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Sclerosing Mucoepidermoid Carcinoma with Eosinophilia of the Thyroid Glands

: A Case Report with Clinical Manifestation of Recurrent Neck Mass

Sclerosing mucoepidermoid carcinoma with eosinophilia (SMECE) is a recently recognized malignant neoplasm of the thyroid gland. About 14 cases of SMECE have been reported and this is the first reported case in Korea. A 57-year-old woman presented with right neck mass for 20 years. Total thyroidectomy was performed under the impression of thyroid carcinoma. The resected thyroid gland showed a poorly circumscribed hard mass. Histologically, the tumor consisted of solid nests of large atypical cells with dense fibrous stroma. The tumor cells showed squamoid appearance with abundant eosinophilic cytoplasm. There were also rare mucin-containing cells within the nests. Within the hyalinized stroma, numerous eosinophils were found. The surrounding thyroid parenchyma displayed Hashimoto's thyroiditis. There was metastasis in a regional lymph node. Two years after initial surgery, she underwent a modified radical neck dissection due to recurrent neck mass. After the radiation therapy for eight weeks, laryngectomy and esophagectomy were performed due to a recurrent carcinoma in the esophageal wall. We report an additional case of SMECE, with metastasis to regional lymph nodes and esophagus. The tumor appears to be more aggressive than previously reported and a correct diagnosis can be rendered by just examining the metastatic lesions.

Key Words: *Mucoepidermoid carcinoma, Sclerosis, Eosinophilia, Thyroid gland*

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INTRODUCTION

Although small islands of squamous epithelium may sometimes be found in various neoplastic and non-neoplastic conditions of thyroid gland, the occurrence of extensive squamous differentiation in a primary thyroid neoplasm is extremely rare (1). When a thyroid tumor exhibits prominent squamous differentiation, the diagnostic possibilities of squamous cell carcinoma (SCC), mucoepidermoid carcinoma (MEC), or sclerosing mucoepidermoid carcinoma with eosinophilia (SMECE) should be considered in the differential diagnosis. SMECE is a recently recognized low-grade carcinoma of the thyroid gland that presents a dense fibrocollagenous stroma and numerous eosinophils (2). At present, only 14 examples of this lesion have been reported in the literature (2-5), and most of those had a relatively indolent clinical course, with three patients developing distant metastasis to lung or bone or both. Herein we report another case of this condition, which showed relatively aggressive clinical course with invasion to the esophagus and lymph node metastases. To the best our knowledge, this is the first reported case in Korea.

CASE REPORT

In April 1997, a 57-year-old woman was referred to Asan Medical Center. Two years ago, the patient initially presented with a chief complaint of a painless thyroid mass for about 20 years without any treatment at Chungnam National University Hospital. The patient had undergone a total thyroidectomy under impression of thyroid carcinoma. In a resected thyroid gland, there was a poorly circumscribed hard mass, measuring 5 cm in greatest dimension. It was diagnosed as undifferentiated carcinoma with squamous differentiation. The tumor extended beyond the thyroid capsule and invaded adjacent tissue. One of five regional lymph nodes revealed metastatic carcinoma. She developed hoarseness due to paralysis of right vocal cord. However, she received no additional treatment except for thyroid hormone replacement.

Nine months after the initial surgery, a follow-up computerized tomography (CT) scan showed multiple enlarged lymph nodes along the right internal jugular vein. Deep cervical lymph node dissection was performed and there was also metastatic carcinoma in the lymph nodes. After one year, the patient developed recurrent neck mass and was referred to Asan Medical Center. On admission, she appeared chronically ill. The results of a physical examination were normal except for the surgical scar in the anterior neck and anteriorly displaced trachea. Thyroid profile tests at Asan Medical Center showed normal free T4 and low TSH level. The results of other routine laboratory studies were within normal limits and there was no peripheral blood eosinophilia. By family history, her sister had suffered from Hashimoto's thyroiditis. The patient underwent modified radical neck dissection and was diagnosed as having metastatic SMECE in the soft tissue of the neck. Subsequently, the patient received radiotherapy, total 6,100 cGY, for eight weeks. Thereafter, the patient developed difficulty in swallowing and hoarseness. The esophagography was performed and showed anteriorly displaced upper esophagus and trachea due to extensive adhesion of anterior neck. Several enlarged lymph nodes along the spinal accessory chains were also noted. The patient underwent total laryngopharyngectomy with gastric pull-up. There was metastatic SMECE in esophageal wall with extensive fibrosis. No tumor was seen in the larynx and pharynx. The patient is alive with no evidence of disease five months after the last surgery.

Grossly, the thyroid mass was ill-defined and firm, measuring 5 cm in greatest dimension. The surface was grayish-white and there was no necrosis or hemorrhage. At the periphery of the mass, there was a rim of residual thyroid parenchyma. On histologic examination, the tumor was ill-defined and consisted of small nests of atypical squamoid cells and densely hyalinized fibrous stroma. There was also numerous eosinophilic infiltration intermixed with lymphocytes and plasma cells (Fig. 1). The tumor cells were arranged in round or irregular nests or formed short narrow strands. The cells were medium to large-sized, round or polygonal in shape, and contained a moderate amount of pale eosinophilic or clear cytoplasm (Fig. 2). The nuclei were usually oval with coarse chromatin and a small but well-defined, centrally placed nucleolus. In some areas, the cells showed obvious squamous differentiation with discernible intercellular bridges and keratin pearl formation. Mucous cells were rarely present either as single cells scattered within the squamous cell nests or forming small aggregates (Fig. 3). In some areas, the tumor cells showed acantholysis and formed pseudovascular pattern. There was vascular invasion, particularly of medium-sized vessels, with luminal obliteration by the tumor cells. The non-neoplastic portion of the thyroid gland showed typical features of Hashimoto's thyroiditis (Fig. 4). The metastatic tumors in lymph nodes, soft tissue of neck and esophagus showed histological features similar to those of thyroid tumor

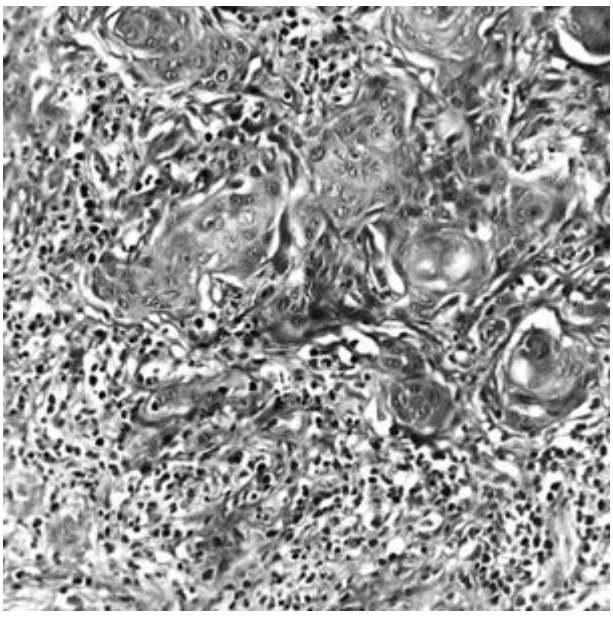

Fig. 1. The tumor consists of irregular nests of squamoid cells and dense fibrous stroma containing inflammatory cells. Tumor cells are round or polygonal and have oval nuclei and eosinophilic cytoplasms (H&E, ×200).

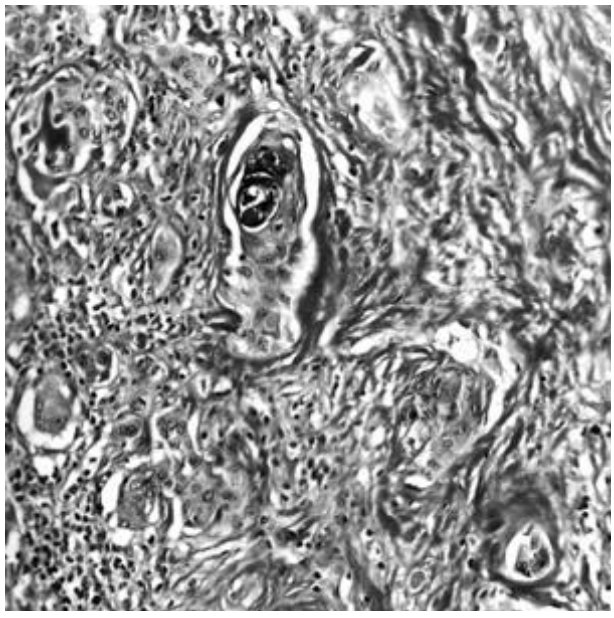

Fig. 2. There is extensive, dense fibrosclerotic stroma infiltrated by many eosinophils (lower left) mixed with lymphocytes and plasma cells (H&E, ×200).

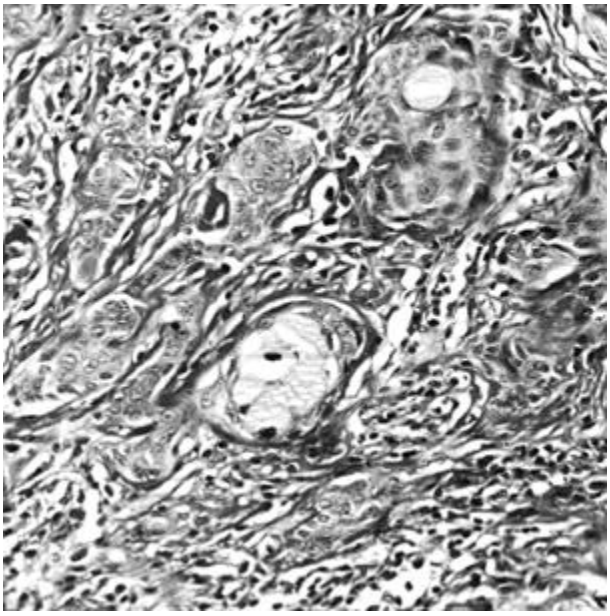

Fig. 3. Mucous cells are rarely seen either as small aggregates or single cells scattered in the squamous cell nests (H&E, × 200).

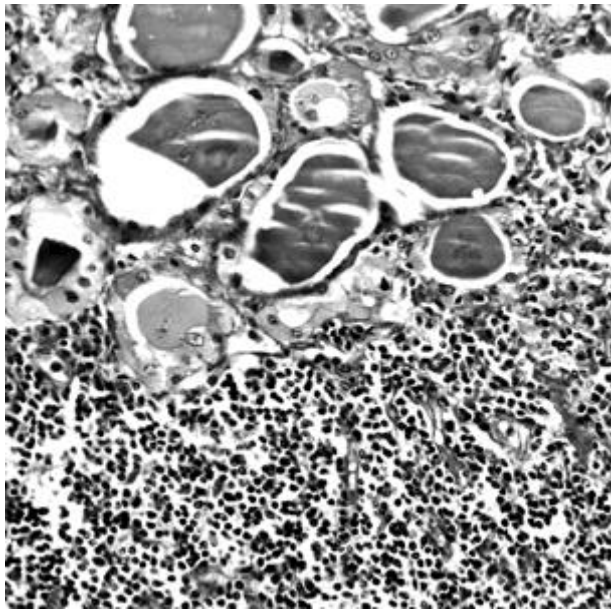

Fig. 4. Features of Hashimoto's thyroiditis are seen in the non-neoplastic portion of the thyroid gland (H&E, ×200).

including fibrosclerotic stroma and abundant eosinophils. Alcian-blue stain showed scattered mucous cells being positive for intracytoplasmic mucin, whereas squamoid cells were negative for mucin stain. By immunohistochemistry, tumor cells were reactive for keratin and CEA but no staining for calcitonin. Immunoreactivity for thyroglobulin was found in the thyroid parenchymal cells but not in the neoplastic cells.

DISCUSSION

The current case was suspected of having SMECE in the metastatic lesion in the neck because the lesion showed characteristic histologic features of SMECE originally described by Chan et al. (2). Subsequently reviewed thyroid tumor showed ill-defined tumor nests or cords with dense fibrous stroma, heavy eosinophilic infiltration, small nests or islands of squamoid cells with rare mucous cells, pseudovascular pattern of the tumor cells, and the background of Hashimoto's thyroiditis in the uninvolved thyroid tissue, which confirmed the diagnosis of SMECE in the thyroid.

On the basis of our experience and review of the literature, this tumor appears to be a histopathologically distinct entity that can be easily diagnosed by pattern recognition even in recurrent or metastatic sites (1-5).

When the squamous cellular component appears obviously malignant with cytological atypia and mitotic figures, primary or secondary SCC should be included in the differential diagnosis. Secondary involvement of the thyroid gland by SCC, either from the adjacent structure or distant organ, is far more common than a primary tumor, so that a careful search for a primary tumor elsewhere is important (6). Pure primary SCC of the thyroid gland is extremely rare. It is a rapidly growing, aggressive tumor with poor prognosis (1, 6). Pure SCC is distinguished from SMECE by its more diffuse growth in the form of sheets and large islands, much greater nuclear atypia, numerous mitoses, frequent necrosis and neutrophilic inflammatory infiltrates (1, 2). In addition, there are no mucous cells or extensive sclerosis.

The distinct morphology of SMECE strongly supports that SMECE is a separate disease entity rather than a variant of conventional MEC of the thyroid gland (2, 7-9). MECs exhibit larger and more confluent tumor nests, abundant mucous cells and show no eosinophilic infiltration and a greater tendency for lymph node metastasis and extrathyroidal extension (3). Vascular invasion with luminal obliteration of the medium-sized vessels is observed in only SMECE. Although there has been a reported case with histologically normal thyroid parenchyma (4), SMECE generally shows a classical Hashimoto's thyroiditis in the uninvloved thyroid parenchyma. However, in MEC the remaining thyroid usually shows lymphocytic thyroiditis in about a half of the reported cases. By immunohistochemistry, tumor cells in SMECE are negative for thyroglobulin, whereas focally positive in MEC.

Based on the clinical information, SMECE affects an

older female patient than MEC and represents a low-grade malignancy displaying local invasion (3). By review of the literature, follow-up of all 14 patients with SMECE showed that all patients are alive with or without evidence of tumor for periods ranging up to 12 years (3, 4). Some SMECEs behave aggressively with distant metastasis (3 of 14 reported cases). One of those patients was a male with pulmonary metastasis (4). Our case was a 57-year-old woman who also displayed lymph node metastases and local invasion into the surrounding soft tissue of the neck and esophagus. At the present time, there was no distant metastasis.

The origin of SMECE of the thyroid remains unclear. On the basis of the findings of the constant association with Hashimoto's thyroiditis with SMECE and gradual merging of the tumor cords and islands from the benign-appearing metaplastic cells of Hashimoto's thyroiditis, it is suggested that SMECE originates from the metaplastic squamous cells that are often found in Hashimoto's thyroiditis (1-3).

In conclusion, SMECE is a newly described distinct disease entity and presents characteristic clinicopathologic features. Our case is the first reported case in Korea which also emphasizes that the histologic features of SMECE are so distinctive that this tumor can be recognized even at a recurrent or metastatic site distant from the thyroid.

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