Recurrent Respiratory Pappillomatosis: National Registry

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Received: 5 March 2012 / Accepted: 9 March 2012 / Published online: 29 March 2012
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Abstract Recurrent respiratory pappillomatosis is a rare condition with unpredictable nature of disease presentation and behaviour. This write up highlights our institutional experience and the importance of having a national registry for the same. An existing registry created by our team is introduced to facilitate maximum participation by our national otolaryngologists.

Keywords Recurrent · Respiratory · Pappillomatosis

Recurrent respiratory papillomatosis (RRP) is mostly seen in otolaryngology practice where small tumours grow in the respiratory tract (predominantly the larynx) of children as well as adults. Human papilloma viruses (HPV 6 & HPV 11) are reported to be significantly associated with this condition in a causal relationship. There is no cure of RRP but (repeated) surgical excision is an accepted method of controlling the disease, or else these tumours have a tendency to grow ultimately blocking the patient's airway. The occurrence of RRP is quite rare. This paper highlights our institutional experience of RRP and introduces an existing national registry for the same.

Aim of the Study

1. To highlight the changing pattern of incidence of RRP during past 3 decades.

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2. To focus the inconstant behaviour of the disease in terms of aggressiveness reflected by the recurrence, curability and the need for additional airway in the long run.

Materials and Methods

A retrospective review of cases in the past 20 years (1972, 1973, 1981–1987, 1989–1996, 2001–2003) was undertaken at department of Otolaryngology, K.G Medical College (CSMMU) Lucknow. The preliminary data about 55 cases was obtained by our index-records. The incidence of disease month-wise was noted along with the duration of disease, recurrence, time to recurrence, treatment undertaken at every visit, need for tracheostomy, frequency of recurrences, associated treatment and overall follow up including quality of life in terms of functional speech and tracheostomy dependence.

Observations and Results

Only 55 cases could be registered during a period of 20 years at our 100-year old institution that happens to be the biggest referral centre in Uttar Pradesh. Our institute is the oldest one in the north India that drains patients from adjoining states and Nepal. Hence considering this fact, the incidence rate seems to be unexpectedly low. The duration of symptoms during first admission ranged from 2 months to 15 years. The annual number of cases varied from 0 to 11 (Table 1). In general the trend of decreasing incidence has been appreciated in the 1990s and is further accentuated in the last decade (Table 1). The males in general dominated females with a marginal difference (Table 2).

Table 1 Incidence of cases

Year Month	1972	1973	1981	1982	1983	1984	1985	1986	1987	1989	1990	1991	1992	1993	1994	1995	1996	2001	2002	2003
Jan	0	0	2	0	1	0	0	0	0	0	0	0	0	0	0	1	1	0	0	0
Feb	1	1	0	0	1	0	0	1	0	2	1	0	0	0	1	0	0	0	0	1
Mar	1	0	2	0	1	0	0	0	0	0	0	0	1	0	1	0	0	0	0	0
April	1	0	0	0	2	3	0	1	0	1	2	2	2	0	0	1	0	0	0	0
May	1	0	0	0	0	0	1	1	1	2	1	0	0	0	0	0	0	0	0	0
June	2	0	0	3	0	0	1	0	2	0	0	0	1	0	1	0	0	0	0	1
July	1	2	0	0	1	1	2	0	0	0	0	0	0	0	1	1	0	0	1	0
Aug	0	0	1	0	1	0	1	1	2	1	1	3	0	1	1	1	0	0	0	0
Sep	1	1	0	0	1	0	0	3	1	1	2	1	1	1	0	1	0	0	0	0
Oct	1	0	0	1	0	0	3	0	1	0	0	1	0	1	0	0	2	0	0	0
Nov	0	0	0	0	1	0	0	2	1	2	0	0	1	1	0	2	0	0	0	0
Dec	0	0	0	0	0	2	0	0	1	2	2	1	1	2	0	0	0	0	0	0
Total	9	4	5	4	9	6	8	9	9	11	9	8	7	6	5	7	3	0	1	2

Only 55 % patients were fortunate enough not to show any recurrence that however was seen in other cases (45 %) ranging from 1 to 8 (Table 3). The total number of recurrences in the latter group was 119. Amongst those that recurred, majority (44 %) revealed a single recurrence within a time period of 2–60 months, while 20 % showed 6 or more recurrences with in 32–93 months duration.

Table 2 Sex distribution/ratio

Year	Male	Female	Ratio
1972	5	4	5:4
1973	1	3	1:3
1981	3	2	3:2
1982	3	1	3:1
1983	6	3	2:1
1984	5	1	5:1
1985	3	5	3:5
1986	5	4	5:4
1987	4	5	4:5
1989	8	3	8:3
1990	8	1	8:1
1991	6	2	3:1
1992	6	1	6:1
1993	4	2	2:1
1994	3	2	3:2
1995	4	3	4:3
1996	3	0	3:0
2001	0	0	–
2002	1	0	1:0
2003	2	0	2:0

Another 32 % revealed 2–3 recurrences with in 21–62 months, while only 4 % showed 4–5 recurrences. The treatment undertaken was primarily endoscopic surgical excision under general anaesthesia and none was subjected to alternative forms of treatment(s). The need for tracheostomy did not correlate with the frequency of recurrences although none without recurrence underwent such procedure. It was carried out only in four cases. There were six dropouts in this study and a single case was referred to chest surgery department for a possible extension in the lower respiratory tract. A single case however became medically serious to be transferred to pediatrics for further management following surgical resection. The prognosis in the rest of the patients was satisfactory as per the criteria of rendering them clinically asymptomatic. However those with persistent tracheostomy were rendered communicative by instituting a speech valve in the tracheostomy tube.

Table 3 Frequency of recurrence

No. of recurrence	No. of patients		Total
	Males	Females	
1	12	8	20
2	7	1	8
3	2	2	4
4	2	1	3
5	1	0	1
6	0	0	0
7	1	1	2
8	2	0	2

Table 4 Coltrera/Derkay staging/severity scheme

PATIENTS INITIALS : _____ DATE OF SURGERY _____ SURGEON _____

PATIENT ID # _____ INSTITUTION _____

1. How long since the last papilloma surgery? _____ days _____ weeks _____ months
_____ years _____ don't know
_____ this is the child's first surgery

2. Counting today's surgery, how many papilloma surgeries in the past 12 months? _____

3. Describe the patient's voice today :
nomal _____ (0) **abnormal** _____ (1) **aphonic** _____ (2)

4. Describe the patient's stridor today :
absent _____ (0) present with activity _____ (1) present at rest _____ (2)

5. Describe the urgency of today's intervention :
scheduled _____ (0) elective _____ (1) urgent _____ (2) emergent _____ (3)

6. Describe today's level of respiratory distress :
none _____ (0) mild _____ (1) mod _____ (2) severe _____ (3) extreme _____ (4)

Total score of questions 3 – 6 = _____

LARYNX

Epiglottis _____

Lingual surface _____ Laryngeal surface _____

Aryepiglottic folder : Right _____ Left _____

False vocal cords : Right _____ Left _____

True vocal cords : Right _____ Left _____

Arytenoids : Right _____ Left _____

Anterior commissure _____ Posterior commissure _____

Subglottis _____

TRACHEA :

Upper one-third _____

Middle one-third _____

Lower one-third _____

Bronchi : Right _____ Left _____

Tracheotomy stoma _____

OTHER :

Nose _____

Palate _____

Pharynx _____

Esophagus _____

Lungs _____

Other _____

TOTAL SCORE ALL SITES : _____ TOTAL CLINICAL SCORE : _____

Discussion

A single institution experience seems to be insufficient to conclude the true clinical behaviour of these patients.

There is a wide variation in the presentation especially in terms of duration of symptoms. The associated unpredictable recurrence amongst the existing health disparities contributes to a certain degree of non-compliance with

follow-up. The low degree of dropouts in our observation possibly signifies the grave nature of symptoms associated with RRP. Hence it may at times pose a threat to those living far away from a tertiary health care centre. The existing adjuvant therapies (interferon alpha [1], photodynamic therapy [2], indole-3-carbinol [3], cis-Retinoic acid, acyclovir, ribovarin [4], cidofovir [5] and therapeutic vaccines) are too expensive to be adapted routinely in Indian perspective. The prognosis on the other hand including recurrences is quite unpredictable with the set of available data. It may seem reasonable that the institution of adjuvant therapies may reduce the aggressive nature of the disease making the recurrences less likely as well as lessen the severity of symptoms. However the decreasing trends of incidence as in our set up remains unexplained with our limited epidemiological evidence. The current increasing immunization (e.g. against mumps), better obstetric / antenatal care or judicious antibiotic usage may possibly be a few of the critical factors involved. The existing management (and associated health) policies for Indian patients are governed by existing norms of the western world. It may be possible to have racial, ethnical and biological differences in the disease behaviour in our subcontinent. Hence it seems reasonable to accumulate sufficient data to predict a better overall prognosis. Accordingly there is a need to establish a national registry for Indian population as such, which is likely to further enhance the research potential and create sufficient scope to refine health policy management-guidelines for a better patient care and follow-up.

Introduction to National Registry

A national registry (www.irrregistry.in) exists in this regard that initially aims to collect the relevant clinical data

only. All otolaryngologists (of India) who are actively involved in management of RRP either in government or private hospital are invited to help strengthen this online registry. After one time registration (free of cost) a single page patient information as per Coltrera/Derkay staging (Table 4) and severity Scheme [6] needs to be submitted online. The same surgeon-registration ID (permanent) would be used further to submit further information of a followed-up or upfront case. The figure shows the view of the home page of the website. The site can be entered by clicking the 'login' icon and subsequently the otolaryngologist needs to sign up (one time only) accordingly. The website is extremely user friendly and a single entry of data hardly takes 5–10 min.

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