ATYPICAL PRIMARY CARCINOID TUMOR OF THE THYMUS

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Background: A 65-year-old man presented with slight cough and throat ache for one month. The complaints largely disappeared under medical treatment. Clinical examination and laboratory results showed no significant abnormalities. A conventional radiography was taken, which showed a mediastinal abnormality and several lung nodules. Additionally, a CT scan of the thorax was performed for further evaluation.

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Work-up

Chest radiography (PA view) (Fig. 1) shows widening of the upper mediastinum without displacement of the trachea. Notice the multiple lung nodules (arrows).

Contrast-enhanced CT scan of the chest (soft-tissue window) (Fig. 2, A: axial section at the level of the upper mediastinum, B: reformatted image in the coronal plane) shows on both images a large, enhancing soft tissue mass at the upper mediastinum, with invasion of the brachiocephalic vein and superior vena cava (arrows). A punctuate intratumoral calcification can also be seen on the axial image (A).

Radiological diagnosis

The CT examination was suggestive for a malignant tumor of the upper mediastinum with metastases in the lungs, which were also seen on chest radiography. Biopsy of the lesion through mediastinoscopy with subsequent pathologic examination revealed an atypical primary carcinoid tumor of the thymus.

Discussion

Thymic carcinoids are rare and can present in all age-groups. A male predominance is noticed. They may be part of multiple endocrine neoplasia syndrome type 1 (MEN 1) and approximately a third of patients present with Cushing syndrome. When symptomatic, symptoms are often secondary to mass-effect or invasion on adjacent structures of the mediastinum.

Thymic carcinoids are usually detected on chest radiographs as a mediastinal mass.

On CT scan and MRI, thymic carcinoids usually present as well-circumscribed anterior mediastinal masses, which can be partially calcified, and may cause superior vena cava obstruction. There is usually evidence of invasive disease and metastasis of the lung, liver or bone are frequently present at the time of diagnosis.

The location anteriorly in the upper mediastinum is indicative for the thymus as origin of the tumor. However, imaging findings are non-specific, and differential diagnosis is difficult. It includes other thymic masses varying from benign to malignant lesions: thymoma, thymic carcinoma, teratoma, exo- or endophytic thyroid nodule, lymphoma, adenopathy.

Thymic carcinoids should be considered aggressive neoplasms as they show invasion of adjacent mediastinal structures, local recurrence, or metastatic spread. An aggressive surgical approach in the management of thymic neuroendocrine tumor offers the best possible treatment, due to lack of effective chemotherapy or radiation therapy. Prognosis is usually poor.

Even in the cases with curative treatment, long-term follow-up is mandatory due to the aggressive nature of thymic carcinoids.

Bibliography