Cavernous sinus syndrome caused by a metastasis of a thymic carcinoid tumor

Cavernöses Sinus Syndrom verursacht durch ein metastasierendes Thymuscarcinoid

Abstract

Thymic carcinoid tumors belong to the group of mediastinal tumors, but are quite rare and usually carry a very poor prognosis. This report illustrates a case in which a thymic carcinoid tumor that had led to cavernous sinus syndrome was treated successfully. This is the first case report of a thymic carcinoid to metastasize to the cavernous sinus.

Zusammenfassung

Neuroendokrine Tumoren des Thymus, auch als Thymuscarcinoide bekannt, gehören zu den seltenen Tumoren des vorderen Mediastinums und haben der Regel eine ungünstige Prognose. Im nachfolgenden Fall wird dargestellt, dass auch ein Thymuscarcinoid, welches zu einem Cavernösen Sinus Syndrom führte, erfolgreich behandelt werden kann.

Introduction

Thymic carcinoids are very rare neoplasms that are highly malignant and aggressive [1]. They have been shown to arise from neuroendocrine cells that are normally found in the thymus.

Thymic carcinoid tumors make up only 4% of all anterior mediastinal tumors and usually occur in patients between the ages of 40 and 70. These tumors are three times more likely to affect men than women [2]. 65% of all cases are discovered incidentally during a routine physical examination.

These tumors tend to invade adjacent structures, recur locally or cause hematogenous metastases in other areas of the body. In central the nervous system, metastases of the thymic carcinoid may lead to a cavernous sinus syndrome.

Treatment of a patient with this tumor calls for prompt diagnosis, surgical treatment, and optimal adjuvant therapy [3].

Case report

A 51-year-old male was admitted to our hospital complaining of pain in the anterior chest wall and shortness of breath. The physical examination detected minimal hepatomegaly. A chest x-ray showed a nodular density at right parahilar zone (Figure 1).

Figure 1: Chest x-ray shows a well-delineated mass (arrow) in the right middle mediastinum.

A thorax CT revealed an anterior mediastinal necrotic mass sized 10 x 11 cm and multiple lymph nodes (Figure 2).
We then performed a mediastinotomy and, based on the histopathological findings, diagnosed this tumor as a thymic carcinoid tumor. Ultrasonography of the abdomen, total bone scintigraphy, and cranial computed tomography were normal. We did not see any evidence of distant metastasis.

During the sternotomy, we saw a brown, hard, calcified concentrated mass with an irregular surface that completely filled the anterior mediastinum and was attached to the medial side of both lungs, the pericardium and the vena brachiocephalica sinistra. The tumor was carefully excised together with the anterior pericardium and some lung tissue. Mediastinal lymph node dissection was also performed, followed by the appropriate closure of the sternotomy. There were no complications in the postoperative period. The patient was discharged after recovery on the 5th postoperative day.

The detailed histopathological findings confirmed the diagnosis of a thymic carcinoid tumor (Figure 3).

During the follow-up, patient complained of diplopia. An ophthalmologic examination revealed left 6th cranial nerve palsy. A cranial MRI showed a mass lesion eroding and protruding through the sella turcica and invading the cavernous sinus (Figure 4).

These findings suggested a cavernous sinus syndrome caused by cavernous sinus metastasis. We then used Cisplatin (100 mg/m²) and Etoposide (80 mg/m²) regimen (4 cycles). Patient underwent mediastinal and cranial radiotherapy. The lesion disappeared completely and the patient recovered without any further complications. He was asymptomatic during his 18 months follow-up after the operation.

**Discussion**

Thymic carcinoid tumors are rare neoplasms with a poor prognosis, especially in the event of an extrathymic invasion or lymph node metastases. They occur in all age groups, but predominantly in men. There is a frequent association with endocrine disorders. First, there is an association with Cushing syndrome due to the ectopic adrenocorticotropic hormone production, which has been reported in 20% to 35% of all patients. Second, Multiple Endocrine Neoplasia Type 1 has been reported in 25% of all thymic carcinoids [3], [4]. Our patient had no associated hormonal hypersecretion.

Most patients present with symptoms of chest pain, cough, and fatigue. Often, a routine chest x-ray has been able to show the carcinoid tumor in asymptomatic patients [5]. Thymic carcinoids vary in size from less than 2 cm to more than 10 cm in diameter. They are of soft consistency, appear grayish-white to brownish on the cut surface, and more than half of them display intraparenchymal necrosis or hemorrhages. The tumors are usually well
circumscribed, but the surrounding structures such as mediastinal fatty tissue, pleura, lung, pericardium, or vena brachiocephalica sinistra are also sometimes affected [6]. In our case, the mass measured 6 x 10 x 11 cm and had invaded the lungs, the pericardium, the vena brachiocephalica sinistra and the aortic adventitia. Chest x-rays, CTs, MRIs and PETs can be very helpful in diagnosing this type of tumor. The therapy of choice for thymic carcinoid tumors is surgery. Resection of these tumors usually includes all of the thymus and the perithymic fatty tissue. Lymph nodes should routinely be dissected. If the tumor has invaded the vena cava superior, the pulmonary vessels, or the aorta or if the surgical resection is incomplete, the diagnosis is poor [7]. An aggressive resection may include the pericardium, adherent lung tissue, mediastinal nodes, and occasionally a phrenic nerve. Complete surgical excision is absolutely necessary for a positive outcome. Even after a resection, most patients develop distant metastases. Local recurrence is also frequent, and the disease-free interval is generally short. Nevertheless, the intermediate-term survival rate is fairly good [8]. This tumor typically spreads by local invasion and metastasizes most commonly to the lung, the liver, the kidneys, the lymph nodes, and the bone and sometimes even to the central nervous system [9], [10]. Extrathoracic metastasis can be observed in 20% to 30% of all diagnosed patients. The cavernous sinus is an extremely rare metastasis site for a thymic carcinoid tumor. It may be hematogenous. In this case, a cavernous sinus syndrome may occur. The cavernous sinus syndrome may be caused by a wide range of pathological processes such as a trauma, a neoplasm, an intracavernous internal carotid artery aneurysm or a metastatic disease. The lesions within the cavernous sinus could simultaneously induce abducens nerve palsy. Patients with this syndrome experience diplopia, ptosis, and facial pain and numbness [11], [12]. We noted the cavernous sinus syndrome caused by cavernous sinus metastasis in the postoperative 8th month. He complained of diplopia. The symptom was treated and disappeared within a few days. We emphasize that this is the first case caused by a metastasis from a thymic carcinoid. The role of adjuvant or neoadjuvant radiotherapy, chemotherapy, or both has not been adequately assessed because of the low number of cases. Adjuvant therapy may have mixed short-term results, although it had little impact on long-term survival [3]. However, in our case, adjuvant radiotherapy and chemotherapy were performed successfully during the postoperative period. In conclusion, thymic carcinoid tumors have a poor prognosis because of their high degree of malignancy, early metastasis, and delayed diagnosis. Thus, treatment of a patient with this tumor calls for prompt diagnosis, surgical treatment, and optimal adjuvant therapy. Moreover, these tumors can totally be removed together with the pericardium with a low morbidity and mortality risk without requiring excessive dissection when it is difficult to remove them due to excessive adhesion to the surrounding tissue. In addition, if cranial symptoms such as diplopia are observed in patients with thymic carcinoid, the physicians should check for cavernous sinus metastasis. If a cavernous sinus syndrome is diagnosed early, treatment with radiotherapy and chemotherapy can achieve very satisfactory early and late results.

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Please cite as
Onen A, Sanli A, Karapolat S, Hayrettayg A, Aciel U. Cavernous sinus syndrome caused by a metastasis of a thymic carcinoid tumor. GMS Thorac Surg Sci. 2006;3:Doc05.

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