Tracheopathia osteoplastica

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Tracheopathia osteoplastica is an unusual disease first described by Wilks in 1857. Even to the naked eye it is easily recognizable by the striking changes present in the trachea and major bronchi. It is a condition characterized by numerous mucosal, cartilaginous, and calcified plaques which project into the lumen.

Since Wilks's original observation, only one additional case (Bowen, 1959) has been reported in Britain, in contrast to other parts of the world where such cases have been recognized fairly frequently. For example, Dalgaard (1947), in a review of the world literature up to 1947, found 90 cases, and by 1959 the total had risen to 120. Huzly (1960) diagnosed two during routine bronchoscopy, while Carr and Olsen (1954) reported seven cases from the Mayo Clinic alone.

It is curious that the following case should be one of such rarity in this country.

CASE REPORT

The patient (A. H.), a welder aged 42 years, was admitted to hospital as an emergency on 15 November 1963 with uraemia and left ventricular failure. He complained of sudden severe breathlessness of two weeks' duration, with expectoration of heavily blood-stained sputum for some days before admission.

On examination he was pale with cold extremities but no oedema. His blood pressure was 220/120 mm. Hg.

The chest was dull over the bases with crepitations present.

Urinalysis revealed considerable albuminuria with 12 to 14 pus cells and a fair number of R.B.C.s per high-power field, in addition to hyaline and granular casts.

Blood count: Hb 10.9 g./100 ml. = 75 %; blood urea 70 mg./100 ml.; serum cholesterol 220 mg./100 ml.; total protein 8.15 g./100 ml.; albumin 3.75 g./100 ml.; globulin 4.40 g./100 ml.; albumin/globulin ratio 0.85/1.0; L.E. latex slide test negative.

A chest radiograph (Fig. 1) on 16 November 1963 was reported on as follows: 'Marked congestive changes in lung fields. Superimposed bronchopneumonia mid-zone areas. No cardiac enlargement.'

The patient was drowsy, restless, and confused from the time of admission; he became increasingly uraemic with the blood urea rising rapidly to 196 mg./100 ml. within four days, and he died on 21 November 1963.

NECROPSY The deceased was a pale man with slight puffiness of the face.

The brain showed slight oedema.

The lungs showed marked oedema, especially of the right side and of the bases.

The heart showed moderate left ventricular hypertrophy. The coronary arteries were patent. The aorta showed no gross atheroma.

The liver was somewhat enlarged and showed a moderate degree of chronic venous congestion.

The spleen was enlarged to almost twice its normal size and was congested.

The kidneys were markedly enlarged. The capsule stripped readily, revealing a smooth, rather haemorrhagic surface. On section the cortex was pale and toxic, in contrast to the rather congested pyramids. There was no evidence of hydronephrosis. On histological examination the kidney showed enlargement of the glomeruli, with marked endothelial proliferation of the tufts typical of acute diffuse glomerulonephritis.

Apart from the trachea, described below, the other organs showed no gross pathological changes, and the cause of death appeared to be acute left ventricular failure due to hypertension and acute nephritis.

The trachea appeared normal externally. On opening it in the routine manner by slitting it along the posterior wall, a remarkable appearance was observed. The whole of the mucosa of the anterior and lateral parts of the trachea down as far as the bifurcation was thickly studded with small grey-white, hard tuberosities measuring up to 3 mm. in diameter. These stood out prominently against a pinkish background of apparently normal mucosa, giving a picture which mirrored very closely that depicted in Huzly's (1960) beautiful endoscopic illustrations of tracheopathia osteoplastica. The absence of any lesions on the posterior membranous part of the trachea was a striking feature.

Microscopically the nodules were seen to be in the submucosa. The majority were calcified, and in many cases the process had gone on to the formation of woven and even of lamellar bone. There were areas where the latter was disposed around a small cavity containing fat and a few small collections of cells resembling erythropoietic tissue (Fig. 2). There were
FIG. 1. Chest radiograph showing marked congestive changes in lung fields. The tracheal outline has a slightly scalloped appearance.

FIG. 2. Section of a nodule in the submucosa showing lamellar bone arranged around a marrow cavity. H. and E., ×24.

FIG. 3. Section showing a nodule arising from the surface of a tracheal ring. No lamellar bone is present in this field. H. and E., ×50.
also islets of cartilage within the submucosa which showed foci of calcification. One of the nodules appeared to be connected with the perichondrium of a tracheal ring (Fig. 3). The mucous membrane appeared intact even over the nodules, and there was surprisingly little inflammatory reaction. Sections stained for elastic tissue by Verhoeff's technique showed no apparent abnormality in the distribution of the elastic fibres when compared with control sections from normal trachea.

AETIOLOGY

For over a hundred years, although there has been considerable speculation on the pathogenesis of this disease, its causation is still unknown. Theories advanced and now largely discarded include congenital disturbance of the tracheal connective tissue, an effect of syphilis, tuberculosis, tumours, or mechanical factors.

Chronic bronchial infection has been suggested as a factor, but, if this were so, then one would expect a relatively high incidence of tracheopathia osteoplastica in England where bronchitis is commonplace, and yet the opposite appears to be the case. Dalgaard (1947) has pointed out that inevitably and entirely coincidentally some cases will be found associated with chronic infection, but in no case has any proved pathological connexion been established between infective foci and the new formations. The virtual absence of inflammatory reaction and apparently normal mucosa in the case described lends little support to the suggestion that chronic bronchitis is an aetiological factor.

Aschoff (1910), who originally proposed the name 'tracheopathia osteoplastica', considered that the condition was a disorder of the elastic connective tissue. Dalgaard (1947), expanding this theory of metaplasia, suggested that this tissue might produce elastic cartilage with the ability to calcify and ossify by means of an osteoplastic activity. He regarded the process as originating in the undifferentiated tissue cells of the internal elastic band. His interpretation of the histological findings, especially the visible continuity of elastic threads into cartilaginous islands, has not been confirmed by Bowen (1959).

In addition, this theory is difficult to uphold in the presence of apparently normal and normally distributed elastic fibres. It would fail to explain also the absence of tuberosities on the membranous portion of the trachea where elastic fibres are prominent.

Hiebaum (1934), without producing any supporting evidence, suggested that the metaplasia was initiated by changes in the functional requirements of the tissues through ageing.

The condition is associated with no alteration in blood calcium and phosphorus levels and is not related to calcinosis of the lungs (Spencer, 1962).

Hempel and Gläser (1958), in their series of eight cases, postulated acidosis arising from metabolic or local inflammatory disturbances as a cause of hyaline swelling, which then progresses to cartilage or bone formation.

Among sclerosing tracheo-bronchopathies, Huzly (1960) considers tracheopathia osteoplastica as a distinct group, characterized by involvement of the trachea alone and never attacking the membranous portion of the wall. It produces less dense infiltration and thickening of the mucosa than the diffuse variety.

Virchow's (1863) theory that the changes are due to an ecchondrosis arising in the tracheal rings appears best to fit the facts. Indeed, we have direct microscopic evidence of an outgrowth from a tracheal ring, and Virchow suggested that where islets of cartilage or bone appear to be completely isolated in the mucosa, continuity of their connecting stalks has been interrupted as a consequence of extreme elongation and thinning.

However, by saying that the condition is an ecchondrosis, one is contributing little beyond a name to an interpretation of the cause of the process, which still remains largely unexplained.

DIAGNOSIS

The majority of reported cases have been diagnosed only post mortem, and usually the condition has not been suspected previously. However, as the bronchoscopic findings are obvious and characteristic, it is not surprising that during life the disease has been recognized by bronchoscopists. The salient features are strikingly illustrated in Huzly's endoscopic photographs.

Lell (1953), in stressing the bronchoscopic appearances, described the blunt spicule-like formations which project into the tracheal lumen, producing a grating sensation as the instrument is passed. The tracheal and bronchial walls can present a beaded appearance due to the presence of numerous sessile and polypoid nodules covered by apparently normal mucous membrane. The projections, beginning 2 to 3 cm. below the cricoid cartilage and limited to the anterior and lateral
walls, produce both narrowing and rigidity of the trachea and bronchi. The affected mucosa is often a dull greyish-red colour (Huzly, 1960).

Using local anaesthesia, Jackson and Jackson (1932) conducted weekly bronchoscopic examinations on one patient, removing large numbers of projecting spicules. In Carr and Olsen's (1954) series of seven cases, satisfactory biopsy specimens could be obtained from only three patients as the lesions were extremely hard. The tendency for biopsy forceps to slip off such hard bony projections will not surprise any bronchoscopist. Even where extensive biopsies are possible, however, very little bleeding has been reported.

AGE AND SEX INCIDENCE

According to Spencer (1962), the condition mainly affects men over the age of 50 years. The youngest case is that reported by Rode in a 12-year-old girl (Dalgaard, 1947).

RADIOLOGICAL APPEARANCES

Radiological findings in this disease are variable and largely depend on secondary lung changes produced by the bronchial obstruction. If the latter is minimal, the lung changes are correspondingly slight. Gross radiological abnormalities are likely to be associated mainly with infection in collapsed or bronchiectatic segments and will include areas of pneumonitis and fibrosis (Carr and Olsen, 1954).

It is difficult to establish a correct diagnosis of the condition by radiography alone, but tomography of the trachea or main bronchi may be helpful if it reveals the characteristic projecting nodules.

In the present case, the P.A. film demonstrates a slightly scalloped tracheal outline without narrowing. The lung fields may become obscured by pulmonary oedema resulting from left ventricular failure.

SYMPTOMS

Factors determining the degree of radiological change will play a similar role so far as symptoms are concerned. Hoarseness, cough, expectoration, and haemoptysis are likely in cases of advanced obstruction, but in many instances chest symptoms have been conspicuous by their absence.

This was so in the present case, where the cardiac and uraemic conditions were probably responsible for the terminal symptoms.

The presence of other diseases may complicate the clinical picture. For example, Dalgaard (1955) describes a case with bony and cartilaginous deposits in the upper bronchus where a bronchial carcinoma was present.

TREATMENT

No specific therapy is known, and the prognosis will depend on the degree of secondary infection. Measures should include avoidance of infection when possible and the prophylactic use of wide-spectrum antibiotics. Bronchodilator drugs and postural drainage are likely to be useful.

SUMMARY

A case of ‘tracheopathia osteoplastica’ is described.

The rarity of this disease in Britain is contrasted with its incidence in other countries. It is unlikely that the condition is being overlooked in view of the unusual macroscopic appearances, which are clearly visible at bronchoscopy and necropsy.

The pathogenesis of the disease is discussed. Ecdysonis arising from tracheal rings appears to be the most likely explanation and is supported by direct microscopic evidence in this case of an outgrowth from the perichondrium of a tracheal ring.

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REFERENCES
