

A clinical sign of adenocystic tumour of the trachea

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Singh, H. M., and Thomas, D. M. E. (1973). *Thorax*, 28, 442-443. A clinical sign of adenocystic tumour of the trachea. Three patients with intrathoracic adenocystic tumour of the trachea presented as difficult diagnostic problems. All three patients had been labelled as psychiatric problems. Present in two of the cases, the sniff sign proved diagnostic and occurred at a stage when occlusion of the trachea during expiration was almost total. In order to breathe, the patients drew in small sharp, sniffing breaths. The sign owes its name to this pattern of breathing.

Adenocystic tumour of the trachea is rare. It ranks second to squamous-cell carcinoma in primary malignant tumours of the trachea. Salm (1964), reviewing 40 cases of primary carcinoma of the trachea in the preceding decade, noted 56% of these to be squamous-cell carcinomata and 30% to be adenocystic tumours. Clark, Clagett, and McDonald (1953) described 15 patients and pointed out that surgery as against irradiation gave better results in the overall treatment of these patients. Zunker, Moore, Baker, and Lattes (1969) as well as Strobel (1966) attribute to irradiation a palliative value with temporary regression of the tumour.

The tumour is believed to grow slowly and to

have high rates of invasiveness and recurrence, and a definite but low potentiality for metastasis. Its slow growth makes it of all malignant tumours of the trachea the most amenable to surgery. Its invasiveness and late detection often contraindicate such therapy.

We present three cases (Table I) with a clinical sign thought to be pathognomonic of slow growing intrathoracic intratracheal tumours. The sign has not previously been described in these tumours.

DISCUSSION

Ranke, Presley, and Holinger (1962) point out the difficulty of diagnosis in these patients and stress that a negative chest radiograph with hacking

TABLE I

No.	Patient	Presenting Symptoms	Past History	Pattern of Breathing	Clinical Findings	FEV ₁ /VC	Radiographs	Bronchoscopy	Treatment
1	J. A. 50 yr Housewife Admitted 27 Apr. '70	Irritating, dry cough 2 yr Streaky haemoptysis 6 mth Dyspnoea 3 mth	Psychiatric treatment for last 6 mth	'Sniffing'	Absent air entry at bases	0.9/3.5	Bilateral obstructive emphysema Laminograms: intrathoracic, intratracheal tumour 6 cm above carina	Adenocystic tumour of trachea occluding lumen in expiration	Resected
2	E. L. T. 52 yr Housewife Admitted 28 June '68	Irritating cough 4 yr Streaky haemoptysis 9 mth Dyspnoea 2 yr Anterior chest pain (anginal type) 3 mth Stridor 5 mth	Treated for paranoid schizo- phrenia with ECT last 6 mth	'Sniffing'	Clubbing of fingers; expiratory wheeze	0.4/1.65	Bilateral obstructive emphysema Laminograms: intrathoracic, intratracheal tumour 2.4 cm above carina	Adenocystic tumour of trachea occluding lumen in expiration	Resected
3	C. C. 44 yr Coalminer Admitted 5 Dec. '55	Irritating cough 4 yr Dyspnoea 2 yr Streaky haemoptysis 3 weeks	Treated for psychiatric ailment for 2 to 3 yr	Not known	Inspiratory and ex- piratory stridor; absent air entry at bases	Not known	Bilateral obstructive emphysema with category III pneumo- coniosis	Adenocystic tumour of trachea oc- cluding lumen in expiration	Died before surgery

and irritating cough, wheeze, and blood-streaked sputum should be adequate for the diagnosis. However, they point out that some of the patients were treated by desensitizing procedures with various allergens before the diagnosis was made. Clark *et al.* (1953) stress that in the majority of their patients signs were present for two to four years before a diagnosis was made. As pointed out by Belsey (1950), signs of tracheal occlusion do not appear until the lumen has been narrowed by 75% or more. All three of our patients were remarkable in that they were considered to be suffering from psychiatric problems and had had several years of symptoms before a diagnosis was made.

The sign that we found characteristic of the lesion was present in the first of our two patients seen in recent years. It occurred at the stage when more than 75% of the tracheal lumen had been occluded. The patients presented with short, sharp, 'sniffing' inspirations as though 'afraid I might not be able to breathe all the air out'. Expiration at the end of a short sharp sniff seemed easier than if they had inspired a greater volume of air. The sign best described as the 'sniff sign' when associated with a wheeze is almost pathognomonic of tracheal occlusion. Bronchoscopic examination in the third patient also points to this phenomenon; the occlusion was found to be almost total in expiration but not so in inspiration. However, no note of the sign being present in this patient was recorded in 1955.

In retrospect three periods in the clinical evolution of a slow growing intrathoracic intratracheal tumour might be recognized (Table II).

TABLE II

Stage	Lumen	Respiration	Symptoms/Signs	Duration
I	Less than 75% lumen occlusion	Inspiration and expiration both normal	Tracheal irritation; hacking cough; bloodstreaked sputum	2 to 4 yr or more
II	75% lumen occlusion	Expiration gradually more difficult; inspiration normal or mildly affected	Shortness of breath; wheeze +; bloodstreaked sputum	Few months to 2 yr or more
III	Further occlusion	Expiration more severely limited; inspiration shorter and sharper; sniffing	Shortness of breath more marked; wheeze +++; sniff sign ++; x-ray obstructive emphysema	Few weeks to few months

Although the sign is a late occurrence, it represents a physiological adaptation in the pattern of breathing and should point to the diagnosis.

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