Tracheopathia osteoplastica

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TRACHEOPATHIA osteoplastica is a rare condition characterized by multiple foci of submucosal calcification, cartilaginous and bony plaques projecting into tracheal lumen. Wilks, in 1857, first described it as an incidental autopsy finding. Carr & Osler (1954) reported seven, Huzley (1960) diagnosed two during routine bronchoscopy and one case each was reported by Bowen (1959) and Baird & Macartney (1966). Gilbert, Mazzorella & Feit (1953) found only one case of tracheopathia amongst forty-three of primary tracheal tumours. Although just over 100 cases are recorded in the world literature, only three have been from Britain (Wilks, 1857; Bowen, 1959; Baird & Macartney, 1966).

Tracheopathia osteoplastica forms a special group among sclerosing tracheobronchopathies (Huzley, 1960) and is recognized by the fact that pathology stops at the entrance to the main bronchi and the membranous posterior tracheal wall is never involved. Many among the reported cases of tracheopathia osteoplastica have really been the diffuse variety of the tracheobronchopathy characterized by involvement of the bronchial tree as well.

The condition mainly affects men over 50 years (Spencer, 1962) and in between the normal tracheal rings deposition of cartilage, calcium and bone takes place, giving rise to a characteristic bronchoscopic appearance. The trachea is rigid, its lumen narrowed and there are numerous yellowish white sessile, polypoid, spiky, hard cartilaginous and bony nodules projecting into tracheal lumen. The nodules, beginning 2–3 cm distal to cricoid and limited to the anterior and lateral tracheal walls, stop at the carina. There is a grating sensation as the instrument is passed down the trachea (Lell, 1953) and a small bronchoscope may have to be used to negotiate the narrow lumen. Histological examination of the nodule shows islands of cartilage and bone and the bony lamellae may even contain marrow filled with fibrofatty tissue (Huzley, 1960). The aetiology is not definitely known.

To just over 100 cases in the world literature and to the three post-mortems reported in Britain (Wilks, 1857; Bowen, 1959; Baird & Macartney, 1966), I wish to add two cases of tracheopathia osteoplastica diagnosed on routine bronchoscopy.

Case 1

C.M., a man of 65, was found to have an opacity in right lower lobe on MMR chest film in April 1966. At Bolton Royal Infirmary under the care of Dr J. B. Mitchell, he was kept under observation till February 1967 when he was transferred to us due to an increase in size of the opacity. He had been asymptomatic, had no history of cough, sputum, haemoptysis, respiratory infection, wheezing, shortness of breath or any general disorder.

Examination revealed nothing abnormal except for very early finger clubbing and dullness with poor air entry in the posterior right lower chest. BP 160/100 mmHg. The PA and lateral chest X-ray showed an opacity in the right lower lobe. Normal tracheal tomograms. On bronchoscopy the trachea presented a remarkable appearance. There were clusters of pale yellow, sessile, round and spiky stony hard nodules spread over the mucosa projecting into the tracheal lumen, an appearance somewhat resembling a rock garden (Fig. 1). The intervening mucosa was thickened and except for one or two, all nodules had complete mucosal cover. They varied in size from 0·25 to 1·25 cm, started about an inch below the cricoid and stopped abruptly at the main carinal level. The membranous posterior wall of trachea was not affected. Both major bronchi were normal; the only abnormality was crowding of the right basal bronchi. Biopsy from the latter showed normal histology. Biopsy was also obtained from tracheal nodules. Though virtually it was biting into bone, a satisfactory specimen was obtained without bleeding. Microscopic examination revealed the presence of normal mucosa containing a small mass of well-formed regular lamellar

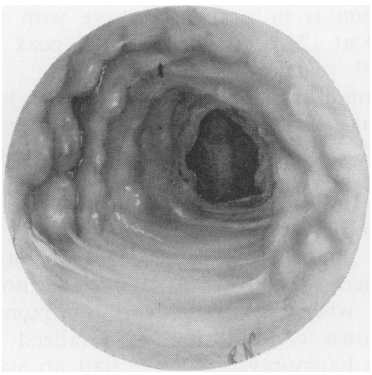

FIG. 1. Tracheopathia osteoplastica (Case 1). Diagram showing appearance of trachea at bronchoscopic examination.

cancellous bone containing fatty marrow with no evidence of any inflammatory change.

Operation. At exploratory right thoracotomy (1 March 1967) a growth was found in the right lower lobe with no glandular secondaries. The trachea, when examined from outside, was found to be normal. Basal segmental resection was done. Histology proved to be that of squamous cell carcinoma of bronchus. He was discharged on 14 March 1967.

Case 2

J.B., a 55-year-old man, was admitted because of sudden haemoptysis to the Bolton Royal Infirmary on 12 October 1962. Having had two big haemoptyses, after admission he brought up only a little blood each time he coughed and it disappeared completely after 36 hr. There was no history of previous haemoptysis, chest infection, shortness of breath, loss of weight, pulmonary or general disorder. No abnormality was discovered at clinical examination. BP 155/100 mmHg. PA and lateral chest X-rays were normal. Tomograms of the trachea in coronal and sagittal planes demonstrated irregularity of the lumen and partly calcified nodular projections situated on the left lateral wall of trachea at the level of upper border of the sternum (Fig. 2). On bronchoscopy over 100 multiple nodules varying in size and shape, greyish-white in colour and bony hard were found scattered over the tracheal mucosa. There was one much larger than the others, about the size of a big pea, situated beyond the vocal cords on the left posterolateral wall of the trachea. The posterior wall was not involved and the nodules were found extending from the upper trachea to the region of the carina. Main bronchi were normal. On attempt to take a biopsy, the nodules proved too hard to cut. No source of haemoptysis was discovered except a little blood at the base of the large nodule which was noted before any attempt at biopsy was made.

He has had no haemoptysis since, and when seen in March 1967 was well and asymptomatic.

Discussion

Several theories have been advocated, yet the pathogenesis remains uncertain. Originally it was thought to be a tumour and Rokitansky labelled a specimen in the Pathology Museum, Vienna, as showing multiple osteomata of the trachea which was later discovered to be tracheopathia osteoplastica. Neither the growth pattern nor the histology supports the tumour hypothesis. Mechanical irritation, syphilis and tuberculosis were named only to find no support as aetiological factors.

Chronic bacterial infection has also been suggested. It is quite inevitable and entirely coincidental to find an associated bacterial infection

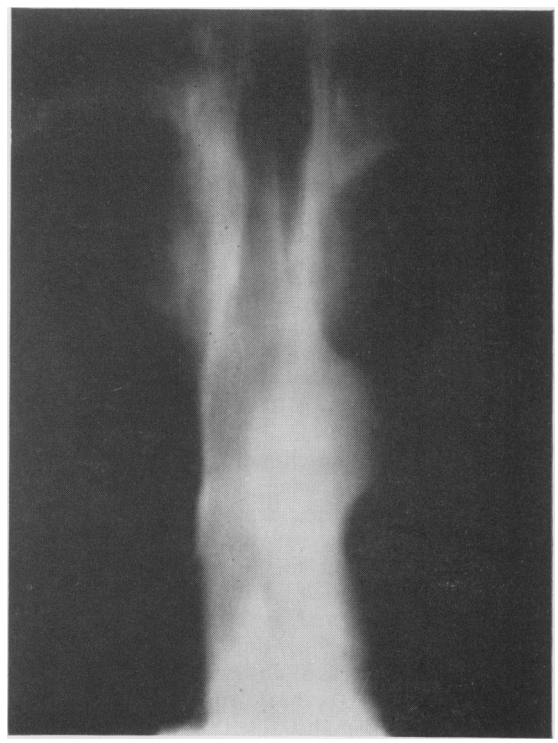

FIG. 2. PA tracheal tomogram (Case 2) showing partly calcified nodular projection on the left lateral wall and irregularity of tracheal lumen.

in some but in no case has any proved pathological connection been established between infection and the formation. If it were true one would expect to find many cases of tracheopathia in Britain where bronchitis and respiratory tract infections are quite common, yet the opposite is the case. In both these cases there had been no evidence of respiratory tract infection, the sputum grew normal bacterial flora and histological examination (Case 1) did not show any evidence of inflammation or fibrosis. All this lends little support to the infection theory.

Hampel & Gläser (1958) postulate acidosis arising from metabolic and local inflammatory disturbances as the cause of hyaline swelling which then progresses to cartilage and bone formation. There is no alteration in blood calcium and phosphorus (also true in these cases) and the condition is not related to pulmonary calcinosis (Spencer, 1962).

Aschoff (1910) considered it a disorder of the elastic connective tissue, and Dalgaard (1947), supporting this theory of metaplasia originating in the undifferentiated tissue cells of the internal elastic band, suggested that the metaplastic tissue produces elastic cartilage likely to calcify and ossify by osteoplastic activity. Hiebaum (1934) considers that the process of metaplasia is initiated by changes in the functional requirements of the tissues through the process of ageing. This theory does not explain the presence of normal and normally distributed elastic fibres on microscopic examination and the absence of nodules on the posterior tracheal wall where elastic fibres are quite prominent.

Huzley (1960) thinks that the process starts as an episodic multifocal submucosal infiltration of plasma cells and fibrinoid necrosis. In the course of time, calcification and deposition of cartilage and bone takes place. Sclerema as a possible cause is ruled out by lack of positive histological proof (Case 1) and negative latex and sheep cells tests (Cases 1 and 2).

Virchow's (1863) ecchondrosis theory is, however, the most convincing. The process starts as an ecchondrosis arising from the tracheal rings and, in due course, isolated islets of cartilage and bone come to lie in the tracheal submucosa, the continuity of their stalks connecting to the rings being interrupted as a consequence of extreme thinning and elongation. Baird & Macartney (1966) report microscopic evidence of an outgrowth from a tracheal ring in a case of tracheopathia. Also this theory explains why the posterior membranous wall is not affected.

Owing to lack of symptoms the diagnosis is usually late. Including the post-mortem cases, the distribution is in a normal curve with youngest recorded at 12½ and oldest 87, the peak being at about 50 years.

The condition is usually found as an incidental finding at necropsy (Wilks, 1857; Bowen, 1959; Baird & Macartney, 1966) or diagnosed accidentally during bronchoscopy for other reasons (Huzley, 1960). The presence of other diseases may, however, complicate the clinical picture, but in contrast to chronic bronchitis cases of tracheopathia osteoplastica have no cough, sputum, wheezing or general symptoms. The only known complication of localized tracheopathia is haemoptysis. Case 1 had no symptoms. The X-ray opacity and haemoptyses were the reasons for bronchoscopy in Cases 1 and 2 respectively. Though obviously there does not seem to be any link between the squamous cell carcinoma of the right basal bronchus as observed in Case 1, Dalgaard (1955) has also reported one case of carcinoma of bronchus arising in a tracheopathia patient. There appears to be no definite association between the two pathologies.

Although plain chest X-ray may rarely show a slightly scalloped tracheal outline (Baird & Macartney, 1966) tracheal tomograms are more likely to reveal irregularity of the lumen and thickening of the tracheal wall (Secrest, Kendig & Belord, 1964). The calcification may or may not be the radiological feature as even the largest nodule may contain only a small amount of calcium. Plain chest X-rays and tracheal tomograms were normal in Case 1. However, tracheal tomograms in Case 2 demonstrated partly calcified nodular projections and slight irregularity of the tracheal lumen (Fig. 2).

The characteristic bronchoscopic appearance has been seen in both these cases (Fig. 1). Owing to the hardness, satisfactory bronchoscopic biopsy, though very interesting, is not always possible (Carr & Osler, 1954). Even in extensive biopsies there is little if any bleeding. In Case 2 biopsy attempts failed as the nodules proved too hard to cut. Histological examination of the specimen from Case 1 showed normal tracheal mucosa, and cancellous lamellar bone containing fatty marrow.

Secondary lung changes due to bronchial obstruction are never encountered and, unlike diffuse disease where antibiotics, antispasmodics and postural drainage are indicated for prophylaxis and treatment of secondary infection, nothing is needed for tracheopathia osteoplastica, a localized tracheal condition not associated with any symptoms.

The rarity of this condition in Britain is in contrast with its incidence in other countries.

The reason remains unknown. In view of its unusual appearance it is very unlikely that the pathology is ever missed at bronchoscopy or autopsy.

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Duodenal obstruction due to an anomalous portal vein

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CONGENITAL malformations of the portal venous system are rare. Of these, the pre-duodenal position of the portal vein is of special surgical significance because of its dangerous situation in operations involving the duodenum and biliary tract, the difficulty it causes in radiological interpretation and because it can be responsible for duodenal obstruction.

We recently encountered a patient in whom duodenal obstruction from this cause produced interesting gastric symptoms.

Case report

Mr M.B., aged 29 years, was admitted to hospital complaining that he had to take two meals in quick succession. He would feel hungry within half an hour of taking the first meal and require another meal. He also complained of occasional attacks of epigastric pain following immediately after the second meal and relieved by vomiting. He had had one haematemesis 3 years previously after the ingestion of a large quantity of alcohol. There was no history of melaena or weight loss.

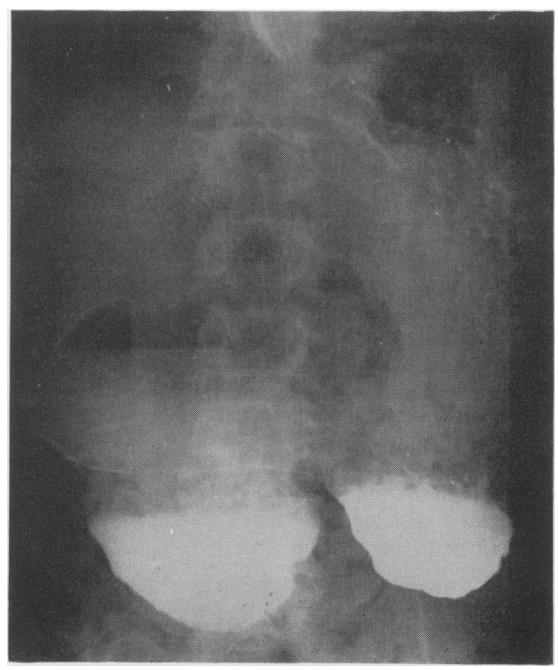

FIG. 1. Showing obstruction in the first part of duodenum and 'double bubble' appearance.

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