Atypical carcinoid of thymus associated with multiple endocrine neoplasia syndrome type 1

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Abstract Thymic carcinoid associated with multiple endocrine neoplasia syndrome type 1 (MEN-1) is a rare tumor. We report a case of MEN-1-related thymic carcinoid. The patient reported herein had already been diagnosed with MEN-1 and was found to have a mediastinal mass. She underwent thymectomy with partial resection of the left innominate vein and lung. Histological examination revealed atypical carcinoid with infiltration. MEN-1 gene mutation was detected by employing the direct nucleotide sequencing method. Postoperative 2-fluoro-2-deoxyglucose positron emission tomography showed probable multiple metastases in the vertebrae and myocardium. However, she has been alive and asymptomatic for 2 years postoperatively. MEN-1-related thymic carcinoid is often insidious with a poor prognosis. We suggest chest computed tomography scan or magnetic resonance imaging for MEN-1 patients and serological or genetic screening for patients with thymic carcinoid to screen for MEN-1.

Key words Thymic carcinoid · Multiple endocrine neoplasia syndrome type 1

Introduction

Multiple endocrine neoplasia syndrome type 1 (MEN-1) is an autosomal dominantly inherited syndrome characterized by tumor development in the parathyroid, anterior pituitary, and endocrine pancreas or duodenum. Thymic carcinoid is also one of the tumors related to MEN-1. We here present a rare case of MEN-1-related thymic carcinoid.

Case

A 56-year-old Japanese woman was referred to our department for investigation and treatment of an anterior mediastinal mass in April 2007. Prior to this, she had been clinically diagnosed with MEN-1 and had undergone two previous operations. One was total pancreatectomy for multifocal well-differentiated endocrine tumors, and the other was resection of three parathyroid glands for hyperplasia. The right lower gland of the parathyroid was not detected during the operation. The patient was asymptomatic and had no other past history except for cholelithiasis. Her father had died of a gastric ulcer, but there was no clinical history of endocrine tumor in her family.

Laboratory tests revealed elevated values of serum hemoglobin Alc (6.8%, normal 4.3%–5.8%) and parathyroid hormone (PTH) (1960 pg/ml, normal 230–560 pg/ml), with normal functioning of the pituitary and adrenal gland. Chest computed tomography (CT) scan showed an anterior mediastinal mass close to the left innominate vein and left lung that was 40 mm in diameter (Fig. 1a). Angiography suggested that the mass slightly invaded her left innominate vein. Chest CT scan also revealed
some masses of the ventricular septum and apex of her myocardium, but their clinical significance was not recognized at that time (Fig. 1b).

In June 2007, thymectomy was carried out with partial resection of the left innominate vein and the upper lobe of the left lung through a median sternotomy. The right lower parathyroid gland was detected at the top of the right lobe of the thymus and was resected simultaneously. The tumor originated from the left lobe of the thymus, was $50 \times 40 \times 20$ mm in diameter, and showed invasion to the left innominate vein and upper lobe of the left lung. The size of the right lower parathyroid gland was $15 \times 8 \times 6$ mm. Microscopic examination confirmed the diagnosis of atypical carcinoid, well-differentiated neuroendocrine carcinoma with infiltrative growth (Fig. 2). Two satellite lesions were detected in the left lobe of the thymus.

The tumor cells had round nuclei of various sizes with fine chromatin. They were arranged in solid, ribbon-like,