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Case report

A case of adult congenital laryngeal cleft asymptomatic until hypopharynx cancer treatment

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ABSTRACT

Laryngeal cleft is an anomaly of failed posterior closure of the larynx. Most cases are diagnosed and need treatment early in life due to respiratory and swallowing problems. We report an unusual case of a 66-year-old man with an asymptomatic laryngeal cleft until treatment for hypopharyngeal cancer. During concurrent chemoradiotherapy (CCRT), despite reduced tumor volume, he presented severe dysphagia and dyspnea, followed by severe pneumonia twice. Because CCRT had to be discontinued, a pharyngolaryngectomy was performed for the cancer treatment. The resected specimen showed total removal of the tumor and a total longitudinal cleft of the cricoid cartilage, classified as a type III laryngeal cleft by the Benjamin and Inglis' classification. A review of computed tomography images indicated that the redundant mucosa from bilateral edges closed the separation of the posterior cricoid cartilage and narrowed the laryngeal airway during CCRT. Adult presentations of laryngeal cleft are quite rare with only ten reported cases in English literature; the present case is of the oldest patient. Undiagnosed cases with laryngeal cleft may exist asymptomatically or without severe symptoms. The awareness of this condition may increase its diagnosis as a cause of diseases such as aspiration and recurrent pneumonia even in adult patients.

1. Introduction

Laryngeal cleft is a rare anomaly of failed posterior closure of the larynx and extends to the membranous portion of the trachea in some cases; the latter condition is precisely termed as a laryngo-tracheo-esophageal cleft. The interruption of cricoid fusion in utero results in an abnormal communication between the laryngo-tracheal respiratory and pharyngo-esophageal digestive tracts, with an estimated incidence of 1 in 10,000–20,000 living births, constituting 0.2%–1.5% of the congenital malformations of the larynx [1]. Presenting signs and symptoms include stridor, hoarseness, dysphagia, cough, cyanosis during feeding, aspiration, and recurrent pneumonia [1,2]. Symptom severity varies with the length of the defect, and the most commonly accepted classification is the one proposed by Benjamin and Inglis [3] and later modified by Sandu and Monnier [4]. A type I cleft is located above the level of the vocal folds, a type II cleft extends into the cricoid cartilage, a type IIIa cleft extends entirely through the cricoid cartilage but not into the trachea, a type IIIb cleft extends to the cervical trachea, and a type IV cleft extends beyond the thoracic inlet. A laryngeal cleft is often associated with other congenital anomalies (16%–68%) of the digestive tract, genitourinary tract,

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cardiovascular system, craniofacial, and tracheo-bronchopulmonary regions. These associated malformations may appear as isolated or syndromic defects [1].

Because the laryngeal cleft would mostly be diagnosed in the neonatal or infantile stage, adult presentations of this congenital deficit are extremely rare, with only ten cases previously reported in the English literature [5–12]. Here, we report a case of a laryngeal cleft that remained asymptomatic until treatment for hypopharyngeal cancer. This case is unique because it presents a report of the oldest patient diagnosed with a laryngeal cleft.

2. Case report

A 66-year-old man visited our department with complaints of dysphagia. The patient had a mass lesion located from the left piriform sinus to the lateral wall of the oropharynx that obstructed the epiglottal movement while swallowing. Hoarseness and dyspnea were not reported or apparent at initial examination. The patient had no past history of pneumonia, asthma or laryngopharyngeal reflex disease. Histological examination detected moderately differentiated squamous cell carcinoma. Supported by evidence from imaging studies, hypopharyngeal cancer of cT4aN2bM0 (UICC 7th ed.) was diagnosed. Because induction chemotherapy (docetaxel/cisplatin/5-fluorouracil) showed a favorable partial response, concurrent chemoradiotherapy (CCRT) with cisplatin and 70 Gy (35 fractions) of intensity-modulated radiotherapy was planned. When approximately half of the planned CCRT dose (32 Gy) was delivered and despite substantial tumor volume reduction, the patient presented with severe laryngeal edema, dyspnea, and dysphagia. A tracheostomy was performed, but the patient had severe pneumonia twice. The insertion of an endoscope for examination or even a nasogastric tube for feeding encountered difficulty in passing through the hypopharynx because of a tendency to enter the laryngo-tracheal lumen. The CCRT had to be discontinued, and a pharyngolaryngectomy, left neck dissection, and reconstruction with a free jejunal graft were subsequently performed.

On examination of the resected specimen, total tumor removal was pathologically proved; additionally, a total longitudinal cleft of the cricoid cartilage was observed (Fig. 1). A review of the computed tomography (CT) images indicated that the redundant mucosa from the bilateral edges had closed the separation of the posterior cricoid cartilage since the initial examination (Fig. 2A), and that swollen redundant mucosa had caused laryngeal airway narrowing during CCRT (Fig. 2B). Postoperatively, the patient recovered well and did not have any signs of recurrence or problems with respiration or swallowing at the 1-year follow-up.

3. Discussion

Adult presentations of laryngeal cleft are quite rare, but ten cases have been previously reported. In 1976, Montgomery and Smith [5] reported a male patient who became symptomatic at the age of 42 years and underwent surgery at the age of 48 years due to gradual progression of dysphagia, dyspnea, and hoarseness. Haskins et al. [6] reported a male patient who underwent surgery at the age of 24 years and had a life-long history of recurrent pneumonia. Lancaster et al. [7] reported the case of a 50-year-old woman with a 3-week history of worsening inspiratory stridor associated with increasing shortness of breath and simultaneous worsening of heart failure. Thornton et al. [8] reported the case of a 19-year-old male patient with type I laryngeal cleft and a history of recurrent pneumonia and gastro-esophageal reflux disease. Birkent et al. [9] reported the case of a 20-year-old man with posterior laryngeal cleft and nonfusion of the thyroid alae, with complains of only dysphonia. Weissbrod et al. [10] reported three cases: a 60-year-old man with liquid aspiration since infancy; a 48-year-old woman with a long history of progressive shortness of breath, dysphagia, cough, and hoarseness; and a 29-year-old woman with a 1-month history

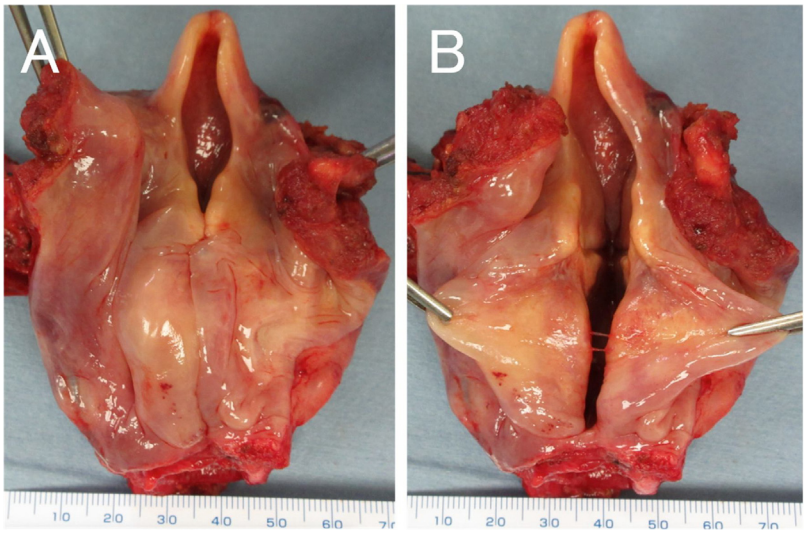

Fig. 1. The posterior view of the resected specimen of pharygolaryngectomy. (A) The posterior wall of hypopharynx was already cut and opened. The swollen mucosa covering the posterior larynx was identified, but it was difficult to detect the laryngeal cleft at a glance. (B) Pulling the redundant mucosa laterally clearly exposed the cleft of the cricoid cartilage. Note that the cleft extended from the arytenoid throughout the entire cricoid cartilage but not into the trachea.

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