biopsy should be considered. On laryngoscopic visualisation it appear as exophytic mass, pedunculated, which can be single or at times multiple also.

Unfortunately, these facilities and experts to perform these procedures are not readily available in many developing nations and rural areas.

In developing countries as there is high incidence of tuberculosis, laryngeal tuberculosis may sometime mimic as respiratory papillomatosis. On histology, papillomas are finger-like projections comprised of stratified squamous epithelium in connective tissue stroma that has abnormal keratinization and hyperplasia of basal cell. While investigating JORRP X-ray chest or HRCT chest scan should also be done, especially for those with a clinical suspicion of pulmonary involvement.

Staging

The Derkay staging system is most widely accepted system for staging of JORRP. This system includes functional assessment of clinical features and anatomic assessment of its distribution. Although there is limitation of this staging system, it is effective in describing the presence of disease, number of sites involved and the bulkiness of lesions, it does not distinguish the various degrees of severity within the site. Shorter intersurgical interval is indicated by high anatomical score.

Human papilloma virus and JORRP

On exploring the literature, it has been observed that. Most studies have found HPV11 positive disease to be more aggressive as compared to HPV6. Hence HPV typing can be used as a potential tool for predicting disease aggressiveness. However, it is also observed that the younger the age at diagnosis, more aggressive is the disease, age at diagnosis is also established as a more significant marker of disease aggressive nature than HPV type. Intratypic high-risk variants of HPV have specific geographical distribution with differing pathogenic potential.

Managing JORRP

By the date this article is being written no definite cure for RRP is present. Management consists of repeated microlaryngoscopic surgeries procedures till the time when the patient spontaneously go into remission. Papilloma removal is performed either with a cupped forceps or laser (CO_2, KTP). Role of powered instruments like microdebrider and coblator is also recommended. In developing countries, mostly it is done by using cold instruments is. The aim is to remove the papillomas and preservation of the normal laryngeal mucosa. The use of laser or microdebrider is not found to be increase the time between surgical interventions. However, in terms of voice treatment with microdebrider gives better result than CO_2 laser. Patients in developing countries often suffer and travel long distances to undergo surgical procedures, this ultimately is a considerable social as well as financial burden on the family that too when this disease is prevalent in low socioeconomic status.

Complications due to multiple surgeries are frequent like anterior commissure synechiae, glottic stenosis, and granuloma formation. They result in abnormal voice in the long term. Even when all visible papillomas are removed, the disease may recur as HPV DNA is present in the adjacent uninvolved laryngeal mucosa as well.

Malignant transformation

Malignant transformation of papilloma to squamous cell carcinoma, is a rare event that, as per literature occurs in 0.5%, and may be found anywhere in respiratory tract including the larynx, bronchi, or even in the lungs. Malignant transformation is more frequent in elder children, time interval between the diagnosis of JORRP and transformation into malignancy is somewhere near 19 years. However, cancer develop in around 16% patients of RRP with pulmonary involvement, predominantly squamous cell carcinoma. Above discussion suggests that children who have pulmonary involvement due to JORRP are more prone to malignant transformation than those with only upper airway disease.

Conclusion

In JORRP aggressiveness is related to three strong factors:

- Age of onset or age at diagnosis;
- HPV typing: HPV11; and
- Multiple resections/Interventions.

Furthermore, JORRP can be a fatal disease as a result of acute airway obstruction, due to pulmonary spread or due to malignant transformation in papillomatous lesions. A significant limiting factor in the early diagnosis and treatment of children suffering from JORRP in developing countries is, non-availability and inaccessibility of healthcare services, particularly ENT specialist.