

Scleroma of the Larynx - A Case Report -

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Scleroma of the larynx is a rare specific granulomatous disease and is usually presented as a laryngeal extension of a primary rhinoscleroma. We report an unusual case of laryngeal scleroma in a 64-year-old female patient whose presented with progressive upper respiratory obstructive symptoms lasted for 2 years. Symptoms were acutely exaggerated during the course of one month. On physical examination, the nasal mucosa was atrophic and crusted, and septal perforation was noted. The fiberoptic laryngoscopy revealed a yellowish granulated epiglottic mass with a dirty surface mimicking laryngeal cancer. Histological sectioning showed the characteristic features of scleroma including Mikulicz cells with scattered plasma cells and some neutrophils. Warthin-starry stains and electron microscopy examination showed numerous short rods inside the cytoplasm of the Mikulicz cells. After the diagnosis, the patient underwent an emergency tracheostomy followed by long-term antibiotic treatment, and her symptoms have slowly improved.

Key Words : Larynx-Scleroma-Mikulicz Cell-Warthin-Starry Stain

INTRODUCTION

Rhinoscleroma is an infectious disease of the upper respiratory tract that has a chronic and slow progressive clinical course. The disease was first demonstrated by Von Hebra,¹ and its histologic features were described by Mikulicz.² It is caused by the bacterium *Klebsiella rhinoscleromatis* and is endemic to eastern Europe, northern Africa, southern Asia, and central America.³ Only one case of non-endemic rhinoscleroma has been reported in Korea.⁴

The disease usually involves the nasal cavity as a rhinoscleroma, but occasionally it gradually progresses into the larynx and the trachea, which causes airway obstruction symptoms.^{5,6} Because of the slow progression, a prompt diagnosis is usually delayed. Although the airway obstruction may be mild at the initial stages of the disease, it can be life threatening unless proper intervention is done. We describe a sporadic case of scleroma with severe airway obstruction involving the larynx. It led to a tracheotomy.

CASE REPORT

A 64-year-old female patient with a 2-year history of minor dyspnea which was admitted to our hospital. Her ailment had been exaggerated during the one month prior to her admission. There were no signs of sore throat, odynophagia or voice change. On physical examination, the nasal mucosa was atrophic and crusted, and septal perforation was noted. There was a small 0.5 × 0.5 cm-sized polypoid mass just behind the left posterior pillar and a mass-like lesion with a dirty surface in the epiglottis.

CT scans of the neck showed a focal nodular mass of the epiglottis with a contour deformity and slight thickening of the tracheal wall. Fiberoptic bronchoscopy revealed an epiglottic mass, and the trachea was almost obliterated by a fibrotic lesion 3 cm below the vocal cord (Fig. 1A). The initial clinical impression of the patient was supraglottic cancer. Then, the biopsy for the epiglottic lesion was performed.

The epiglottic biopsy specimen was fixed in formalin and stain-

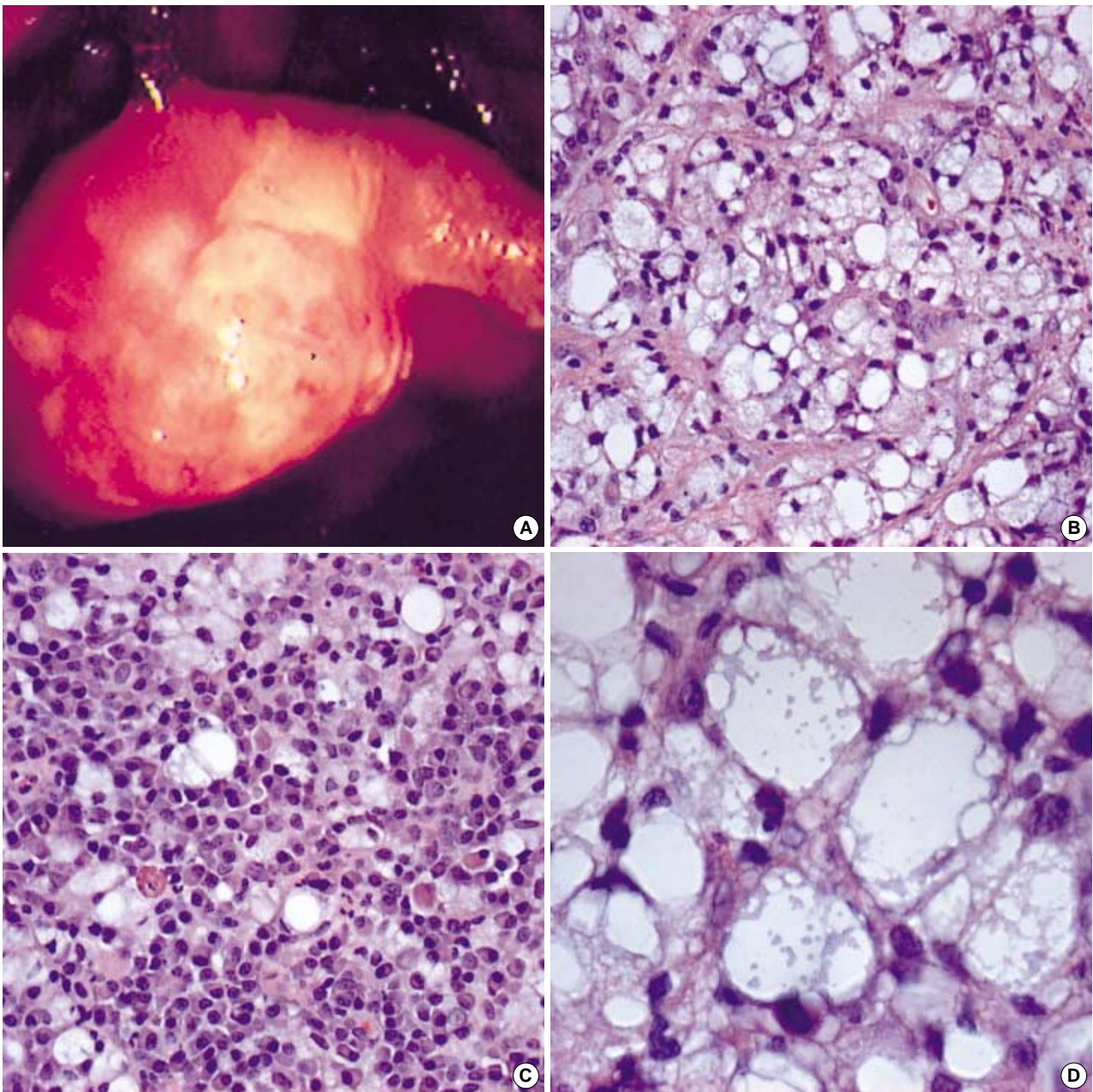


Fig. 1. (A) The bronchoscopic view of the larynx shows an epiglottic nodular mass with dirty yellowish surface. (B) Microscopically the dense infiltration of foamy macrophages (Mikulicz cells) are accompanied by some neutrophils. (C) Some scattered macrophages are admixed with plasma cells with Russell bodies. (D) The foamy macrophages show cytoplasmic bacilli consistent with *Klebsiella rhinoscleromatis*.

ed with hematoxylin and eosin. The section showed a diffuse infiltration of foamy histiocytes (Mikulicz cells) containing bacillary organisms which had a slimy mucopolysaccharide coating (Fig. 1B&D), scattered plasma cells with Russell bodies, and some neutrophils (Fig. 1C). Special stainings for acid fast bacilli, Gram, D-PAS, Gomori-Methenamine Silver (CMS), and Warthin-starry were performed to identify the organism. Remarkably, only the organisms stained by the GMS and Warthin-starry stains led to

the histologic diagnosis of laryngeal scleroma. Warthin-starry stains showed numerous short rods inside the cytoplasm of the Mikulicz cells (Fig. 2A). On electron microscopic examination, irregularly shaped Mikulicz cells with oval vacuolation of the cytoplasm were noted. The organelles in the cytoplasm were located in the intervesicular bridges and presented a variety of alternations according to the extent of the vacuolization. There were numerous small bacillary rods with their slimy mucopolysaccharide coatings inside

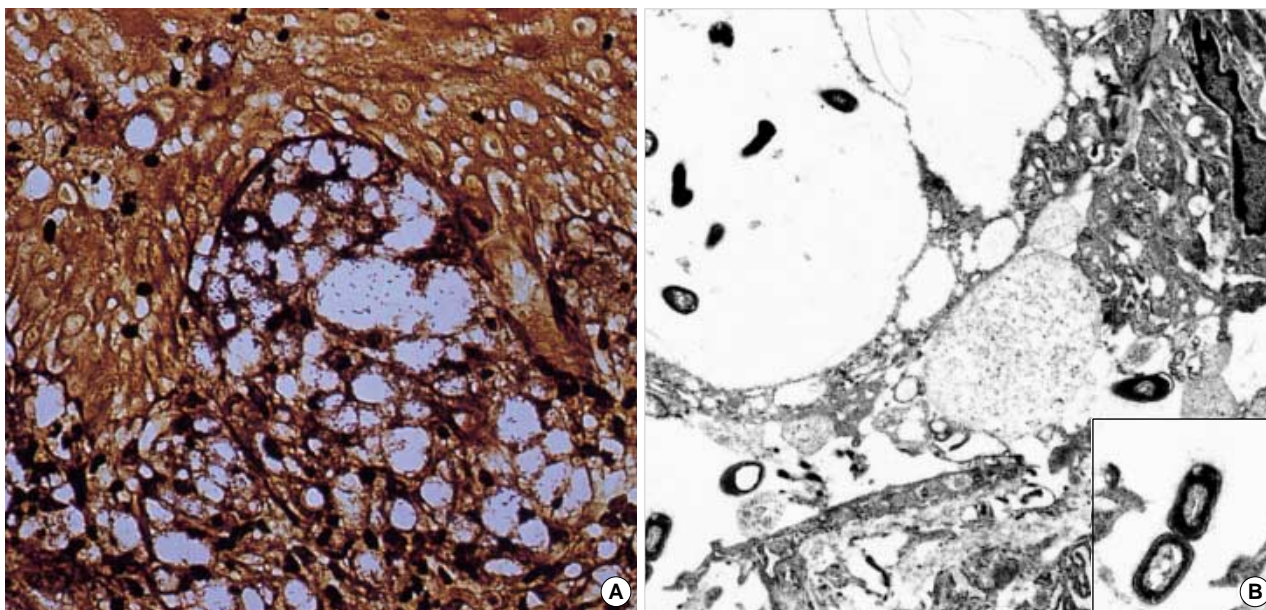


Fig. 2. (A) The Warthin-Starry stain displays numerous bacilli in the cytoplasm. (B) The electron micrograph of the Mikulicz cells shows several vacuoles and *Klebsiella rhinoscleromatis*. Inset, Note its slimy mucopolysaccharide coating of the organism.

the vacuoles (Fig. 2B).

The patient's obstructive symptoms got worse, which led to a tracheotomy. She was transferred to another medical facility for tracheal stent insertion, but the procedure was eventually not performed. Her condition has improved under long-term antibiotic treatment.

DISCUSSION

We describe a 64-year-old female patient with a laryngeal scleroma. The diagnosis was made from the characteristic histologic features on a laryngeal biopsy. As far as we know, this is the second Korean case of scleroma, which follows the previously reported rhinoscleroma.⁴ Moreover, this is the first experience of a laryngeal manifestation of a scleroma mimicking laryngeal cancer. The long-standing history of upper respiratory tract obstruction was consistent with the diagnosis.

Klebsiella rhinoscleromatis is an opportunistic, gram-negative diplobacillus of the family Enterobacteriaceae.⁷ Prompt treatment with antimicrobial agents can reduce the obstructive symptoms and prevent a relapse, but a high prevalence of resistant strains are usually the major problem in progressive, non-remissive cases. Such cases can lead to potentially life-threatening obstructions.⁸

Rhinoscleroma is usually encountered in areas such as Central America, Eastern Europe, southern Asia and northern Africa; but

sporadic cases have been reported.³ Laryngotracheal scleroma is relatively uncommon and is usually a secondary extension of rhinoscleroma. Primary laryngeal scleroma is extremely rare.⁹ As the intranasal mucosa was atrophic with crust formation in the present case, the larynx seemed to be involved secondarily. The incidence of laryngeal lesions in the scleroma varies in different series, ranging from 12 to 40 percent.¹⁰ The lesion occurs most often in the subglottic region where the character of the mucosa changes from squamous epithelium to columnar epithelium.¹⁰ There have been no reported cases of primary laryngeal scleroma in Korea. Malignant transformation of rhinoscleroma is also very rare and only a few cases have been reported.¹¹

Rhinoscleroma usually undergoes three distinctive clinical stages.¹² The exudation stage or the ozena phase, as it is termed, is mainly characterized by a purulent discharge and mucosal congestion. The proliferation stage has granulomatous inflammation and nodule formation, and the cicatricial stage has extensive scarring. These three stages usually overlap, and an atrophic phase can be seen between the first and the second stages.

There are numerous methods of diagnosing scleroma, such as the examination of a culture from the affected area, histopathologic study with special stains from the biopsy specimen, and serologic and immunochemical studies. Culture studies are diagnostic, but the limitation is that only 60% of the biopsy proven cases were positive for *K. rhinoscleromatis*.¹³ Complement fixation tests and agglutination tests can be used, but they are diagnostic only when

the tests show positive results. Histopathologic determination of the scleroma is by far the most accurate and the most widely used method of diagnosis. The presence of Mikulicz cells, Russell bodies, plasma cells, lymphocytes, and gram-negative bacilli showing slimy mucopolysaccharide coating are not pathognomonic, but characteristic of the scleroma.

In summary, we report an unusual case of sporadic laryngeal scleroma in a 64-year-old female patient. She presented a long-standing history of upper airway obstruction. The laryngoscopic findings indicated laryngeal cancer. The laryngeal biopsy showed characteristic features of Mikulicz cells containing bacilli that resembled *Klebsiella rhinoscleromatis*. This is the second Korean case of scleroma and the first experience of laryngeal manifestation. Further distinctions characterized by other specific and nonspecific granulomatous inflammations are necessary.

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