Laryngeal diphtheria: still a problem

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ABSTRACT

**Background:** Globally diphtheria has been showing a declining trend due to effective childhood vaccination programmes. However, inadequate healthcare delivery systems and other social factors have led to diphtheria re-emerging and remaining endemic in many regions of the world. Securing airway by emergency tracheostomy plays a pivotal role in the management of laryngeal diphtheria in addition to antitoxin.

**Methods:** This was a retrospective study conducted in Department of ENT, Maulana Azad Medical College from January 2015 to December 2018. The study included cases of diphtheria with stridor or impending airway obstruction. The information collected included age, gender, socioeconomic status and immunization status. The month of presentation was noted to establish the seasonal trend of the infection.

**Results:** Maximum number of cases was in the age group of 3 to 9 years (62.3% of total cases). Male children predominated over female children with overall sex ratio of 3:1. Most of the cases presenting to us were partially immunized (51.8%) or unimmunized (44.6%). The maximum number of cases was reported in the month of August to November (96 out of 122 cases). Emergency tracheostomy was done in 86.9% (106 out of 122 cases).

**Conclusions:** Diphtheria still continues to be a major public health issue in India. Early recognition and treatment are necessary to prevent morbidities and mortalities. Tracheostomy is a speedy and efficient lifesaving procedure in the management of laryngeal diphtheria.

**Keywords:** Diphtheria, Immunization, Stridor, Tracheostomy

INTRODUCTION

Laryngeal diphtheria is one of the causes of stridor in pediatric age group. It is a highly contagious disease caused by toxin-producing strains of Corynebacterium diphtheriae (or rarely by Corynebacterium ulcerans or Corynebacterium pseudotuberculosis) and transmitted through respiratory droplets during close contact. Although primarily infecting the pharynx, tonsils and nose diphtheria may affect other organs far from the initial area of infection, such as the heart, nervous system, and kidneys. It usually localizes in the upper respiratory tract, ulcerates the mucosa, and induces the formation of an inflammatory pseudo membrane which is implicated for the cause of stridor due to mechanical airway obstruction.

Globally diphtheria has been showing a decline in trend due to effective childhood vaccination programmes. However, inadequate healthcare delivery systems, poverty and other social factors have led to diphtheria re-emerging and remaining endemic in many regions of the world. Diphtheria still prevails in several countries in Africa, the eastern Mediterranean, eastern Europe, south America, southeast Asia and the south Pacific. Nearly half of the diphtheria cases reported globally during
2001-2015 were from India. Central bureau of health intelligence (CBHI) data showed that during 2005 to 2014, 41672 cases were reported with 897 deaths in India. The national family health surveys during 2015-2016 reported coverage of three doses of diphtheria vaccine to be 80%.\(^2\) Massive immunization coverage has led to significant changes in the age wise distribution of immunity pattern. With widespread immunization, diphtheria became rare and exposure to these bacteria became uncommon. Adults, who did not have natural exposure to diphtheria causing organism or received booster dose diphtheria toxoid, became susceptible to disease as the immunity induced by childhood immunization weaned off. But in developing countries like India where the pool of immunized person is not yet large, immunity is still maintained through natural mechanisms like frequent skin infections caused by C. diphtheria.

Change in age distribution is also contributed by other factors like improvement in socio-economic status and changing lifestyles.

Successful treatment of diphtheria depends on rapid administration of equine diphtheria antitoxin in combination with antibiotics. However, if there are signs of impending airway obstruction such as inspiratory stridor, fast respiratory rate, desaturation or cyanosis, immediate steps must be taken to secure the airway by intubation or tracheostomy. One should never wait too long as intervening early has better final outcome. There are considerable controversies regarding the merits and demerits of tracheostomy and intubation of larynx as a means of relieving stridor in diphtheria.\(^3\)

Tracheostomy is preferred over intubation as the latter can cause ulceration in the trachea and increases the risk of pushing a piece of membrane further down into the airway.

If a patient can be treated with antitoxin serum and the first 24 to 36 hours can be tided over, the necessity for tracheostomy may be altogether avoided.

**Aim of study**

The aim of the study was to know the prevalence, age distribution, seasonal variation and trend of diphtheria cases presenting in ENT emergency, to emphasize the role of emergency tracheostomy in the management of laryngeal diphtheria.

**METHODS**

This was a retrospective study conducted in department of ENT, Maulana Azad Medical College from January 2015 to December 2018. The medical files were recovered from the emergency registration record and information was collected about the cases of diphtheria presenting with stridor to the emergency. The study included suspected, probable and confirmed cases of diphtheria as per the WHO definition guidelines with stridor or impending airway obstruction.\(^4\) The information collected included age, gender, socio-economic status and immunization status. The total number of emergency cases was also noted so as to estimate the load of diphtheria cases to ENT emergency. The month of presentation was also noted to establish the seasonal trend of the infection. The data collected was analysed using microsoft excel and presented in number and percentages.

**RESULTS**

**Case distribution**

Diphtheria accounted for 0.8% total number of cases presenting in ENT emergency.

| Table 1: Age distribution of diphtheria cases. |
|-----------------|---|---|---|---|---|
| Age group (in years) | Male | Female | Total | Male female ratio | Total (%) |
| 0-3 | 12 | 4 | 16 | 3:1 | 13.1 |
| 3-6 | 30 | 10 | 40 | 3:1 | 32.8 |
| 6-9 | 24 | 12 | 36 | 2:1 | 29.5 |
| 9-12 | 20 | 4 | 24 | 5:1 | 19.7 |
| >12 | 4 | 2 | 6 | 2:1 | 4.9 |
| Total | 90 | 32 | 122 | - | 100 |

**Age and sex distribution**

A total of 122 cases of diphtheria presented to ENT emergency from January 2015 to December 2018. Maximum number of cases were in age group of between 3 to 6 years (32.8%) followed by 6-9 age group (29.5%), 9-12 (19.7%), 0-3 (13.1%). The minimum number of cases was above 12 years of age. Male children predominated over female children with overall sex ratio of 3:1.

**Seasonal variation**

The maximum number of cases was reported in the month of August to November. (96 out of 122 cases). There were no cases of diphtheria during the month of March to June.
Table 2: Month wise distribution of diphtheria cases.

| Month    | Number of cases |
|----------|----------------|
| January  | 8              |
| February | 3              |
| March    | 0              |
| April    | 0              |
| May      | 0              |
| June     | 0              |
| July     | 3              |
| August   | 26             |
| September| 18             |
| October  | 24             |
| November | 28             |
| December | 12             |
| Total    | 122            |

Immunization status

Out of total 122 cases, reliable immunization history could be obtained from 112 patients. Most of the cases presenting to us were partially immunized (51.8%) or unimmunized (44.6%).

Table 3: Immunization status.

| Immunization status | N (% ) |
|---------------------|--------|
| Fully immunized     | 4 (3.6)|
| Partially immunized | 58 (51.8)|
| Unimmunized         | 50 (44.6)|
| Total               | 112 (100)|

Figure 1: A) Pseudo membrane in the oropharynx, B) airway secured with tracheostomy and C) pseudo membrane removed from tracheostome.

Treatment and outcome

Out of the total cases presented to us, 6 cases did not need any airway intervention and was sent for antitoxin administration to infectious disease hospital. Since the study was conducted at a tertiary care hospital most of the patients presented to us in the advanced stage of disease necessitating airway intervention in majority of them (106 out of 122 cases, 86.9%). Ten patients died before intervention was done due to late presentation. Emergency tracheostomy was successfully done in remaining patients and started on antitoxin treatment.

DISCUSSION

Diphtheria still continues to be a major public health problem in India. Our study presents a retrospective analysis of diphtheria cases presenting to the ENT emergency. In our institution, most of the cases presented with stridor and diagnosis was made on clinical grounds.

Diphtheria cases accounted to about 0.8% of the total emergency cases. The disease shows a seasonal variation with maximum cases occurring during month of August to November which was in concordance with several studies carried out at different places of our country over last 30 years.

In our study, maximum number of cases was in the age group of 3 to 9 years (62.3% of total cases). Male children predominated over female children with overall sex ratio of 3:1. Another study conducted in Delhi on epidemiological aspects of diphtheria reported that maximum number of cases was seen in the age group less than 9 years (93%) with a sex ratio of 1.6:1. Bhattacharya et al reported a case on outbreak of diphtheria where the mean age group was 10.23 years with male preponderance of 53.33% and case fatality rate of 28.6%. Our study reported a case fatality rate of 8.2%. Study conducted in Hyderabad during 2003 to 2006 also noted a highest incidence among children aged 5 to 19 years. Although previously considered to be a disease common to under five children, in recent years there has been a shift in the age group which can be attributed to widespread immunization programmes. But the persistence or resurgence of diphtheria in the country points to low coverage of primary immunization and booster doses.

Most of the cases that presented to us were either unimmunized or partially immunized. This indicates the unsatisfactory immunization coverage that still prevails in some parts of India. Various reasons attributed include sociocultural aspects like lack of awareness, frequent vaccine and antitoxin shortages, unnecessary contraindications to vaccination, religious beliefs and illiteracy.

It was observed in our study that 4 patients developed diphtheria inspite of completed DPT vaccination. Fortunately, disease in previously immunized children is milder and is considered to have better prognosis.

Tracheostomy is a lifesaving intervention in the management of diphtheria. Other indications of pediatric tracheostomy include foreign body in upper airway,
tetanus, subglottic stenosis, congenital anomaly, post traumatic brain injury and cases of anticipated prolonged mechanical ventilation. Unlike tracheostomy in adults, pediatric tracheostomy has a higher rate of morbidity and mortality. It requires great expertise and thorough knowledge of anatomy to reduce complications. Most important intraoperative complications include hemorrhage, injury to adjacent structures, and sudden apnea. Subcutaneous emphysema, pneumothorax, accidental decannulation and pneumonia are the common immediate postoperative complications. Delayed complications like subglottic stenosis, tracheoarterous fistula, tracheoarterial fistula and difficult decannulation are also seen associated with pediatric tracheostomy. In our follow up the most common delayed complication was difficult decannulation owing to tracheal granuloma and subglottic stenosis. Common sites of tracheal granulations were suprastomal granulations and peristomal granulations.

**Diphtheria** is a disease with high rate of morbidity and mortality. C. diphtheria is transmitted through aerosol route from close contact with an infected person; incubation period is 3-5 days for respiratory diphtheria. Toxin causes necrosis of epithelium leading to mucosal ulceration, hyperemia, and edema leading to fibrinosuppurative exudates which ultimately forms the pseudo membranes. Apart from causing mechanical airway obstruction, the exotoxin produced in the pseudo membranes reaches into bloodstream and gets into systemic circulation. High toxicity of diphtheria toxin, poor socioeconomic status and poor host response to diphtheria antitoxins are some of the important reasons for the same. The magnitude of disease as per our study is just a projection of a small section of the country. The cases presenting to us are mainly in its most severe form. It is just the tip of iceberg. The actual magnitude of the disease lies unknown, unrecognized and unreported. Lack of knowledge of disease and value of immunization are the fundamental issues that need to be addressed for the disease prevention. Further promotion of health education, immunization and strengthening of disease surveillance are required to address this problem.

**CONCLUSION**

Diphtheria still continues to be a major public health issue in India. Early recognition and treatment including securing airway is necessary to prevent morbidities and mortalities. Tracheostomy is a speedy and efficient lifesaving procedure in the management of laryngeal diphtheria. There is a need to strengthen the immunization strategies and disease surveillance to all sectors of our population.

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**Ethical approval:** The study was approved by the Institutional Ethics Committee

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