

# Thymic carcinoid tumours

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Carcinoid tumours of the thymus are rare. The case of a 57-year-old asymptomatic man with a carcinoid tumour of the thymus, who showed a widened mediastinum by chest x-ray, is presented. Fine needle aspiration suggested the diagnosis, which was confirmed by biopsy.

**Key Words:** *Mediastinal mass; Thymic carcinoid tumour*

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## Les tumeurs carcinoïdes du thymus

**RÉSUMÉ :** Les tumeurs carcinoïdes du thymus sont rares. Le cas d'un homme asymptomatique de 57 ans atteint d'une tumeur carcinoïde, qui présentait un élargissement du médiastin aux rayons X, est décrit. Une aspiration à l'aiguille a mené au diagnostic, qui a été confirmé par biopsie.

## CASE PRESENTATION

A 57 year-old man was referred to the Department of Respiratory Medicine at the Hospital Monte San Isidro, León, Spain, because a widened mediastinum had been incidentally discovered in a chest x-ray taken during a routine medical examination. He had suffered from no serious illnesses apart from hepatitis of unknown etiology and had smoked 20 cigarettes/day for 20 years. He had no symptoms, and results of a physical examination were normal.

The hemogram and screening biochemistry results were normal, as were his levels of alpha-phetoprotein, carcinoembryonic antigen and CA 19-9. Urinary vanilmandelic acid and 5-hydroxyindolacetic acid levels were also normal.

A bronchoscopy and transbronchial pulmonary biopsy were performed, which revealed a normal bronchial tree and normal microscopy of pulmonary parenchyma, with negative cytology and cultures of the bronchial aspiration.

A computed tomography scan of the thorax, abdomen and pelvis revealed a mass in the anterior mediastinum (Figure 1) characterized by uneven and clearly defined edges; it measured 10 cm in diameter, was highly vascular and contained calcification. The mass was in contact with the heart, the ascending aorta, the arch of the aorta and the trunk of the pulmonary artery, displacing them dorsally. The pulmonary parenchyma, liver, spleen, pancreas and kidneys were normal. No adenopathies were identified.

Fine needle aspiration was suggestive of a carcinoid tumour with positive neuroendocrine markers (chromogranin and neuron specific enolase). The diagnosis was corroborated in a biopsy obtained through a left anterior mediastinotomy, in which a moderate atypia was observed (Figures 2, 3).

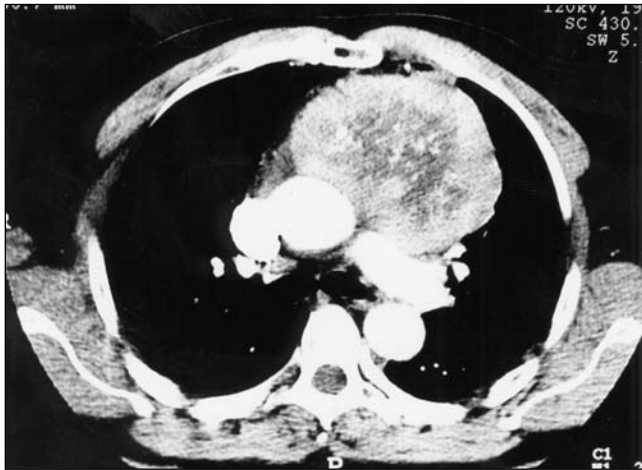
The patient was unable to undergo surgical intervention, because the great vessels were affected. At present, he is being treated with chemotherapy (cisplatin and etoposide) as an outpatient.

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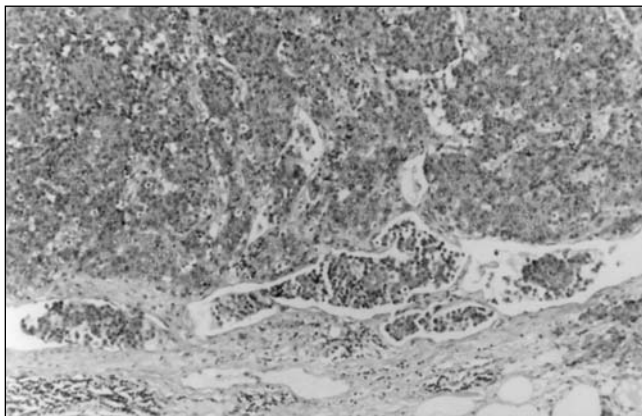
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**Figure 1)** A chest computed tomography scan showing a mediastinal mass with calcification

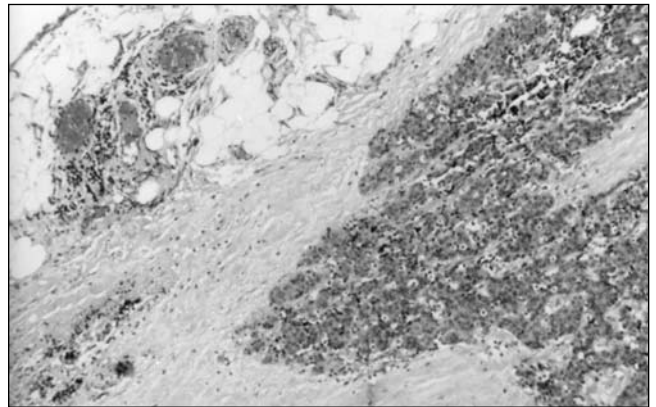


**Figure 2)** Solid ribbons and nests of small, uniform cells, with lymphatic invasion. (Hematoxylin and eosin stain, original magnification  $\times 300$ )

## DISCUSSION

Thymic carcinoid tumours are rare. Since Rosai and Higa (1) distinguished it from thymoma in 1972, slightly more than 100 cases have been described in the literature (2). This primary neoplasm, also termed primary neuroendocrine carcinoma of the thymus (3), predominantly affects males (in a ratio of 3:1) mainly between the ages of 40 and 60 years (3).

Approximately one-third of patients do not have any symptoms when diagnosed. These tumours are discovered by chance when the patients undergo x-rays of the thorax for another reason, as in the present case. Another one-third of patients present with associated endocrinopathies such as Cushing's syndrome or multiple endocrine neoplasia types I and II. Other symptoms may be present if the tumour is large, and compresses or invades adjacent structures, causing thoracic pain, dyspnea, cough and superior



**Figure 3)** Infiltration of mediastinal fat; also shown are thymic remnants. (Hematoxylin and eosin, original magnification  $\times 300$ )

vena cava obstruction syndrome (4). Rarely, it is associated with a carcinoid syndrome (5).

Classification of the tumour into three grades has been proposed: well-differentiated, moderately differentiated or atypical (the most frequent) and poorly differentiated (small cell carcinoma) (6). In this patient, the thymic carcinoid tumour was moderately differentiated.

Surgical removal is the treatment of choice for thymic carcinoid tumours, but local invasion and metastasis is frequent, even after surgery (7,8). The prognosis is poor, and if the tumour is associated with Cushing's syndrome or multiple endocrine neoplasia, the prognosis is even worse (9,10).

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