Primary Atypical Carcinoid Tumor of the Thymus with Parathyroid Adenoma: A Very Rare Finding

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Abstract

About 2 - 5% of the total thymic malignancies and 0.4% of total carcinoid tumors, primary thymic neuroendocrine tumors (NETs) are the zebras of the thymic neoplasms and in 25% of an association with multiple neuroendocrine neoplasia type 1 (MEN1) is reported. Thymic Atypical carcinoid are exceptional. We present a 36 years-old man with resectable thymic atypical carcinoid and parathyroid adenoma in a syndrome MEN-1.

Keywords: Carcinoid; Neuro Endocrine Tumor; Parathyroid Adenoma; NEM-1

Introduction

Representing about 2 - 5% of the total thymic malignancies and 0.4% of total carcinoid tumors, primary thymic neuroendocrine tumors (NETs) are the zebras of the thymic neoplasms. Approximately 40% of patients have paraneoplastic syndromes such as Cushing syndrome related to ACTH tumoral secretion [1]. About 25% of the NETs are in association with multiple neuroendocrine neoplasia type 1 (MEN1) [1-3]. Thymic Atypical carcinoid are exceptional.

Case Report

We present a 36 years-old man with resectable thymic atypical carcinoid and parathyroid adenoma in a syndrome MEN-1 based on positive somatic mutation test of MEN1 gene. A 36 years old man presented with 11 months history of chest pain. Negative family history of MEN1 syndrome. Physical examination was normal. There were no associated complaints of hemoptysis, cough or any significant medical or surgical history. Laboratory tests results were normal apart from: total serum calcium level 115 mg/L and PTH 162 ng/L. Radiological investigations (chest radiograph, CT and MRI) revealed a resectable mediastinal mass measuring 8 cm in its largest dimension without calcifications nore vessel invasion (Figure 1a and 1b). Parathyroid scintigraphy showed nodular fixation lower to the right thyroid lobe (Figure 2A). Physical examination, laboratory evaluation, and radiological tests of other organs revealed no significant abnormalities. Transsternal resection of mediastinum mass together with systemic lymphadenectomy was performed using a midline approach through a sternotomy (Figure 1c) that was enlarged by cervicotomy to ensure right parathyroid nodule resection (Figure 2B). Preoperatively we perform jugular blood evaluation of PTH after adenoma resection. Clips were inserted in suspected sites of mediastinal dissection. Macroscopically, the mediastinal mass was 8.5 cm × 4.5 cm with gray, soft and smooth section. Microscopically, postoperative hematoxylin and
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eosin (HE) staining showed thymic tissue, together with 2 - 4 nuclear mitosis per 10 HPF and focal necrosis. And immunohistochemistry (IHC) staining was positive for chromogranin A, CD56, CK19 and negative for CD5, CD117, S-100, CD99, CD20 with a Ki-67 index of 15%. Through HE and IHC stainings, final diagnosis of thymic atypical carcinoid was established with positive lymph node metastasis (1N+/1N) and parathyroid adenoma. Post-operative course was favorable. The patient underwent genetic tests with EXISTAT mutation of MEN-1 gene. An octresocan (A SPECT- TC with In111 Pentetreotide) noted no others sites of NEM-1 tumor. The patient refused any postoperative chemotherapy. Local radiotherapy was prescribed with 45 GY on the mediastinum and boost (15GY) of the clipped sites. Two years later the patient was symptom-free and had no clinical or radiological symptoms of relapse.

**Figure 1:**

1a: Chest CT noted the encapsulated mass of the thymic area. 1b: Chest MRI noted the thymic mass with no vessels involvement. 1c: Preoperative view of the mass dissection through a median sternotomy.

**Figure 2:**

1a: MIBI parathyroid scintigraphy view noted a right paratracheal parathyroid adenoma. 2B: Preoperative view of the parathyroid adenoma (PA).
**Discussion**

MEN-1 syndrome is a very rare incidence of 0.25% determined from postmortem studies, and an estimated prevalence of between 0.02 and 0.2 per thousand [1-3]. It is inherited in an autosomal dominant pattern. MEN-1 is characterized by hyperparathyroidism that occurs in about 90% of patients [4,5] and it is usually the first manifestation of the syndrome. Hyperparathyroidism is commonly diagnosed during the second decade of life and secondary to parathyroid neoplasm or adenoma. The most frequent islet cell neoplasm in patients with MEN-1 is gastrinoma and Pituitary tumors but carcinoid tumors can also occur more frequently [6-8]. Although they have been reported in a variety of locations, bronchial carcinoids occur more commonly in women, and thymic carcinoids occur more commonly in men such our described case. Most of non-pulmonary NETs arising in the mediastinum are of thymic origin. Patients with MEN-1 and gastrinoma who are on long term H2 blockers or proton pump inhibitors may develop gastric carcinoids [9]. A diagnosis of MEN1 can be established by any one of three criteria recommended by a consensus statement: 1) An individual with a known MEN1 gene mutation but does not have clinical or biochemical evidence of disease, 2) An individual with one MEN1-associated tumor and a first degree relative diagnosed with MEN1, and 3) An individual with at least two MEN1-associated tumors [10,11]. In patients without a family history, the diagnosis of MEN-1 requires a high level of clinical suspicion. Patients presenting with hyperparathyroidism or hypergastrinemia should be carefully questioned regarding a family history [5,7,12]. Compared with bronchopulmonary carcinoid, primary NETs of the thymus are characterized by a poorer prognosis due to their high propensity for local recurrence and earlier distant metastases [3,13]. On the other hand, one of the recent studies has shown a better prognosis of atypical thymic carcinoids as compared to pulmonary carcinoids. The clinical behavior of Atypical carcinoids varies in several studies. It has been reported that even encapsulated carcinoids bear a significant risk for recurrence, metastasis and tumor-associated death [13,15]. The role of chemotherapy is debated; in this case chemotherapy was not performed. In the literature, some cases of mediastinal NETs successfully treated by a combination of chemotherapy [13-15] or by radiotherapy alone [14] have been described. Somatostatin-analogue therapy could represent a good option in non-operable patients or in case of systemic recurrence in SSTRs (SomatoStatin Tumor Receptor) positive NETs. Hormone-expressing tumors show an unfavorable prognosis. Further, regarding the view that thymic neuroendocrine tumors are clinically more aggressive than morphologically identical neuroendocrine tumors of the lung, we also observed that primary thymic tended to exhibit less neuroendocrine differentiation.

**Conclusion**

In conclusion, thymic atypical carcinoids carry a poor prognosis however early complete surgical treatment with local radiotherapy can allow good prognosis even with lymph node metastasis. Hyperparathyroidism should be evaluated when a MEN-1 syndrome or thymic neuroendocrine tumor are suspected.

**Conflict of Interest**

There is no conflict of interest to declare.

**Bibliography**

2. Aaron Vinik., et al. "Multiple Endocrine Neoplasia Type 1".
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