TRACHEOPATHIA OSTEOPLASTICA

BY

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Tracheopathia osteoplastica is a rare benign abnormality of the trachea and bronchi characterized by numerous cartilaginous and bony nodules within the tissues of the submucosa without ulceration of the surface.

The trachea, and, to a lesser extent, the bronchi, become narrow, rigid, and inelastic, the surface of the air passage presenting a corrugated and "cobblestone" appearance due to the projection of many sessile plaques and polyps into the lumen from the underlying tissues.

In most instances the age incidence has been 40 to 60 years of age, with exceptions at 12 and 70 years, the sexes being equally affected.

According to Hiebaum (1934), Luschka described the condition in 1856, a year before the first generally recognized publication by Wilks (1857). The latter found evidence of bony deposits in the larynx, trachea, and bronchi in a 38-year-old man who died of pulmonary tuberculosis.

Dalgaard (1947) reviewed the literature of some 90 cases recorded at that time and included a detailed histological examination of the trachea in a personal case from a man 67 years of age. Since then further reports have been published by Rønnov-Jessen (1952), Pedro (1953), Lell (1953), Padovan (1954), Carr and Olsen (1954), Delord (1954), Moser and Them (1954), Braun and Dhom (1954), Heyden (1954), Dalgaard (1955), Caliceti and Lura (1955), Sors, Rose, Villeneuve, and Cottin (1955), Sobczynski (1955), Simonin, Grilliat, Massinot, and Simon (1957), Israel, Gilbert, and Lechien (1957), Pieri and Casalonga (1957), Hutás and Zsámbéky (1957), and Hempel and Gläser (1958), who collected the largest single number of cases (eight) so far recorded.

Case Report

A man of 61 years of age, a retired civil servant, and in good health previously, developed a left-sided hemiplegia and died the following day in hospital.

Necropsy Report.—The body was that of an elderly man of average build and nutrition, with no external

injuries. The surface of the right cerebral hemisphere was flattened, with loss of the normal convoluntary pattern due to an underlying area of infarction in the substance of the right cerebral hemisphere. This was in the distribution of the right middle cerebral artery which was obstructed by atheroma one inch from its origin, the arteries of the brain showing advanced atheromatous changes.

The heart weighed 15 oz. and showed some hypertrophy of the left ventricle. There was no valvular disease, the aorta contained numerous plaques of atheroma and the coronary arteries patchy atheromatous changes without occlusion. The remaining systems of the body, with the exception of the respiratory tract, were quite healthy. The cause of death was cerebral infarction due to cerebral arteriosclerosis.

The upper respiratory tract was healthy and the epiglottis and larynx free from disease. From a point 3 cm. below the level of the vocal cords to the level of the second divisions of the bronchi, the respiratory tract was converted into a narrow triangular-shaped channel, the rigid walls showing numerous bony, hard plaques and polypoidal outgrowths from the underlying tissues (Fig. 1a and 1b). There was no evidence of mucosal ulceration and the membranous portion of the trachea was uninvolved. The normal pattern of the tracheal rings was completely obscured, being identified again only in the bronchial subdivisions. The smaller bronchi were unaffected, and the lungs showed no gross evidence of acute or chronic infection.

Histology.—Sections of the trachea showed the mucosa replaced by a layer of collagen, the submucosal tissues containing evidence of chronic inflammation, including groups of plasma cells and dilated capillary vessels. In the trachea, two distinct layers of partly cartilaginous and partly bony islands were seen. First, immediately beneath the mucosal surface a number of small cartilaginous nodules were evident, some showing calcification and in a few places bone formation (Fig. 2). This layer was intimately related to the internal elastic band of the trachea which showed fragmentation, especially in the neighbourhood of the islands, but at no point did the collagenous fibres enter the newly formed cartilage, nor did the collagenous fibres stain with elastic stains. In the deeper layers, larger islands

FIG. 1a.—Necropsy specimen showing appearance of the trachea and bronchi in tracheopathia osteoplastica.

FIG. 1b.—Radiograph of necropsy specimen showing irregular distribution of calcification in tracheal nodules.

FIG. 2.—Section of tracheal wall with superficial submucosal nodules lying between the tracheal lining and normal cartilage which is evident at one edge. (Van Gieson, $\times 30$.)

FIG. 3a.—Section of the tracheal wall showing the relationship of bony nodules to two normal hyaline tracheal rings. A centrally placed space of adipose tissue is evident in one nodule. (Haematoxylin and eosin, $\times 12$.)

FIG. 3b.—Section of bronchial deposit with well-defined perichondrial membrane. (Haematoxylin and eosin, $\times 50$.)

FIG. 4.—Section of tracheal cartilage merging with the edge of an ectopic bony island, there being no visible perichondrial membrane. (Haematoxylin and eosin, $\times 125$.)

FIG. 5.—Section of bony island showing haemopoietic tissue within the marrow space. (Haematoxylin and eosin, $\times 120$.)

were present (Fig. 3a), many of them osseous and all attached to the superficial aspect of the tracheal rings. In many instances a perichondrial membrane was interposed between the surfaces. This was especially prominent in some of the bronchial deposits (Fig. 3b), but occasionally no such demarcation was evident, the bone merging with the hyaline tracheal cartilage (Fig. 4). In the largest islands a centrally placed space was evident, filled with adipose tissue and normal haemopoietic cells (Fig. 5).

The lungs showed well-marked congestion with fibrinous material in many alveoli as well as evidence of acute inflammation and exudate in the smaller bronchi.

Discussion

The aetiology of the condition is unknown, although numerous theories have been advanced by various writers. These include non-specific chronic infection, tuberculosis, syphilis, mechanical or chemical irritation, and tumour formation. Delord (1954) considered it to be a degenerative process associated with age and chronic infection.

It is likely that infection is coincidental or the result of the condition rather than a factor in its production, and the irritative and tumour theories are not supported by the histological findings.

Two views of more substance are that the condition is the result of an *ecchondrosis* from the normal tracheal rings, or a metaplasia of the connective tissue of the submucosa of the trachea.

Virchow (1863) considered that the plaques were outgrowths from the tracheal ring, a view supported in later years by serial sections and stereoscopic radiography. However, in many instances the plaques have been crowded into tracheal interspaces, and histological examination has shown no connecting stalk with the tracheal rings, findings that do not support the theory of *ecchondrosis*.

Aschoff (1910) gave the present name to the condition, regarding it as a metaplasia of the elastic connective tissue cells, whereby undifferentiated connective tissue cells in the superficial layers of the submucosa in close proximity to the elastic fibres became elastic cartilage cells. Other writers, notably Dalgaard (1947), visualized the deposition of calcium in these cells, their invasion by vascular connective tissue to give the so-called "marrow spaces," and their subsequent conversion into bone, following the activity of osteoblasts. No mention is made of haemopoietic tissue in the central spaces, and Pedro (1953) considered the spaces to be submucosal adipose tissue enclosed in the process of formation of an island.

Microscopy in the present case does not support the theory of origin as metaplasia. In the smaller islands elastic connective tissue was found close to the cartilaginous and bony islands, but there was no visible continuity of the elastic threads into the abnormally placed cartilage, and no evidence that the cartilage was elastic in nature. The relationship between the cartilage and elastic tissue appeared to be no more than a geographical one. In the larger, more deeply placed islands, where they were fused with the tracheal rings, and especially where there was no visible perichondrial margin, the appearances suggest an origin from the hyaline tracheal ring.

Clinically the condition is probably not as rare as reports suggest and it is surprising that more instances have not been recorded after bronchoscopy. As far as can be ascertained, there has been no published report in this country since Wilks's original publication. The majority of some 120 cases reported have been found incidentally at necropsy, as in the present instance.

the remainder having been discovered on bronchoscopy, generally in patients with long-standing chronic lung infection and recurrent haemoptysis. The bronchoscopic findings are quite characteristic, the instrument grating and rasping along the surfaces of the bony nodules in the narrowed air passages. Histological examination of material obtained at biopsy has aided diagnosis in some instances.

No specific treatment is available, antibiotics being required where obstruction of the air passages had led to infection. Dalgaard (1955) reported a carcinoma arising in an upper lobe bronchus affected by the condition and regarded this as a complication, an event which could equally well have been fortuitous.

Summary

A case is reported of tracheopathia osteoplastica found incidentally at necropsy. In this rare benign condition of the trachea and bronchi numerous submucosal cartilaginous and osseous nodules project into the lumen causing distortion and narrowing and, where the smaller air passages are involved, chronic lung infection. Histologically ectopic nodules are seen superficially in the submucosal tissues and in the deeper layers, fused to the tracheal rings. The aetiology

of the condition remains unknown, although many theories have been proposed including development as an ecchondrosis or as a metaplasia of the elastic connective tissue with subsequent calcification.

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