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Nebulized racemic ephedrine in the treatment of acute exacerbations of laryngeal relapsing polychondritis

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Abstract
A case of relapsing polychondritis of the larynx and upper trachea is presented. We report the use of nebulized racemic ephedrine, as a new treatment modality of the life threatening airway oedema which characterizes acute exacerbations of this disease.

Introduction

Relapsing polychondritis was first described in 1923 by Faksch-Wartenhorst. Since then 114 cases have appeared in the literature (Hughes *et al.*, 1972). It is a disease of unknown aetiology which presents as recurrent inflammation of cartilage, especially of the auricle, nose, larynx and trachea. Arthropathy and conjunctivitis are often present. Histologically there is chondritis with subsequent degeneration of cartilage, the end stages of the disease being characterized by marked fibrosis (Valenzuela *et al.*, 1980; van den Broek, 1987). These changes result in airway narrowing and eventually airway collapse.

Symptoms of laryngeal involvement are hoarseness, dyspnoea and stridor. These arise during acute exacerbations of the condition and are due to mucosal oedema. In the early stages of the disease the patient is asymptomatic between acute attacks. Progressive laryngeal fibrosis, leading to stenosis occurs in all cases, though the speed of progression varies greatly from patient to patient. Death occurs from bronchitis and airway collapse, (Dolan *et al.*, 1966).

Case report

A 65-year-old woman was urgently referred by her general practitioner to our casualty department because of increasing stridor. On arrival she was able to give a history of gradual onset of hoarseness during the previous week associated with increasing difficulty in breathing during the preceding 24 hours.

The past history revealed that she had two thyroid adenomata excised previously. In July 1987, four years after the last thyroid operation, she suffered her first attack of acute subglottic oedema. During the first few attacks she spent two to three days in the intensive care unit being treated with antibiotics and corticosteroids. She was discharged on a maintenance dose of 5 mg of prednisone daily.

At presentation she was pale, with a respiratory rate of 22 per minute, and no evidence of cyanosis. There was no stridor at rest but it became quite marked on minimal exertion. Indirect laryngoscopy demonstrated subglottic oedema resulting in an approximately 40 per cent narrowing of the airway. The remainder of her physical examination was normal as were routine investigations, including arterial blood gas evalution. She was admitted to the intensive care unit, and was treated with nebulized racemic ephedrine (0.5 ml of ephedrine in 0.5 ml normal saline given four hourly) and intravenous corticosteroids.

She made a rapid recovery and the following day was fit for transfer to the ward.

Subsequent exacerbations were treated by increasing the steroid dosage and in addition giving nebulized racemic ephedrine. Within minutes of starting the nebulized ephedrine the patient reported a marked improvement in her breathing difficulty. On each occasion the oedema was observed to be greatly reduced over the first two hours of starting the nebulizer, with a corresponding improvement in stridor. This rapid improvement allowed the patient to be transferred from the ICU to the ward the following day.

Investigations of possible causes had been carried out; these included angioneurotic oedema, autoimmune thyroid disease, disorders of calcium metabolism and parasitic infestation of the gastrointestinal tract.

Because of repeated attacks of subglottic oedema, and the development of persistent stridor at rest, a tracheostomy was performed. At operation extensive fibrosis was noted in relation to the cricoid and upper tracheal cartilages with gross mucosal oedema in the trachea. Histologically the cartilage demonstrated early degeneration due to an inflammatory reaction (Fig. 1). The patient made a full recovery and has remained well on a maintenance dose of prednisone 5 mg/daily.

Discussion

Reviewing 52 cases of relapsing polychondritis Dolan *et al.* (1966) and Hughes *et al.* (1972) found that acute exacerbations were most often treated by increasing the patient's maintenance dose of corticosteroids. This proved to be effective in the non-life threatening cases e.g. auricular and nasal disease. In cases of laryngeal and tracheal relapsing polychondritis, however, the delay in onset of action of steroids means that during acute exacerbations life threatening airway obstruction is a particular hazard. It was in an attempt to reduce this danger that we chose to use nebulized racemic ephedrine.

Racemic ephedrine is a synthetic form of ephedrine. It is optically inactive being a mixture of equal parts of the dextro and laevorotatory isomers, and hence the name 'racemic'. It is a very powerful vasoconstrictor which acts immediately and has a duration of action of four hours.

We found that racemic ephedrine, with its immediate onset

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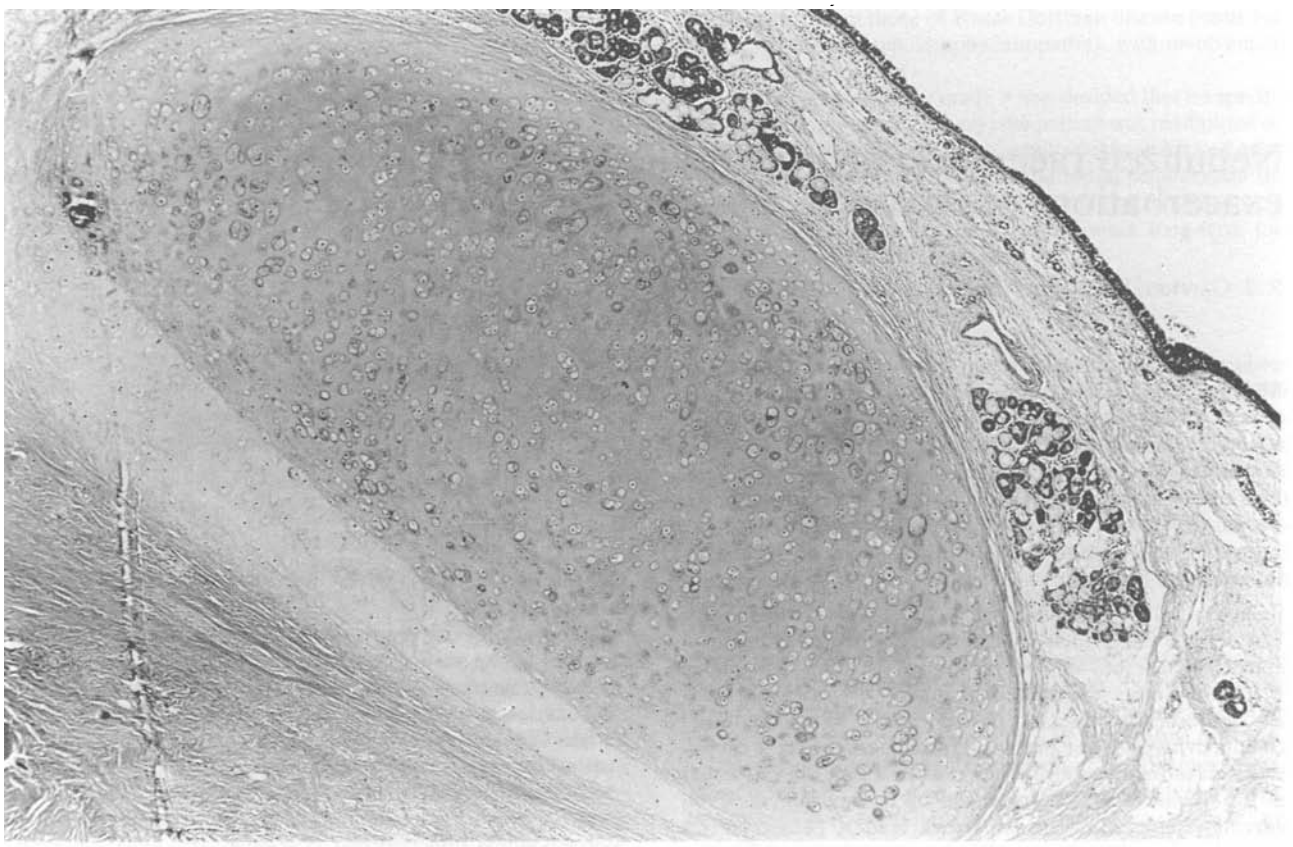

FIG. 1
Inflammatory cell infiltration of tracheal cartilage in polychondritis. H&E stain. Magnification ×40.

of action, was very effective in the treatment of the acute exacerbations of the disease, and provided good relief of airway obstruction while awaiting the onset of action of steroids.

Conclusion

Relapsing polychondritis of the larynx is characterized by acute attacks of laryngeal oedema which can be life threatening. Nebulized racemic ephedrine has proven to be very effective in the treatment of these acute attacks.

We advocate that it should be used while awaiting the onset of action of steroids during acute exacerbations of this disease.

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