

# **A Saddle Nose with Acute Respiratory Failure**

—\_Medical Illustration —

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## Case Report

A 17-year-old youth came to the emergency department (ED) with stridor in March, 2009. Commencing six months prior, he had developed bilateral progressive hearing loss along with frequent vertigo. Both auricles appeared erythematous and swollen over the prior 2 months. Intermittent dyspnea with episodic stridor and easy choking had developed. The neck soft tissue X-ray study showed a narrowing of the airway (Fig 1). The nasal fiberoptic examination revealed bilateral vocal paralysis with edematous swelling of the arytenoids and false vocal cords. Left external ear destruction (Fig 2A), right ear induration (Fig 2B), and a saddle nose deformity (Fig 2C) were noted. Because of the acute respiratory stress and stridor, the patient was intubated immediately treated with endotracheal intubation at ED. Due to soft tissue swelling, the intubation could only be performed with 6.5 mm endotracheal tube. A computed tomography (CT scan) of the neck showed circumferential thickening of the tracheal wall along with tracheal lumen narrowing of the subglottic trachea (Fig 3). The patient then underwent tracheostomy, and was successfully weaned from mechanical ventilation. The biopsy of the trachea showed acute and chronic inflammatory infiltrates surrounding the elastic cartilage with foci of perichondrial erosion and chondrocyte destruction. Biopsy of the ear revealed acutely inflamed granulation tissue with patchy organizing microhemorrhage.

A diagnosis of relapsing polychondritis was established by the combination of clinical typical findings (McAdam's criteria)<sup>1</sup>, imaging studies, and biopsies of the trachea and ear.

Saddle nose is a condition associated with congenital syphilis, Wegener's granulomatosis, cocaine abuse, direct trauma, and relapsing polychondritis. Direct trauma and cocaine abuse can be quickly rule out by history taking. Late congenital syphilis is associated with multiple dental and bony abnormalities and sometimes associated with mulberry molars saber shins, Hutchinson`s incisors, and Clutton`s joint. Wegener's granulomatosis not only has renal and pulmonary involvement but also have central or peripheral neurological damage.<sup>5</sup> Wegener's granulomatosis does not involve the external ear.

Relapsing polychondritis is a rare multisystem inflammatory process that may potentially compromise the structure and function of all bodies' cartilage, especially the ears, nose, eyes, joints, and respiratory tract. Auricular chondritis is the most common initial presentation<sup>2</sup>, and the prevalence of airway involvement is about 21%. Large respiratory tract involvement can result in life-threatening situations.<sup>3</sup> Tracheal collapse is reported to be from 14% to 42%<sup>4</sup>. It is very easy for these patients to complete close off the airway, and they may require emergency cricothyrotomy. Moreover, if the stenosis is distal to the cricoid cartilage, they may still die from

respiratory failure, as the cricothyrotomy has not relieved the obstruction.

We report a case of relapsing polychondritis initially presenting with hearing impairment, along with chondritis of the nose, an auricle, and the development of acute respiratory failure due to involvement of the laryngeal cartilage.

The patient was successfully weaned from mechanical ventilation after tracheostomy.

## Reference

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### **Figure legends**

**Figure 1.** Cervical soft tissue x-ray showing tracheal thickening with a resulting subglottic stenosis

**Figure 2.** Left external ear destruction (A), right ear induration (B) and a saddle nose deformity (C).

**Figure 3.** Computed tomography showed circumferential tracheal wall thickening and tracheal lumen narrowing of the subglottic trachea.