Tracheopathia osteoplastica

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Tracheopathia osteoplastica is a rare disease characterized by an abnormal presence of cartilage and bony tissue in the tracheal and bronchial walls. It was first described by Rokitansky in 1855 (Ragaini and Piccoli, 1957). Its incidence has been studied by several authors: Kertes and Kulka (quoted by Golémanov and Dimitrov, 1965) found three cases in 6,000 bronchoscopies and Scrobogna and Zmajevich (1961) found two in 5,230 bronchoscopies. Ragaini and Piccoli (1957), after perusal of the literature, report the incidence as three in 1,000 autopsies.

The literature contains several analyses in which the authors have tried to collect previously reported cases, reviewing them in respect to aetiology, symptoms, signs, and prognosis (Dalgaard, 1947; Ragaini and Piccoli, 1957). Among the suggested aetiological factors there are infectious metaplasia of respiratory epithelium into squamous epithelium and of submucous connective tissue into cartilage. Thereafter, the cartilage is supposed to have undergone ossification, often with formation of bone marrow. In his review Dalgaard (1947) suggested that the cartilage might have formed from the elastic fibrils in the submucosa, the fibrils continuing into the cartilage or being split by the cartilage, but this has not been confirmed by others (Bowen, 1959; Salm, 1961).

Taddel and de Wech (1962) have discussed the possible causes, viz. (*a*) inflammatory process; (*b*) mechanical action; (*c*) hormonal; (*d*) dysembryonic disease. Batzenschlager (1959) mentioned (1) systemic disease of the elastic connective tissue; (2) tumour formation; (3) dysembryonic growth, and (4) metaplasia. One author (Salm, 1961) stated that ossification with bone marrow had been found in a lipoma affecting a cardiac valve. Many believe that an aetiological relationship exists between tracheopathia osteoplastica and other diseases from which the patients have suffered, e.g. tuberculosis, syphilis, cancer of the lung, chronic bronchitis, ozoena.

The microscopic appearances were first described in 1857 by Wilks. The disease owes its name to Aschoff (1910). The first reported cases were usually found at autopsy, while during recent years many have been detected by bronchoscopy or tomography of the trachea (Ragaini and Piccoli, 1957). The lesion affects the trachea and main bronchi. In rare cases it starts in the larynx and occasionally the changes continue down

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into the finer bronchi. Bronchoscopy shows a characteristic and peculiar appearance, the surface of the trachea being reminiscent of a stalactite cave with firm, whitish nodules of varying size, narrowing the tracheal lumen. Frequently tracheopathia osteoplastica is asymptomatic. When symptoms are present they consist in dryness of the throat, a dry cough, moderate dyspnoea, and haemoptysis; frequently the disease is accompanied by pulmonary infections. The prognosis is favourable when the pulmonary infections are adequately treated. Dalgaard (1947) found the disease to be most common in the age range 30–60, with no sex difference. Ragaini and Piccoli (1957) found it to be most common in the age range 30–70, with a slight male preponderance.

After we observed a case of tracheopathia osteoplastica in our Department, we tried to supplement Ragaini and Piccoli's studies (of 187 cases) by the cases published since then. We analysed the distribution in respect to sex and age and the possible relation to other diseases. Ragaini and Piccoli's studies did not indicate any aetiological relationship to other diseases.

Case report

A male, aged 48, admitted to a chest department in 1965. During the Second World War he had been in a concentration camp where he had contracted pneumonia twice. When he came home he was found to have tuberculosis of the left lung. He attended regular follow-up for some years. Before his admission an infiltration had been found in the right lung, and the patient was put on antituberculous medication because of a suspicion of tuberculosis. He had not had any symptoms, in particular no dyspnoea worth mentioning. Bronchoscopy showed a cushiony thickening of the mucous membrane, from the subglottis and downwards; there were almost nodular prominences, extending at least ½ cm. into the lumen. The mucosa was pink, studded with small whittish, firmly adherent patches. In several sites the tissue felt very hard, and the biopsy forceps slipped off. Biopsy showed only metaplasia into squamous epithelium.

Tomography of the trachea confirmed the hypertrophic, irregular mucosal relief and narrowed lumen (Fig. 1).

Since a satisfactory diagnosis had not yet been made, another bronchoscopy was performed 2 weeks later. A biopsy specimen from the lowest part of the trachea was obtained. Microscopic examination revealed a connective-tissue stroma with infiltration by lymphocytes and plasma cells. In this stroma, several parts of the bronchial branches were coated with a normally stratified ciliated columnar epithelium; in addition, formations made up of collagen cartilage were seen. Within this collagen cartilage there were calcifications as well as changes highly reminiscent of incipient ossification with formation of bone marrow. Lastly, one of the tissue pieces showed part of a surface with normal respiratory epithelium and underneath some infiltration by lymphocytes and plasma cells. There were no definite glandular formations.

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FIG. 1.
Tomogram of trachea showing irregular tracheal outline and narrowed lumen.

No signs of specific inflammation or malignancy. Histological diagnosis: A bronchial biopsy with ossifying collagen cartilage, identical with tracheopathia osteoplastica (microscopic examination by Ch. Johansen and I. Fabricius, Bispebjerg Hospital).

The patient continued on chemotherapy for about 6 months. At follow-up 2 years later there were still no symptoms. A biopsy specimen of the nasal mucosa was taken, as some authors have demonstrated a relationship between ozoena and tracheopathia osteoplastica (Jepsen and Sørensen, 1960). Grossly, the mucous membrane was of normal appearance. Microscopic examination showed non-specific inflammation. On indirect laryngoscopy the whitish coatings of the trachea were barely visible. Tomography of the trachea was unchanged.

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In this case the patient had no symptoms of the osteoplastic tracheopathy. Apart from this disease, he was also suffering from tuberculosis. A co-existence of these two diseases has often been reported previously, but not so often in recent years.

Among the cases that we have found in the literature after 1957 (all authors of the reference list except Dalgaard (1947), Ragaini and Piccoli (1957) and Aschoff (1910)), a total of twenty-nine, the co-existing diseases have been as follows:

Tuberculosis	5 patients
Chronic bronchitis	4 patients
Asthma	3 patients
Cancer of the lung	3 patients
Ozoena	3 patients
Silicosis	1 patient
Benign chest tumour	1 patient
Heart disease	1 patient
Neurological disease	2 patients
Extra-pulmonary cancer	3 patients
No co-existing disease	8 patients

As is apparent from the tabulation, a large proportion had no other diseases; only five had tuberculosis and three ozoena.

TABLE.

AGE DISTRIBUTION AND SEX RATIO.

	0–30	31–40	41–50	51–60	61–70	71–80	81–90	?age	Total
Females	1	0	1	2	3	4	0	0	11
Males	0	3	5	2	3	4	0	1	18
Total	1	3	6	4	6	8	0	1	29

The duration of symptoms before the diagnosis was made was less than 1 year in six cases and more than 1 year in twelve. In the remaining cases no mention is made of the duration.

The diagnosis was based upon bronchoscopy in twenty cases and made at autopsy in nine. In all cases but one the diagnosis had been confirmed histologically.

In one case exploratory thoracotomy was carried out, with a fatal outcome, as the patient developed haemorrhage from the pulmonary artery. A tumour which was removed proved to consist of tracheopathia osteoplastica (Schuttleworth *et al.*, 1960).

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Conclusion

In the literature after 1957 we have found twenty-nine cases of tracheopathia osteoplastica. With our own case this makes thirty in addition to Ragaini and Piccoli's 187. Among these thirty cases the age and sex distribution was very like that found previously by Ragaini and Piccoli.

The distribution of co-existing diseases was just as motley among the thirty patients as among Ragaini and Piccoli's 187. In other words, we also found no indication that tracheopathia osteoplastica bears a special relation to any other disease.

The relation to tuberculosis mentioned by several authors may have been due to chance coincidence considering the high prevalence of tuberculosis a few years ago. In addition, the frequently asymptomatic tracheopathia osteoplastica is more likely to be diagnosed in a patient having a pre-existing pulmonary disease, as in such cases the diagnostic efforts are directed at the respiratory tract.

Summary

A case of tracheopathia osteoplastica diagnosed by bronchoscopy is reported.

The disease is rare, Ragaini and Piccoli (1957) found 187 cases in the literature, and we found another twenty-nine cases to have been reported since then.

Several authors have discussed a possible relation to other diseases.

Ragaini and Piccoli found nothing to indicate such a relationship, and our review of the literature has confirmed this.

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