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Progressive dysphagia

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A 47 yr old male had been in good health until he was referred in September 1990. He complained of epigastric pain, nausea and vomiting and in particular eating his favourite food, potatoes, became increasingly difficult until it had become impossible. His pulmonary history was unremarkable. He was a 25 pack-year smoker and had never abused alcoholic liqours. Physical examination was unremarkable. Routine laboratory investigations revealed a clearly elevated lactate dehydrogenase (LDH) 825 $U \cdot l^{-1}$ (normal <235 $U \cdot l^{-1}$).

On the postero anterior (PA) chest roentgenogram the lateral border of the descending aorta was not visible, and within the heart shadow the contours of a mass were seen. On the lateral roentgenogram a large mass was demonstrated behind the heart (fig. 1a and b). The computed tomographic (CT) scan revealed two enlarged mediastinal lymph nodes between trachea and superior caval vein and a huge, inhomogeneous tumour, with a diameter of 11 cm, in the lower half of the middle mediastinum embracing the concentrically narrowed oesophagus and the descending aorta.

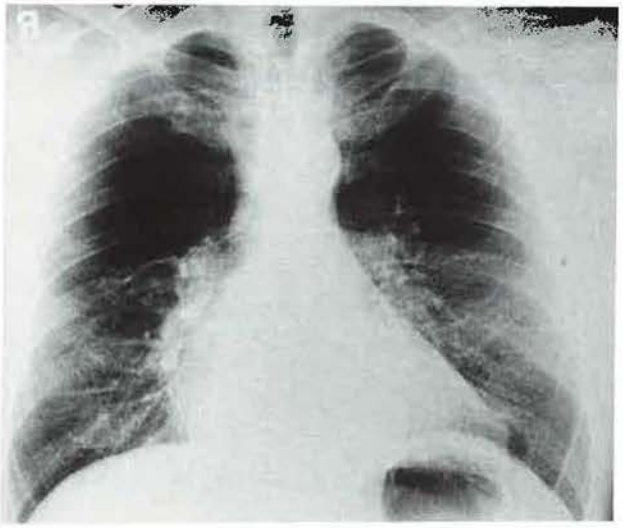
Oesophagoscopy showed concentric narrowing of the distal part of the oesophagus. The mucosa was macroscopically normal and a superficial biopsy revealed no abnormalities.

Screening for tumour markers revealed a high serum level of neuron-specific enolase.

Ultrasound of the upper abdomen as well as bronchoscopy were normal. Biopsies of the mediastinal lympnodes procured by cervical mediastinoscopy showed normal lymphoid tissue.

Isotope bone scintigraphy showed hot spots in the left hip and the first rib on the right, roentgenograms of these areas were normal. A bone marrow biopsy of the left posterior iliac crest was performed.

a



b

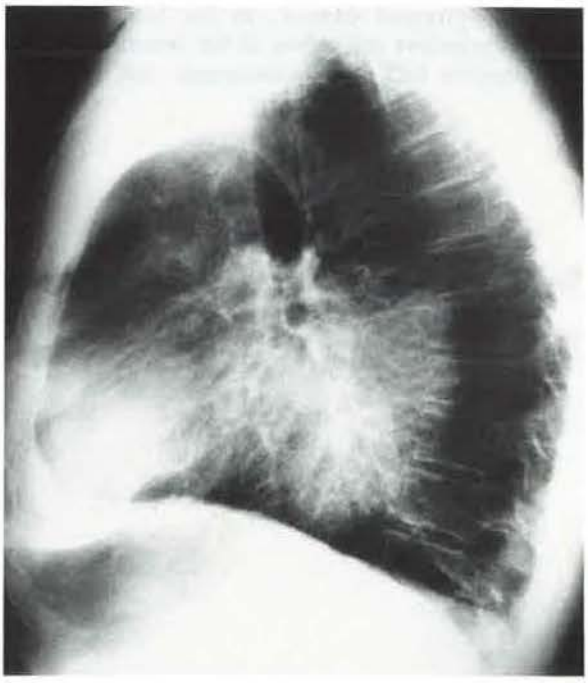


Fig. 1. – a) Postero anterior (PA) chest roentgenogram. The lateral border of the descending aorta is not visible, within the heart shadow the contours of a mass are seen. b) Lateral roentgenogram: a large mass is present behind the heart.

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TURN PAGE FOR DIAGNOSIS

Examination of the bone marrow biopsy showed small cell carcinoma.

Diagnosis: small cell carcinoma (of the oesophagus?)

Treatment with combination chemotherapy was started. Already after one week the dysphagia disappeared and roentgenologically there was an impressive reduction of the tumour size.

Discussion

Small cell carcinoma is a common pulmonary neoplasm, but it occasionally arises in extrapulmonary sites, such as the oesophagus, skin, urinary bladder, uterine cervix, trachea, gastrointestinal system, pancreas, prostate, thymus and breast [1].

Although it is impossible to prove the primary origin of the tumour, the clinical history of the patient described is highly suggestive of a primary small cell carcinoma of the oesophagus. In a recent review of 130 patients with small cell carcinoma of the oesophagus 75% complained of dysphagia, in more than half of the patients the tumour was located in the lower part of the oesophagus and at the time of diagnosis almost 75% of the patients had disseminated disease [2].

Small cell carcinoma of the oesophagus is a rather rare tumour. Its incidence in a retrospective study of 17 yrs from Memorial Sloan Kettering Cancer Centre was only 1.1% of all cases of oesophageal cancer [3].

For oesophageal cancer, as for lung cancer, a totipotent primitive cell serves as the common precursor for squamous cell, adenocarcinoma, and small cell carcinoma [4]. Its clinical behaviour is also very similar to its pulmonary analogue.

In the patient reported the elevated level of neuron-specific enolase proves the neuroendocrine character of this tumour. By this tumour marker other solid tumours and lymphoma are excluded. Together with the clinical behaviour this is diagnostic for the presence of a small cell carcinoma [5].

Therapy of this rare tumour is not well outlined and due to the low incidence prospective studies will be impossible. Regarding the similarity between pulmonary and extra-pulmonary small cell tumours, combination chemotherapy should be considered as the cornerstone of therapy for this tumour [2].

References

1. Fer MF, Levenson RM, Cohen MH, Greco FA. – Extrapulmonary small cell carcinoma. *In*: Small cell lung cancer. F.A. Greco, R.K. Oldham, P.A. Bunn eds, Grune & Stratton, New York, 1981, pp. 301–325.
2. McFadden DW, Rudnicki M, Talamini MA. – Primary small cell carcinoma of the esophagus. *Ann Thorac Surg*, 1989, 47, 477–480.
3. Nichols GL, Kelsen DP. – Small cell carcinoma of the esophagus. The Memorial Hospital Experience 1970 to 1987. *Cancer*, 1989, 64, 1531–1533.
4. Ho KJ, Herrera GA, Jones JM, Alexander CB. – Small cell carcinoma of the esophagus. Evidence for a unified histogenesis. *Hum Pathol*, 1984, 15, 460–468.
5. Carney DN, Marangos P, Ihde DC, Bunn PA, Cohen MH, Minna JD, Gazdar AF. – Neuron specific enolase: a marker for disease extent and response to therapy of small cell lung cancer. *Lancet*, 1982, i, 583–585.

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