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An Unusual Cause of Dyspnea and Hoarseness: A Recurrent Respiratory Papillomatosis

Nefes Darlığı ve Ses Kısıklığının Ender bir Nedeni: Tekrarlayıcı Respiratuar Papillomatozis

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Abstract

Recurrent respiratory papillomatosis is a rare respiratory condition associated with human papillomavirus. It is characterized by exophytic, wart-like, squamous lesions within the respiratory tract. Presently described is case of a 24-year-old Caucasian male who presented with recurrent laryngotracheal papillomatosis. Laryngoscopy and excisional biopsy had been performed at approximately 1½ years of age. Biopsy specimen was determined to be squamous cell papilloma. The patient received interferon alpha-2a and inhaled interferon, but treatment was discontinued due to lack of regression or delay in reappearance of symptoms. Rigid bronchoscopy has been performed for recurrent papilloma excision approximately every 3 to 4 months for 3 years.

Key words: *Bronchoscopy, papilloma, papillomaviridae, trachea.*

Özet

Rekküren respiratuar papillomatozis Human Papilloma Virus'ün neden olduğu nadir bir hastalıktır. Hastalık solunum yollarında siğil benzeri squamoz lezyonlarla karakterizedir. Sunduğumuz olgu 24 yaşında kafkas ırkından bir erkek olup rekküren trakeobronşial papillomatozis saptanmıştır. Olguya 1,5 yaşında iken laringoskopi yapılmış ve lezyon eksize edilerek tanı konulmuştur. Biyopsi materyali squamoz hücreli papilloma olarak raporlanmıştır. Olgu daha önce interferon α-2a ve inhaler interferon tedavisi almış fakat semptomlarda duraklama veya gerileme olmaması nedeniyle sonlandırılmıştı. Hastaya 3 yıldır her 3 ile 4 ayda bir papilloma eksizyonu yapılmaktadır.

Anahtar Sözcükler: *Bronkoskopi, papilloma, papilloma virüs ailesi, trakea.*

Recurrent respiratory papillomatosis (RRP) is a rare respiratory condition associated with human papillomavirus (HPV) (1). It is characterized by exophytic, wart-like, squamous lesions within the respiratory tract (2). It frequently occurs in the larynx, and rarely, may be seen in lower airways of trachea, primary bronchi, and the lungs (1). Patients with RRP may develop bronchiectasis secondary to respiratory obstruction and infection. Malignant transformation rate of 2% to 3% within 10 years of diagnosis has been reported (3).

Described herein is a case report of a 24-year-old male patient who presented with recurrent laryngotracheal papillomatosis.

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CASE

A 24-year-old Caucasian male patient has been receiving treatment for 3 years for shortness of breath and hoarseness that had increased over time. His medical history revealed that problem began at fourth postpartum month when he developed hoarseness. Laryngoscopy was performed when he was about 1½ years of age. Papilloma was identified in the larynx and excised. However, complaints returned approximately 5 months later. He repeatedly underwent laryngoscopic excision until he reached 10 years of age, and no lesion distal to the vocal cord was identified in that period. The patient had no family history of papillomatosis or immunosuppression. He received treatment for pneumonia when he was 10 years old, and since then, tracheal lesions had developed in addition to those in the larynx. Following detection of tracheal papilloma, rigid bronchoscopy was performed approximately every 5 to 6 months to excise papillomas. Interferon alph-2a was used for a while and interval to excision of papillomas increased to 8 months; however, use of interferon alpha-2a was discontinued when time until excision required was reduced to just 6 months. Inhaled interferon was also used, but discontinued due to lack of any regression or delay in recurrence of symptoms. Physical examination indicated body mass index of 18, presence of hoarseness, and scoliosis with mild tendency to lean to left. During period before papilloma excisions he would develop mild stridor, which disappeared following excision. Other systemic examinations were normal except persistent rales at base of the left lung. The patient has been monitored at our outpatient clinic for 3 years, during which he has undergone papilloma excision at 3 to 4-month intervals. Chest radiography confirmed scoliosis with lean to the left (Figure 1). Chest tomography indicated deviation of the mediastinum to the left, prominent bronchiectasis involving posterobasal portion of the left lower lobe, and irregularity of the tracheal lumen in sections of the mediastinum (Figure 2). Blood cell count and routine biochemical tests were normal except for PO2 value of 72 mmHg in arterial blood gases sampled before bronchoscopy. Rigid bronchoscopy under general anesthesia revealed polypoid lesions resembling bunch of grapes, from level of vocal cord to the distal trachea, obliterating 90% of the lumen; after the carina, the lumen was patent. Debulking of polypoid lesions was performed with rigid tube. Cryotherapy and excision with biopsy forceps were also occasionally performed during the procedure. As lesions bled easily from beginning of the procedure merely when touched, short intubation and mechanical ventilation were used due to deep desaturation, and then excision was resumed. Complete patency was achieved in the larynx and trachea following the procedure (Figure 3). Biopsy specimen was determined to be squamous cell papilloma.

Written informed consent to publication was obtained from the patient.

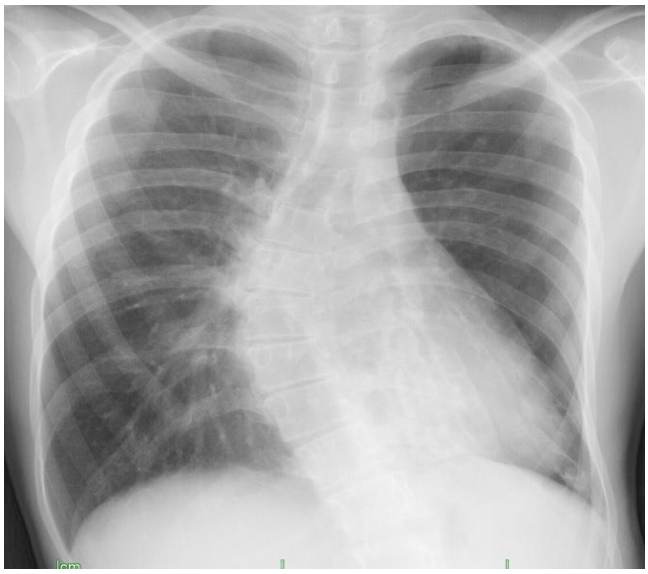

Figure 1: *Chest X-ray indicated scoliosis with a tendency to lean to the left*

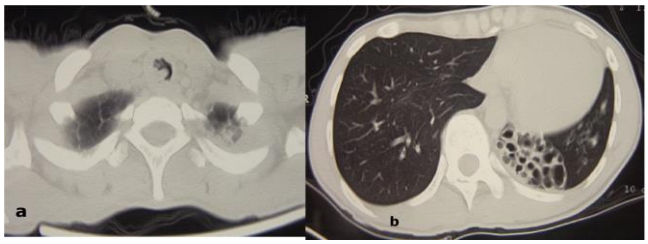

Figure 2: *Polypoid lesion in* ***(a)*** *trachea and* ***(b)*** *bronchiectasis in the left lower lobe are visible in chest tomography images*

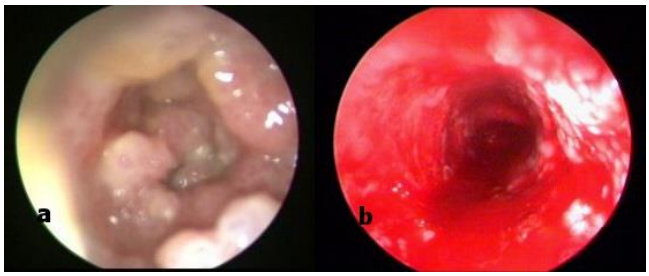

Figure 3: *Images taken* ***(a)*** *before and* ***(b)*** *after bronchoscopic excision*

DISCUSSION

RRP is a rare condition, most commonly associated with HPV types 6 and 11 (4). It equally affects women and men, and there is no difference in incidence by ethnicity. Although it is a benign condition, life-threatening respira-

tory problems or malignant transformation may develop (3).

RRP is the most common benign laryngeal lesion of childhood. Incidence has been reported as 4.3 per 100,000 children, and 1.8 per 100,000 adults per 100,000 in the USA (5). In pediatric series, RRP was typically diagnosed at between 2 and 4 years of age, with time to diagnosis of approximately 1 year (6). In our patient, symptoms started at fourth month of life; however, period between first diagnosis and onset of symptoms was approximately 1 year, in accordance with the literature.

The disease frequently involves the vocal cord area of the larynx, and rarely, there is involvement of the distal respiratory tract (1). Entire aerodigestive tract may also be involved (4). Spontaneous remission may be experienced, or repeated surgical excision may be required (4). Direct transmission of HPV may occur via the birth canal during intrauterine period or during labor (7). Childhood RRP is potentially more threatening due to rapid growth in childhood (8).

Clinically, hoarseness, chronic cough, recurrent pneumonia, retarded development in children, dysphagia, or stridor may occur (8). Our patient had no symptoms other than hoarseness and stridor. In this case, hoarseness remains following excision; however, stridor disappears after bronchoscopic excision.

HPV types 6 and 11 are the most common viral subtypes, the latter being more aggressive (8). In our patient, we didn't attempt to identify the viral agent and conduct typology; diagnosis of RRP was based on clinical, macroscopic, and microscopic evaluations.

As in present case, patients may require repeated bronchoscopic interventions in order to achieve and maintain airway patency. Although repeated surgical procedures may result in various complications, including synechia and narrowing in the laryngeal region, tracheoesophageal fistula, and stricture, no other effective treatment method has yet been developed to maintain airway patency. There is no report of specific complication during endobronchial treatment for RRP (9). However, as reported in previous publications, care should be taken for possible complications, such as hypoxia, bleeding, and arrhythmia associated with general anesthesia and rigid bronchoscopy (10,11). Although interferon alpha-2a, inhaled interferon, acyclovir, and cidofovir have been used, there is still no medical treatment that eliminates need for surgical treatment.

In conclusion, RRP is a rare disease that may involve the entire respiratory system. It is particularly seen in children between 2 and 4 years of age, and in adults between 20 and 40 years of age. Although it is a benign disease, it requires close monitoring due to severely restricted airway and potential malignant transformation. Utmost care should be taken during bronchoscopy due to potential bleeding.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - M.A.Ö., E.Ç., M.Ç., D.Ö.O.; Planning and Design - M.A.Ö., E.Ç., M.Ç., D.Ö.O.; Supervision - M.A.Ö., E.Ç., M.Ç., D.Ö.O.; Funding - M.A.Ö., E.Ç., M.Ç., D.Ö.O.; Materials - M.A.Ö., E.Ç., M.Ç., D.Ö.O.; Data Collection and/or Processing - M.A.Ö., E.Ç., M.Ç., D.Ö.O.; Analysis and/or Interpretation - M.A.Ö., E.Ç., M.Ç., D.Ö.O.; Literature Review - M.A.Ö., E.Ç., M.Ç., D.Ö.O.; Writing - M.A.Ö., E.Ç., M.Ç., D.Ö.O.; Critical Review - M.A.Ö., E.Ç., M.Ç., D.Ö.O.

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