CASE REPORT

Life-threatening stridor presenting in a patient with rheumatoid involvement of the larynx

D. F. McGEEHAN, J. N. CRINNION & D. R. STRACHAN

Accident and Emergency Department, Leeds General Infirmary, Leeds, England

SUMMARY

A case of a female patient with extensive rheumatoid arthritis who presented to the Accident and Emergency Department with life-threatening stridor is described. Although clinical involvement of the larynx is found in over a third of patients with severe rheumatoid arthritis, acute airways obstruction is fortunately a very rare complication. Stridor is probably precipitated in the acute situation in such patients as a result of upper respiratory tract infection.

INTRODUCTION

Arthritis of the crico-arytenoid joints is most commonly seen in rheumatoid arthritis, but it can also be found in other arthropathies including Reiter's syndrome, systemic lupus erythematosus, progressive systemic sclerosis, Tietze's syndrome and relapsing systemic sclerosis. It was clearly shown by Lofgren *et al.* (1962) that clinical involvement of the crico-arytenoid joints is found in 25-35% of patients with severe rheumatoid arthritis. At necropsy it could be demonstrated that there was even greater involvement of the larynx but the incidence of acute airway obstruction is extremely rare. With the increasing arthritic progression in the larynx the vocal cords tend to assume a fixed and adducted position. The laryngeal symptoms tend to run a long and very protracted course and may be present for several years. The symptoms usually result in a hoarsening and weakening of the voice (Wojtuleski *et al.*, 1973). Chronic upper airways obstruction can produce hypoxaemia, hypercapnoea and respiratory acidosis leading to pulmonary hypertension and Cor pulmonale.

CASE REPORT

A 62-year-old lady presented to the Accident and Emergency Department of Leeds General Infirmary on the afternoon of the 17th October 1988 with an onset of severe stridor of less than 1 h in duration. There was an 8 year history of generalized sero-positive rheumatoid arthritis involving all limbs and the cervical spine. More recently there was a 2 month history of increasing hoarseness of the voice but no dyspnoea. On admission to the resuscitation unit she was markedly distressed with central cyanosis and audible stridor. She then became increasingly more dyspnoeic over the next few minutes with flaring of the alar nasa, retraction of the intercostal muscles and a tracheal tug. The peak expiratory flow rate was unrecordable and the estimation of the blood gases on room air showed a pH of 7.26, PCO_2 of 6.4 KPa and a PO_2 of 7.6 KPa. She was started on supplemental oxygen and the anaesthetic and ear, nose and throat registrars were asked to urgently come to the unit. In the meantime the possibility of cricothyroidotomy was being considered. With the anaesthetist present in the unit, the ENT registrar passed a flexible fibre-optic laryngoscope and this showed the vocal cords to be fixed in a mid-line position and no movement could be elicited. She was then transferred to the main operating theatre and cervical tracheotomy was performed and after this she made an uncomplicated recovery. Due to the extensive rheumatoid involvement of her cervical spine, she was deemed unfit for more definitive laryngeal surgery. There was however no radiological evidence of atlanto-axial instability on the views taken of the cervical spine. She was discharged from hospital 3 weeks later with a permanent tracheotomy.

DISCUSSION

Although the incidence of clinical involvement of the crico-arytenoids has been given as over 30%, these series have almost certainly been acquired from specialized hospital clinics and probably represent patients with a more severe type of disease. The relatively uncommon incidence of acute upper airways obstruction may well reflect the sedate activity and poor locomotor function in these groups of patients. The presentation of acute stridor could well be an end-stage of a long-term pathological involvement of the larynx that has been aggravated by a respiratory tract infection. In our case presentation there was no radiological evidence of vertical atlanto-axial subluxation which makes the possibility of brain-stem involvement contributing to laryngeal stridor unlikely. However in patients with evidence of brain-stem compression by the odontoid peg, then this may have to be considered as a factor in the aetiology of acute airways obstruction.

The management of acute airways obstruction in these patients can present marked technical problems due to the involvement of the cervical spine and the temporo-mandibular joints. This makes rigid instrumentation of the larynx extremely difficult. Any attempts to extend the neck can produce fracture and dislocation of the spine and resultant neurological sequelae. The preferred method of viewing the vocal cords is by use of the fibre-optic laryngoscope and endotracheal intubation can be achieved by use

of the fibre-optic bronchoscope. However such instrumentation is extremely hazardous and can provoke complete airway obstruction and should only be undertaken in optimum circumstances with the support of ENT or anaesthetic colleagues. One should be prepared to perform an emergency cervical tracheotomy or crico-thyroidotomy in the accident unit if there is any compromise of the upper airway, but fortunately this is a rare occurrence.

REFERENCES

- Libby D. M., Shain Schley W. & Smith J. P. (1981) Crico-arytenoid arthritis in ankylosing spondylitis. *Chest* **80**, 641–3.
- Lofgren R. H., Montgomery W. W. *et al.* (1962) The incidence of laryngeal involvement in rheumatoid arthritis. *New England Journal of Medicine* **267**, 193–5.
- Wojtuleski J. A., Sturrock R. D., Branfoot A. C. *et al.* (1973) Crico-arytenoid arthritis in ankylosing spondylitis. *British Medical Journal* **3**, 145–6.