Thorax (1966), **21**, 355.

Scleroma of the trachea associated with *Pseudomonas pyocyanea*

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Scleroma is a chronic granulomatous condition affecting the submucosa and mucosa of the upper respiratory tract, particularly the nose and larynx, and less often the trachea and bronchi. In the fully developed state the histological appearances are diagnostic, with plasma cells, lymphocytes, larger more deeply staining cells or Russell bodies, and large vacuolated cells of Mikulicz containing intracellular bacilli (Ellis, 1952). The disease is usually associated with a particular organism, the Frisch bacillus, which is a member of the Friedländer or Klebsiella group (Morgan, 1958). Ellis (1952) stated that there was good evidence that this bacillus was the cause of the disease and that a complement fixation reaction was diagnostic. Goldzieher (1965) claimed that the complement fixation reaction became negative when the patient was adequately treated. The alternative view is that infection has not been proved to be the cause and that there is a possible symbiotic role of these organisms in the disease (Editorial, 1964). Fisher and Dimling (1964) reported a case in which the clinical and histological appearances of scleroma were associated with a negative culture for Klebsiella but a positive culture for *Proteus mirabilis,* which was grown from nasal secretions and from biopsy material. The electron microscope appearances of the bacteria in the histological section appeared to resemble Proteus more than Klebsiella.

In the case reported here, the typical clinical and histological appearances of scleroma of the trachea were associated with repeated positive cultures of Pseudomonas but no evidence of Klebsiella.

CASE REPORT

The patient, a 35-year-old housewife, had been born in Poland but had lived in Australia for 15 years. Her presenting complaint was of non-productive cough and husky voice for 18 months. Her nose had been dry and crusted for as long as she could remember. She was otherwise well. The nose was considered to show an atrophic rhinitis, but there was no thickening or obstruction of the nasal airway. The vocal cords appeared to be more rigid than usual and on bronchoscopy there was narrowing and irregularity of the upper third of the trachea. Physical examination was otherwise normal. A tracheogram showed that the upper 4 in. (100 mm.) of the trachea was narrowed, and in the lower portion of this segment the narrowing was more marked and there were several small rounded

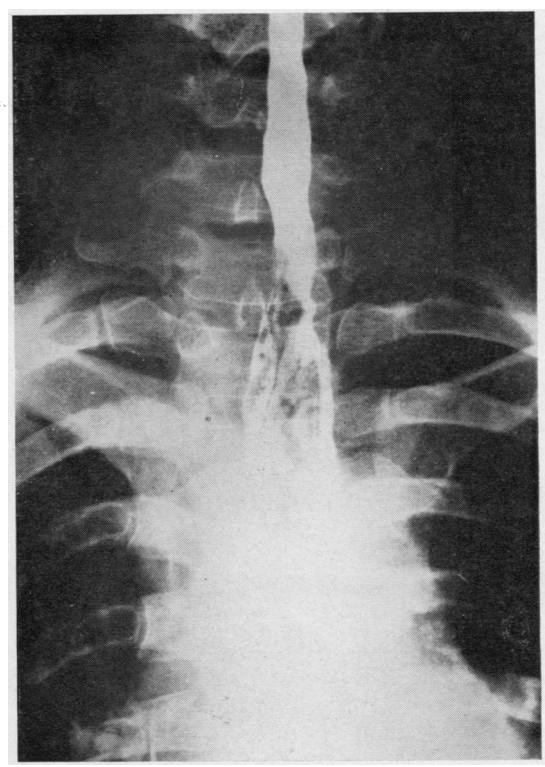

FIG. 1. *Tracheogram using iodized oil inserted via a nasal catheter. The upper* 4 *in.* (100 *mm.*) *of the trachea are narrower than the lower part, and nodular projections can be seen at the lower end of the narrowed segment.*

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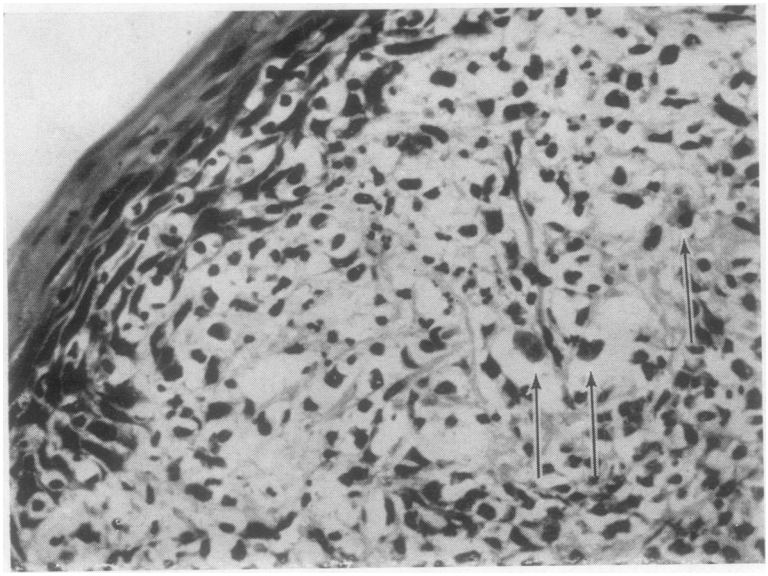

FIG. 2. *High-power view of trachea showing stratified squamous epithelium, submucosal cellular infiltrate and oedema, and absence of fibrosis. Arrows point to examples of Russell bodies. H. and E.,* × 50.

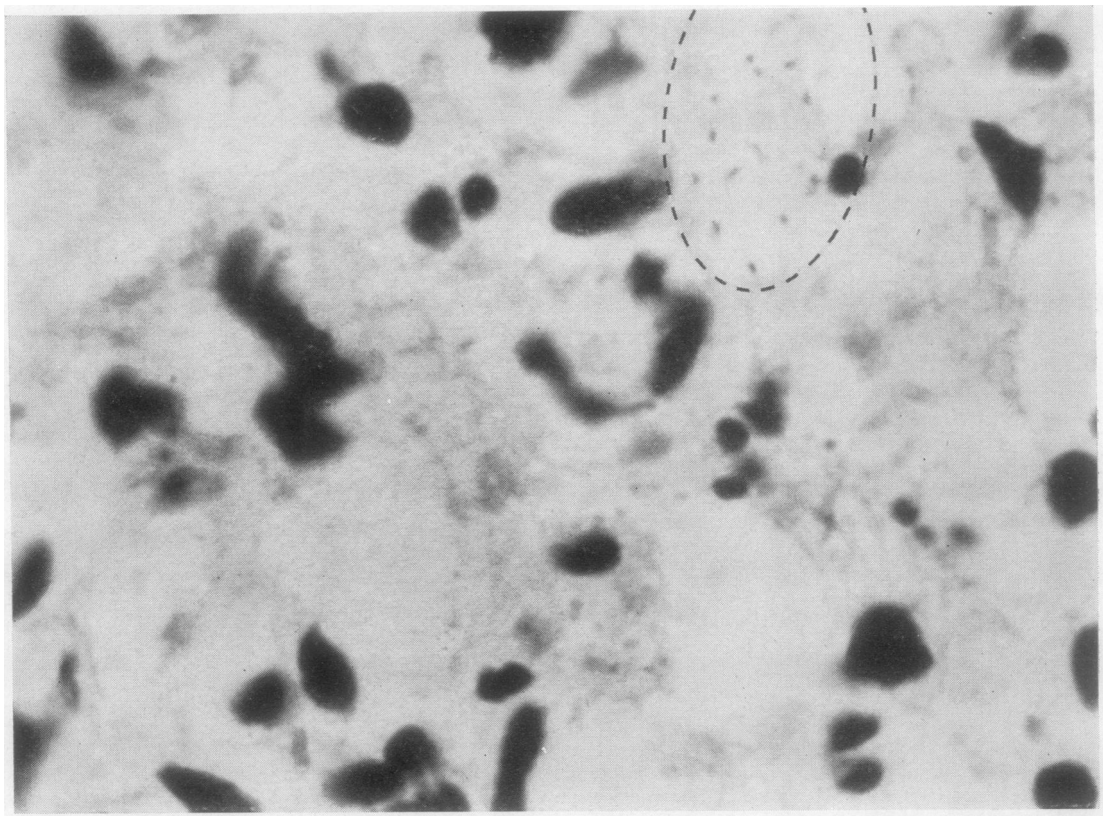

FIG. 3. *Oil-immersion view of submucosa of trachea. The dotted line surrounds a group of intracellular bacteria which had the histological appearances of Frisch bacilli. The cell outlines are not visible. Giemsa,* × 750.

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marginal filling defects (Fig. 1). Swabs of the nose and throat grew *Staphylococcus aureus*, sensitive to all routine antibiotics. A full blood count was normal, serological tests for syphilis were negative, and a chest radiograph was normal.

Nodules from the trachea were removed for culture and for histology. Cultures grew *Pseudomonas pyocyanea* and *Staph. aureus*. The Pseudomonas was sensitive only to streptomycin. On histological section the tracheal biopsy specimen showed that the epithelial lining was a rather hyperplastic stratified squamous epithelium. In the submucosa there was a heavy chronic inflammatory infiltrate of lymphocytes, macrophages, and a large number of plasma cells. Russell bodies were present in some of the plasma cells (Fig. 2). Mikulicz cells were present but were scanty; they consisted of foamy cells containing short Gram-negative bacilli compatible with the histological appearances of Frisch bacilli (Fig. 3). No acid-fast bacilli were seen. Granulation tissue was present, but reticulin stains showed little increase in reticulin fibres.

The patient was given a course of intramuscular streptomycin, 2 g. per day for five days. She was then discharged and readmitted for review in one month. At the time of her return she stated that the cough and throat irritation had been lessened, but that the condition of her nose was unaltered. Bronchoscopy on this occasion showed considerable improvement in the trachea; no granuloma was present and no further biopsy was taken. Six months later she was again reviewed, and on this occasion she had a return of the unproductive cough. Bronchoscopy revealed an area resembling a papilloma in the subglottic region on the anterior wall of the trachea. This lesion was removed for culture and biopsy. Both procedures were done independently at the Brisbane Hospital as well as at the Princess Alexandra Hospital. Both cultures grew *Ps. pyocyanea* alone and both biopsies were reported as showing the histological appearances of scleroma. As the Pseudomonas was still sensitive to streptomycin, the patient was given a further course of injections to a total of 10 g. in five days. The nasal mucosa appeared unaltered at the end of this treatment.

DISCUSSION

The patient came from a known endemic area of scleroma, but for 15 years had been in Australia, where scleroma is almost unknown. The only other case report from Australia seems to be that of a migrant from Sicily (Bulteau, 1963; *Med. J. Aust.*, 1963). Bulteau's patient had been in Australia for seven years, the nose was obstructed by pink fleshy tumours, the histology was said to show the appearance of rhinoscleroma, and *Klebsiella rhinoscleromatosis* was cultured from the lesions.

In our patient the nose was the site of an atrophic rhinitis, which may be one of the early features of scleroma (Manson-Bahr, 1957). The bronchoscopic and radiological appearances of the tracheal lesions were suggestive of scleroma. The diagnosis was confirmed by the histological findings of hyperplastic epithelium, infiltrate rich in plasma cells, with Russell bodies, and Mikulicz cells containing Gram-negative bacillary forms (Ellis, 1952; Fisher and Dimling, 1964). Russell bodies in plasma cells are highly suggestive of the disease but are not specific, as they may be seen in other conditions when an infiltrate rich in plasma cells is present, for example syphilis, tuberculosis, squamous-cell carcinoma, and mycosis fungoides (Lever, 1961). The Mikulicz cells are considered to be diagnostic (Manson-Bahr, 1957; Editorial, 1964). These features were associated with repeated cultures of Pseudomonas and repeated failure to culture Klebsiella. This case thus has similarities to that described by Fisher and Dimling (1964). Their patient showed mucosal changes, infiltrate rich in plasma cells, Russell bodies, and larger cells containing intracellular bacillary forms. They also were unable to grow Klebsiella but cultured *Proteus mirabilis*. They considered that the electron microscope appearances of the intracellular bacilli resembled those of *Pr. mirabilis* from culture rather than those of Klebsiella. The case of Fisher and Dimling and the patient described now support the view that the Klebsiella organisms may have a symbiotic role in the causation of this disease (Editorial, 1964). In both cases the histological appearances by light microscopy were compatible with the Frisch bacillus, but other organisms were cultured and could be presumed to play a similar role.

The response to streptomycin in our patient suggested that a streptomycin-sensitive organism was associated with the tracheal granuloma but not necessarily with the atrophic condition of the nasal mucous membrane. The history supports the view that the original mucosal change began in Poland 15 or more years previously and that a further influence (*i.e.*, superadded Pseudomonas infection) produced a granulomatous reaction in the trachea over the last 18 months. In spite of the length of history there was minimal fibrosis. The amount of streptomycin given was less than the three- or four-week course advocated by Manson-Bahr (1957), but our patient did not have the gross lesions that have been described.

SUMMARY

A case history is presented in which typical changes of scleroma appeared in a Polish woman who had been in Australia for 15 years. These

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changes were associated with repeated positive cultures for *Ps. pyocyanea* and repeated negative cultures for Klebsiella. There was considerable improvement in the trachea after short courses of intramuscular streptomycin, but the accompanying atrophy of nasal mucosa appeared unaltered. The possible role of bacteria in the pathogenesis of this disease is discussed.

The diagnosis of scleroma was originally suggested by Dr. J. Dickson, of Toowoomba. Bronchoscopies and biopsies were performed by Dr. C. Wark. Original histological sections were reported on by Drs J. Little and A. Pound and the repeat sections by Drs J. Little and B. Gutteridge. Drs A. Rao and J. Harper were responsible for the cultures. The photographs were provided by Dr. A. Burry and Mr. D. Crowley.

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