

## TRACHEOPATHIA OSTEOPLASTICA

BY

R. SALM

*From the Rivers Pathological Department, Camborne-Redruth Hospital, Redruth, Cornwall*

(RECEIVED FOR PUBLICATION MAY 5, 1961)

The condition for which Aschoff (1910) coined the name of tracheopathia osteoplastica has been known for over a hundred years and has been reported from many countries. It is characterized by the development of multiple small, cartilaginous and bony islets in the connective tissue of the mucosa and submucosa of the trachea and, to a lesser extent, of the main bronchi. Dalgaard (1947) collected about 90 cases from the literature, and Bowen (1959) added another 30. Further cases were recorded in 1960 by Jepsen and Sørensen in Denmark, and by Shuttleworth, Self, and Pershing in the United States. In Britain there have been only two previous reports in the course of a century: the original observation of the condition by Wilks in 1857, and Bowen's case reported in 1959. The purpose of this communication is to record the third case from this country.

### CASE REPORT

The patient, who had been an actor of stage and screen, was admitted in October, 1960, aged 75, complaining of pain in the upper abdomen and shortness of breath for one year, and anorexia, loss of weight, and lassitude for three months. A large mass was palpable in the right upper abdomen, and radiological examination of the chest showed another mass in the upper mediastinum which was thought to be an intrathoracic goitre. His condition gradually deteriorated and he died one month after admission.

**LABORATORY EXAMINATIONS.**—Haemoglobin 68% ; E.S.R. 115 mm. after one hour ; blood urea 65 mg.% ; serum alkaline phosphatase 47 units ; serum glutamic-oxalacetic transaminase 62 units ; serum glutamic-pyruvic transaminase 28 units ; occult blood test repeatedly weak positive.

**NECROPSY (P.M. 60/108).**—A polypoidal gastric adenocarcinoma, measuring  $7.5 \times 4 \times 3$  cm., was present 6 cm. above the pylorus. The liver was grossly enlarged, weighing 5,150 g., due to very large numbers of small secondary deposits. The right thyroid lobe was adenomatous, measured  $10 \times 7 \times 6.5$  cm., weighed 175 g., and was situated largely intrathoracically. The heart weighed 390 g. and was

distended and flabby in consistency. The lungs showed diffuse emphysema and confluent bronchopneumonic consolidations were present in the right lower lobe. The spleen was enlarged and chronically congested, weighing 430 g.

**Upper Respiratory Tract.**—The larynx was healthy, as was the adjoining part of the trachea. From about 5 cm. below the vocal cords the inner tracheal surface was of a corrugated appearance due to numerous bony-hard protuberances (Fig. 1). These did not appear to transgress above the epithelial lining, as the mucosal surface felt uneven but otherwise quite smooth. They presented mainly as thin, narrow ridges, measuring up to 1 cm. in length and 3 to 4 mm. in width, and were placed longitudinally and very frequently across the interannular spaces. The membranous part of the trachea was not involved. The main bronchi showed only a few scattered, similar tiny bony nodules, the abnormality being mainly confined to the lower three-quarters of the trachea, as was well demonstrated on radiological examination (Fig. 2). The tracheal wall was more rigid than usual, but its lumen was but slightly narrowed.

**HISTOLOGICAL EXAMINATION (60/2639).**—The sections confirmed the presence of multiple, small, bony and occasionally cartilaginous islets within the tracheal mucosa and submucosa (Fig. 3). They were covered by intact tracheal epithelium. Many of the osseous nodules contained comparatively large centres of fatty or slightly cellular bone marrow. Many bony ridges were placed across the interannular spaces and were mostly separated from the cartilaginous tracheal rings by bulky mucous glands. These were normal in appearance, except for dilatation of an occasional excretory duct beneath a bony plaque (Fig. 4). In the narrower mucosal layers the bony islets would sometimes be found adjacent to a tracheal ring, but they were always separated from it by intact perichondrium, which itself showed no connexion with the bony tissues. Special stains demonstrated the presence of normal elastic fibres. The external elastic fibres could usually be traced running above the bony protuberances, whereas the internal elastic system had been interrupted by the nodules, without there being any demonstrable connexion between them (Fig. 5). There was no evidence of any inflammatory condition or of any degenerative changes.

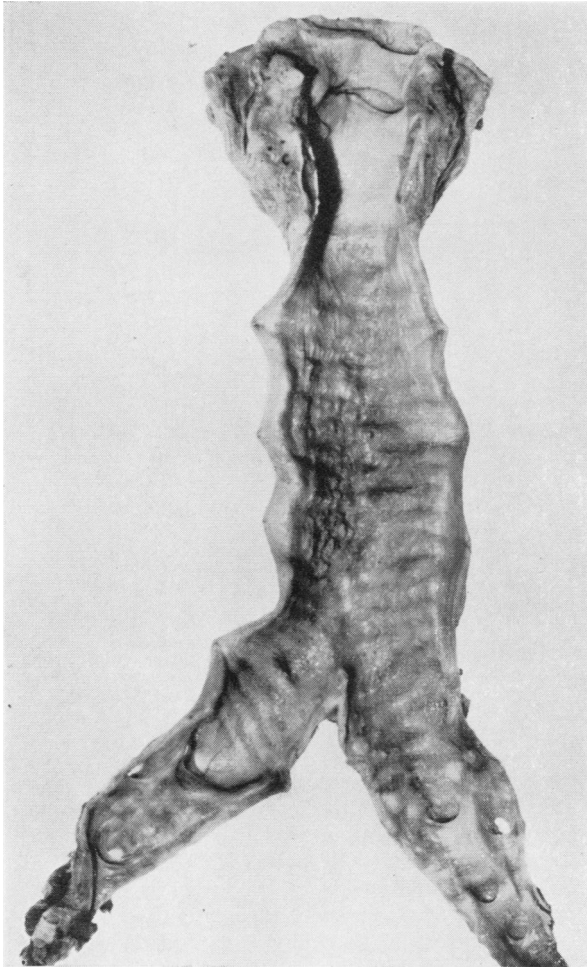


FIG. 1.—Opened tracheal specimen, taken with light source to the left, throwing bony ridges on left side into relief,  $\times$  about  $\frac{1}{2}$ .

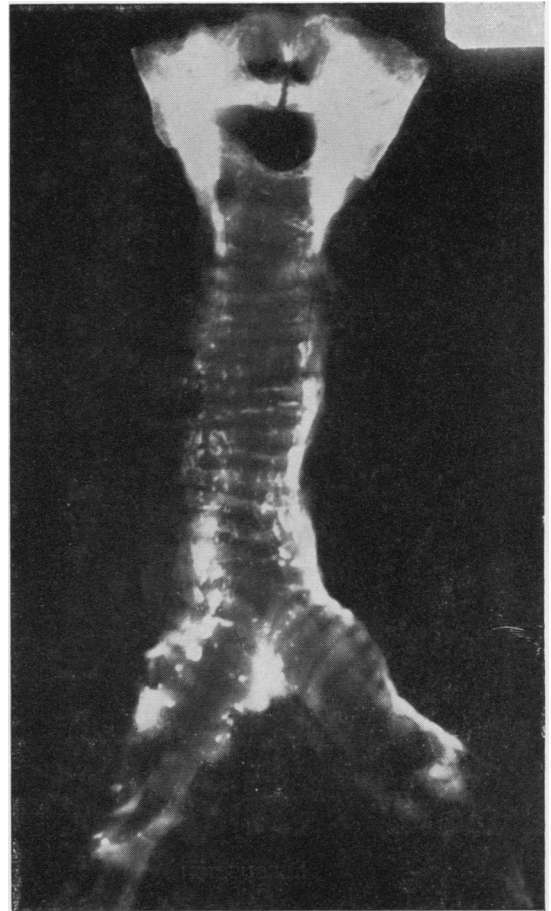


FIG. 2.—Radiograph of tracheal specimen showing many small bony shadows of lower trachea and bifurcation.  $\times$  about  $\frac{1}{2}$ .



FIG. 3.—Trachea (low-power view) showing subepithelial bony ridges with much fatty marrow spanning the inter-annular spaces. Haematoxylin and eosin.  $\times 10$ .



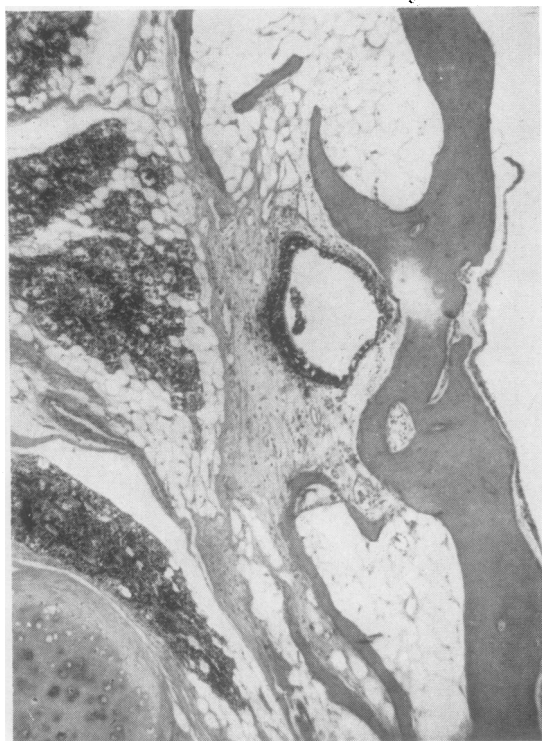


FIG. 4.—Part of bony plaque, separated from tracheal cartilage by mucous glands, with dilated excretory duct beneath it. Haematoxylin and eosin.  $\times 36$ .

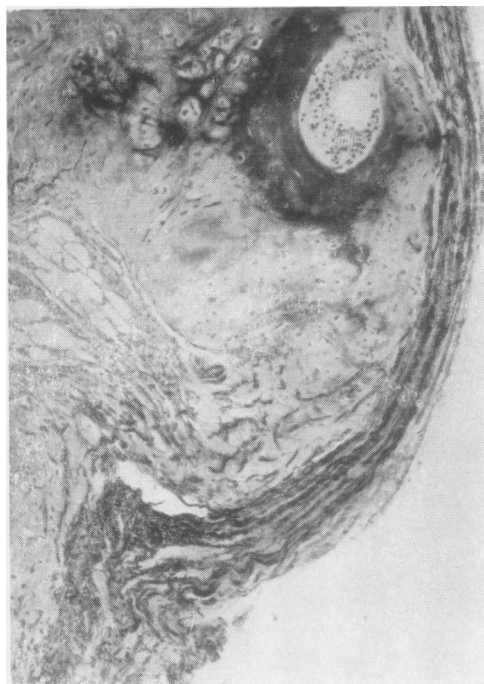


FIG. 5.—Subepithelial chondromatous nodule showing central ossification with sparsely cellular marrow. Intact external elastic fibres running above it; internal elastic fibres are interrupted. Verhoeff-van Gieson.  $\times 90$ .

### DISCUSSION

Tracheopathia osteoplastica is a condition of some interest on account of its rarity and the problem of its histogenesis. Most cases, like the present one, have been asymptomatic and were discovered incidentally at necropsy. Occasionally the condition is reported to have produced haemoptyses (Clerf, 1944; Shuttleworth *et al.*, 1960). But when discovered during bronchoscopy the features were impressive: Dalgaard (1955) speaks of a "veritable mountainscape," and Jepsen and Sørensen (1960) of "stalactite formations." They as well as Clerf (1944) stress the characteristic, "unforgettable" grating sound of metal against bone during insertion of the bronchoscope.

As a rule the condition tends to be confined to the lower two-thirds of the trachea, but the lesions may extend into the bifurcation and, as in the case of Shuttleworth *et al.* (1960), the abnormality may be confined to a large bronchus, producing bronchiectasis, chronic inflammation, fibrosis, and atelectasis in the distal lung segment.

Several theories have been advanced to explain the histogenesis of the lesions. A developmental causation has been considered by Dalgaard (1947) and Hempel and Gläser (1958) but rejected, as the condition has never been observed in young children. Multiple ecchondroses with or without secondary ossification may exceptionally occur in the trachea (Liebow, 1952), and tracheopathia osteoplastica has been regarded, mainly in the older literature, as ossified cartilaginous outgrowths from the tracheal rings. However, the collective evidence of subsequent studies has been against this view. In almost all cases the subepithelial islets consisted preponderantly of bony tissue, and lacked evidence of secondary ossification. In some instances the bony structures were placed almost exclusively horizontally over the tracheal interspaces (Wilks, 1857; Clerf, 1944; Dalgaard, 1947) and were quite unconnected with the cartilaginous tracheal rings. In those cases in which an occasional fusion between islands and tracheal rings had occurred this was due to a secondary, incidental process (Aschoff, 1910).

Aschoff propounded the theory that the abnormality was due to a systemic disease of the tracheal elastic system. This, possibly due to a misunderstanding of his thesis owing to language difficulties, caused considerable confusion, although he had been careful to point out that the elastic fibres themselves are not transformed into cartilage and bone, but that this development merely takes place in the topographical area of the elastic system. Later workers, however, have misinterpreted his thesis and have sought to demonstrate—unconvincingly—a direct relationship between elastic fibres and elastic cartilage, with transitions between them (Dalgaard, 1947). Hempel and Gläser (1958) attributed the lesions to local acidosis or a generalized metabolic disturbance, for which there is no proof and which can only be regarded as speculation.

The most likely explanation of tracheopathia osteoplastica is that the lesions are due to bony and, less frequently, cartilaginous metaplasia of the subepithelial tracheal connective tissue. That the metaplastic islets are often surrounded by, or interrupt, elastic fibres is entirely due to the chance of their topographical relationship. What stimulus triggers off this process of metaplasia is unknown, as it is largely unknown in other conditions. In this connexion it may be recalled that the lower respiratory system, as Willis (1958) has pointed out, may likewise develop bony, although not cartilaginous metaplastic lesions, either as the entity known as pulmonary microlithiasis, or as trabecular, branching ossifications.

Cartilaginous and bony metaplastic lesions in other non-neoplastic conditions, and not associated with haemorrhage, degenerative or inflammatory processes are rare, but are known to occur, for example, as bone in scar tissue, as bony plates in the dura mater, as the solitary "osteoma" cutis, in myositis ossificans, or in the urinary tract, especially under experimental conditions (Willis, 1958).

Another interesting feature is the observation of bone marrow in the osseous nodules and ridges of tracheopathia osteoplastica (Dalgaard, 1947 ;

Bowen, 1959 ; and present case), which has likewise been observed in other metaplastic lesions: Micheli and Fogliati (1952) described a massive bone marrow centre in an ossifying lipoma ; bone marrow is occasionally demonstrable in bone developing in the walls of calcified arteries, and has been found in calcified heart valves (Willis, 1958) ; and personal observations include bone marrow in bone of abdominal scar tissue, osteoma cutis, and silicotic nodules of lung. The haemopoietic tissue in all these different sites can only have arisen by a metaplastic process from dedifferentiated, "uncommitted" mesenchymal cells, and hence the finding of bone marrow within the bone of tracheopathia osteoplastica would appear to lend support to the interpretation of a metaplastic causation.

#### SUMMARY

The occurrence of tracheopathia osteoplastica, discovered incidentally at necropsy, in a man of 75 is recorded.

The literature and theories as to histogenesis are reviewed, and it is concluded that the condition is due to a process of metaplasia.

I wish to record my indebtedness to Professor R. A. Willis for Fig. 3 and for reading the manuscript. I am grateful to Dr. T. S. Wilson for the clinical data, and to Miss Phyllis E. Coleman for the photography.

#### REFERENCES

- Aschoff, L. (1910). *Verh. dtsch. path. Ges.*, **14**, 125.
- Bowen, D. A. L. (1959). *J. clin. Path.*, **12**, 435.
- Clerf, L. H. (1944). *Ann. Otol. (St. Louis)*, **53**, 839.
- Dalgaard, J. B. (1947). *Acta path. microbiol. scand.*, **24**, 118.
- (1955). *Nord. Med.*, **53**, 572.
- Hempel, K. J., and Gläser, A. (1958). *Virchows Arch. path. Anat.*, **331**, 36.
- Jepsen, O., and Sørensen, H. (1960). *Acta oto-laryng. (Stockh.)*, **51**, 79.
- Liebow, A. A. (1952). *Tumors of the Lower Respiratory Tract*, p. 119. *Atlas of Tumor Pathology*, Sect. V—Fasc. 17. Armed Forces Institute of Pathology, Washington, D.C.
- Micheli, E., and Fogliati, A. E. (1952). *Minerva chir. (Torino)*, **7**, 178.
- Shuttleworth, J. S., Self, C. L., and Pershing, H. S. (1960). *Ann. intern. Med.*, **52**, 234.
- Wilks, S. (1857). *Trans. path. Soc. Lond.*, **8**, 88.
- Willis, R. A. (1958). *The Borderland of Embryology and Pathology*, pp. 539–540. Butterworth, London.