Tracheobronchopathia osteochondroplastica

THREE CASE REPORTS

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TRACHEOBRONCHOPATHIA osteochondroplastica (TO), also known as tracheopathia osteoplastica (Aschoff, 1910), is a rare condition which was first described by Wilks (1857). Up to 1957 there were only 187 cases of TO reported in the literature according to Pedersen and Worsøe-Petersen (1969). Further cases have since been described (Baird and McCartney, 1966; Campbell and Gould, 1968; Soulas *et al.*, 1969). Ragaini and Piccoli (1957) give the frequency of TO as 3 per one thousand autopsies. Eimind (1964), however, reported that no instance of TO had been observed at bronchoscopy of some 25,000 patients at Ullevål Hospital in Oslo.

TO is characterized by multiple wax-like papules which coalesce, to form irregular plaques consisting of submucous osseocartilaginous foci which sometimes assume the character of papillomas and bulge into the lumen of the affected air passage. The lesions are usually confined to the lower two-thirds of the trachea, but sometimes estend down into the main bronchi and occasionally up into the larynx and nasopharynx (Schnitzer, 1920). In the trachea and bronchi the changes are confined to the pars cartilaginea; only one case with changes in the pars membranacea is on record (Way, 1967). According to Spencer (1962) TO occurs mainly in men, aged 50–60, and rarely in women. Only one case of TO has been seen in a child (Zajaczowska, 1969).

Case 1

In July 1969 a 53-year-old woman was admitted to the department of internal medicine, Umeå hospital, for investigation because of muscle pain. Scleroderma was assumed.

In December 1970 she was re-admitted, this time because of acute severe muscle pain, abnormally high level of muscle enzymes and tachycardia. She died one week later in a state of severe shock. Autopsy revealed necrotizing myositis (dermatomyositis), widespread partly confluent pulmonary haemorrhages and severe fatty degeneration of the liver. The trachea, particularly in the lower part, and the main bronchi showed submucous, white firm nodules largely following the hyaline cartilages (Fig. 1). The mucosa was macroscopically intact. The changes were very widespread, but confined to the pars cartilaginea.

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Microscopically, the submucous lesions were built up of cartilage and ramifying bone trabeculae with Haversian canals and marrow spaces filled with fatty marrow. The surrounding loose connective and fatty tissues contained remnants of mucous glands and normal elastic fibres. Here and there the osseocartilaginous islands were seen to extend from the tracheal cartilage (Fig. 2).



FIG. 1 (Case 1).
Pars cartilaginea of lower part of trachea shows yellow-white, raised, confluent nodules. The changes extend down to the carina.

Case 2

A 56-year-old man suddenly died while in hospital because of hypertension (180/110) and angina pectoris. Autopsy showed generalized arteriosclerosis, pulmonary oedema and congestion, but no distinct, recent myocardial infarction. The pars cartilaginea in the upper trachea showed streaky, longitudinal, hard whitish, partly calcified submucous plaques, which microscopically consisted of subepithelial islands of cartilage and bone (Fig. 3). No ecchondrosis was seen with certainty. The elastic fibres were of normal appearance.

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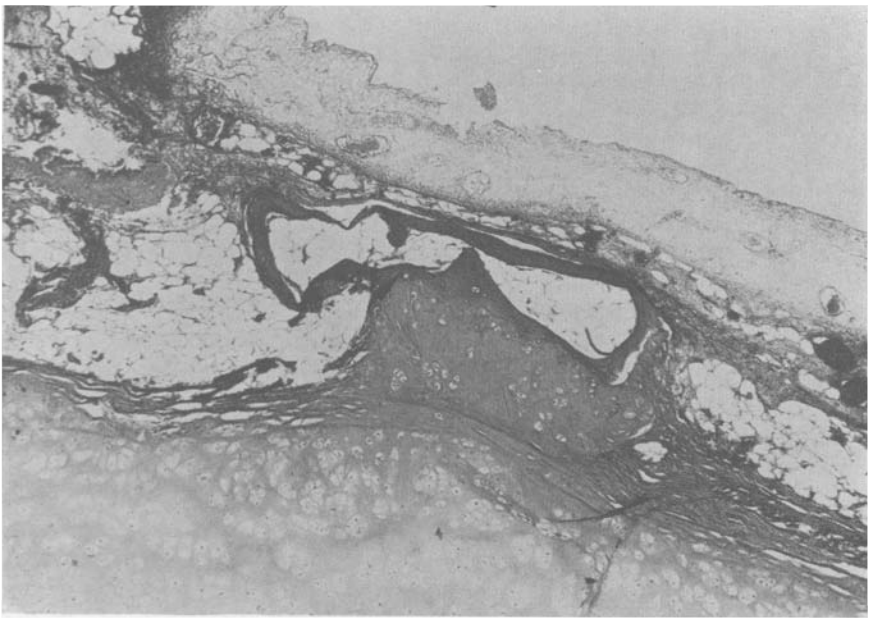


FIG. 2 (Case 1).
Ecchondroma with overlying bone, extending from the tracheal cartilage. The mucosa covering the lesion is loose. (Haematoxylin-eosin × 50.)

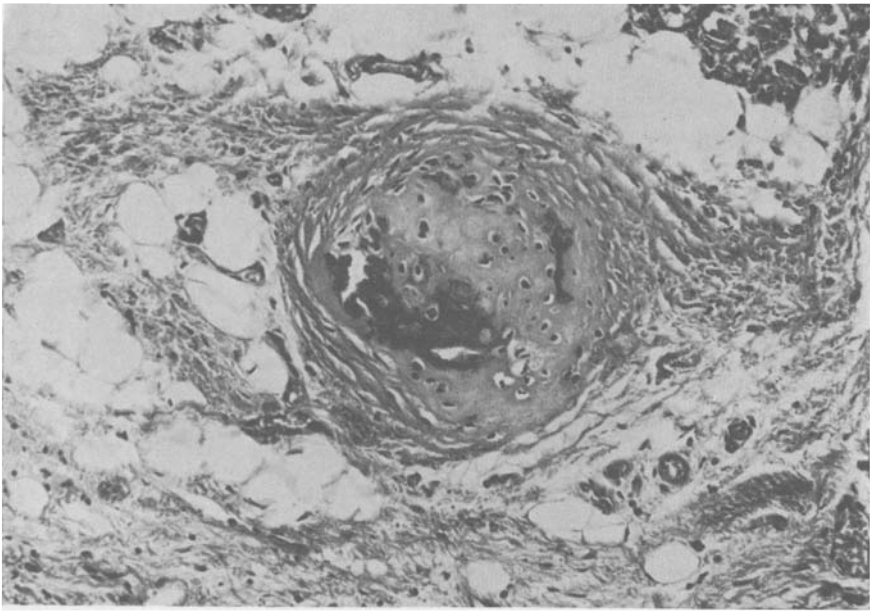


FIG. 3 (Case 2).
Submucous fibrous focus with cartilage and bone formation without visible connection with the tracheal cartilage. Elastic fibres in the vicinity are of normal appearance. Deposition of lymphocytes in the mucosa. (Haematoxylin-eosin × 170.)

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Case 3

A 55-year-old man who had had respiratory symptoms and repeated sinusitis for twenty years. He had repeatedly been treated because of chronic bronchitis and bronchiectasis. During the last 3–4 years he had gradually lost his sense of smell and he complained of continuous irritation in the nose and of a green, mucous nasal discharge. His general condition was good. Chest X-ray showed no parenchymal changes. Culture of the sputum gave abundant growth of two variants of *Proteus mirabilis*. After treatment of the patient with gentamycin^R, culture of the sputum was negative. Physiologic tests showed the lung volumes and ventilation to be normal.

Bronchoscopy disclosed crowded papilla-like formations in the trachea, main bronchi and stem bronchi, but not in the lobar bronchi. Histologic examination of biopsy specimens showed a well-differentiated osseo-cartilaginous tissue with fat-filled marrow spaces. In some areas the mucosa showed squamous epithelial metaplasia but no atypia. Sparse lymphocytes and plasma cells were seen subepithelially.

Discussion

Tracheobronchopathia osteochondroplastica has been known for more than a hundred years, but its aetiology is still unknown. Many theories have been advanced, such as tumour formation, dysembryonic disease, systemic disease of the elastic connective tissue and possibly also mechanical and hormonal factors. The existence of some relation between TO and certain other disease, such as tuberculosis or chronic bronchitis, has also been considered, but no supportive evidence has been found in any of the 216 cases on record (Ragaini-Piccoli, 1957; Pedersen and Worsøe-Petersen, 1969).

According to Virchow (1863), the changes in the trachea arise in the form of ecchondromas, which afterwards undergo ossification and calcification. In those cases where no connection can be seen between the tracheal cartilage and osseocartilaginous islands, it may have been interrupted by extreme elongation and thinning. This theory may explain why no changes are seen in the pars membranacea. Aschoff (1910) and later Dalgaard (1947), ascribed the changes to metaplastic conversion of the elastic connective tissue to elastic cartilage, which is afterwards ossified and calcified. But the absence of changes in the pars membranacea, where elastic fibres are abundant, argues against this theory. Furthermore, normal elastic fibres are often seen close to the TO-changes without being continuous with them (Bowen, 1959).

According to Hempel and Gläser (1958), the changes are due to metabolic or inflammatory disorders producing acidosis in the tissue with hyaline degeneration as a result and secondary formation of new cartilage and bone. This theory is compatible with the other two mentioned above.

The changes in our three cases appear to be compatible with both Virchow's theory and with Aschoff-Dalgaard's. Ecchondromas were seen

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in Case 1, while in Case 2 the changes consisted of isolated islands of metaplastic cartilage and bone. In Case 3, in which only a biopsy specimen was available, the relation of the metaplastic bone tissue to the underlying cartilage could not be examined.

TO is, as a rule, asymptomatic for a long time and is often diagnosed accidentally at autopsy. The symptoms are non-characteristic and consist of cough and hoarseness, dyspnoea and occasionally haemoptysis (Eimind, 1964). Narrowing of the lower part of the trachea and main bronchi has occasionally been observed (Clerf, 1944) as well as bronchiectasis distally to the lesions. If the changes are severe, they will probably impair pulmonary function and cause progressive dyspnoea.

Roentgenologic changes in TO are uncommon and are due mainly to secondary lung changes produced by obstruction. Pneumonia and atelectasis may develop distally to local bronchial lesions (Eimind, 1964). Tomography, possibly with contrast medium, is diagnostically useful and will show narrowing of the lumina and the submucous osseocartilaginous formations in the trachea and bronchi (Howland and Good, 1958; Secrest *et al.*, 1964). But confirmation of the diagnosis *in vivo* requires bronchoscopy (Lell, 1953). The bronchoscopic picture is very characteristic: the wall of the trachea and bronchi show irregular, non-pedunculated or polypoid nodules, partly coalescing into plaques (Lell, 1953; Huzly, 1960). Passage of the bronchoscope over these changes produces a harsh, scraping sound as if the bronchoscope were being passed through sand (Baird and McCartney, 1966).

Summary

Three cases of tracheobronchopathia osteochondroplastica are described, one diagnosed bronchoscopically *in vivo*, the other two at autopsy. The disease, which is rare, has a typical gross appearance which is readily recognised at bronchoscopy. Microscopically, the picture shows isolated islands of metaplastic bone and cartilage as well as ecchondromas. The pathogenesis and clinical manifestations are discussed.

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