CASE REPORT
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Metastatic Hemangiopericytoma of the Thyroid Gland

Ahnsoo Na[1], Seon Kwang Kim[1], Hyun Jo Youn[1,3], Ho Sung Park[2,3] and Sung Hoo Jung[1,3]

Department of Surgery[1], Department of Pathology[2] and Research Institute of Clinical Medicine[3], Chonbuk National University Medical School, Jeonju, Korea

Hemangiopericytoma (HPC) is a mesenchymal neoplasm constituting a minor portion of vascular tumors. Although HPCs are generally benign, some progress in a malignant course, leading to recurrence and metastasis even after radical surgery. We present a case of metastatic HPC of the thyroid gland, a very rare site of occurrence. Histological findings revealed a single, well-margined mass with high cellularity. Tumor cells were tightly packed spindle-shaped to round cells arranged around numerous thin-walled vessels. Tumor cells showed cytoplasmic immunoreactivity for CD34 and CD99. To our knowledge, this is the eleventh case reported worldwide and the first case in South Korea.

Key Words: Hemangiopericytoma, Thyroid, Metastasis, Solitary fibrous tumor

Introduction

Hemangiopericytoma (HPC) is a mesenchymal neoplasm constituting a minor portion of vascular tumors.[1] It was first reported in 1942 by Stout and Murray as a distinctive vascular tumor derived from Zimmerman pericytes.[2] According to the World Health Organization classification of the soft tissue and bone tumors, the term "hemangiopericytoma" refers to a wide group of tumors sharing common characteristics such as a thin-walled branching "staghorn" vascular pattern and cellular similarities of solitary fibrous tumors.[3] The term has been used roughly to describe various neoplasms that share common morphological features. Up to 15% of soft tissue neoplasms show the characteristics of HPC focally.[4] As a result, many entities have been excluded from this ambiguous category over time. HPC is now no longer considered a specific condition, but rather as a growth pattern observed in a variety of tumors. Confusion has arisen regarding this ill-defined category, leading to difficulties in predicting clinical behavior for a given neoplasm, and thus in establishing specific treatment modalities.[4]

HPCs are generally benign in nature. However, some progress in a malignant course, leading to recurrence and metastasis even after radical surgery. Atypical features, such as high mitotic rate, high cellularity, and necrotic foci, may be observed in these malignant cases. Despite the awareness of these malignant progressions, treatment criteria and accurate prediction of prognosis are not fully established.

We present a rare case of metastatic HPC of the thyroid gland with a review of characteristics and clinicopathologic features.

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Correspondence: Hyun Jo Youn, MD, PhD, Division of Breast · Thyroid Surgery, Department of Surgery, Chonbuk National University Medical School, 634-18 Geumam-dong, Deukjin-gu, Jeonju 561-712, Korea
Tel: 82-63-250-2389, Fax: 82-63-271-6197, E-mail: yhj0903@jbnu.ac.kr

Case Report

A 40-year-old female was referred to the Department of Breast·Thyroid Surgery, complaining of discomfort due to a palpable mass on the left anterior neck. On physical examination, a 3-cm-sized, hard, fixed mass was palpated on the left thyroid. The patient had previously operated fifteen years ago at another institution for HPC of the cervical spine. The tumor was completely removed at the time. However, after a long disease-free interval, she was diagnosed with recurrent HPC of the brain (right fronto-temporal area) combined with multiple metastases of the lung, thoracic spine, and liver three years ago. She underwent brain surgery due to persistent headache and received radiotherapy afterwards. At the time of referral, she was receiving chemotherapy at the Oncology Division of Internal Medicine.

Preoperative chest computed tomography revealed a 3.3-cm-sized, well-circumscribed, round mass within the left thyroid (Fig. 1), and fine needle aspiration cytology (FNAC) result was suspicious of a mesenchymal tumor. Thyroid function tests were within normal range. Due to multiple metastases at the point of consultation for operation, left lobectomy of the thyroid was performed with palliative intent, only for cosmetic reasons and relief of neck discomfort. Parathyroid glands and recurrent laryngeal nerve were preserved.

On gross findings, there was a 3.6-cm-sized, well-defined, relatively soft mass within the left lobe of the thyroid (Fig. 2). There was no visible invasion of the neighboring tissues. Microscopic findings revealed a well-marginated, encapsulated mass with evenly distributed cellularity (Fig. 3). Tumor cells were tightly packed and arranged around numerous thin-walled vessels. They were spindle-shaped to round cells of uniform size and contained small amounts of pale or eosinophilic cytoplasm with indistinct margins. Delicate, branching vessels were accentuated. Tumor cells showed immunoreactivity for CD34, CD99, and bcl-2. They were negative for desmin, cytokeratin, S-100, Congo-red stain, EMA, SMA, synaptophysin, and chromogranin. Other findings included high mitotic rate (>10/10 HPF) and high cellularity. Histopathologic diagnosis was compatible with malignant HPC.

Afterwards, the patient continued several regimens of combined chemotherapy including anthracyclines, taxanes, and gemcitabine. However, each time the disease progressed. She is currently taking oral pazopanib, a protein kinase inhibitor. Recent exams have shown a stable disease, and her general condition is well for postoperative 15 months.

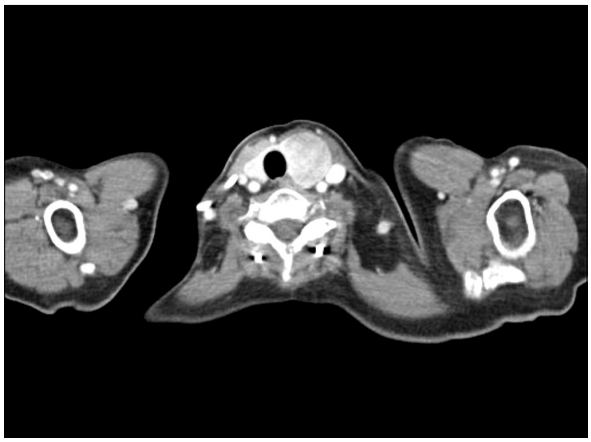

Fig. 1. Preoperative chest computed tomography. A 3.3×2.6-cm-sized, well-circumscribed, round mass was observed within the left lobe of the thyroid. The mass showed heterogeneous enhancement on contrast phase. Calcification was not observed, and there was no evidence of adjacent tissue invasion.

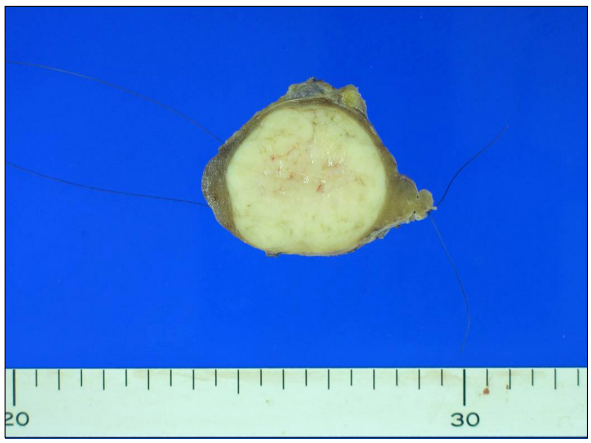

Fig. 2. Gross findings of the thyroid mass. A 3.6×3.2-cm-sized mass was observed within the left lobe of the thyroid. The mass was yellowish-white in color, relatively soft, and well-defined.

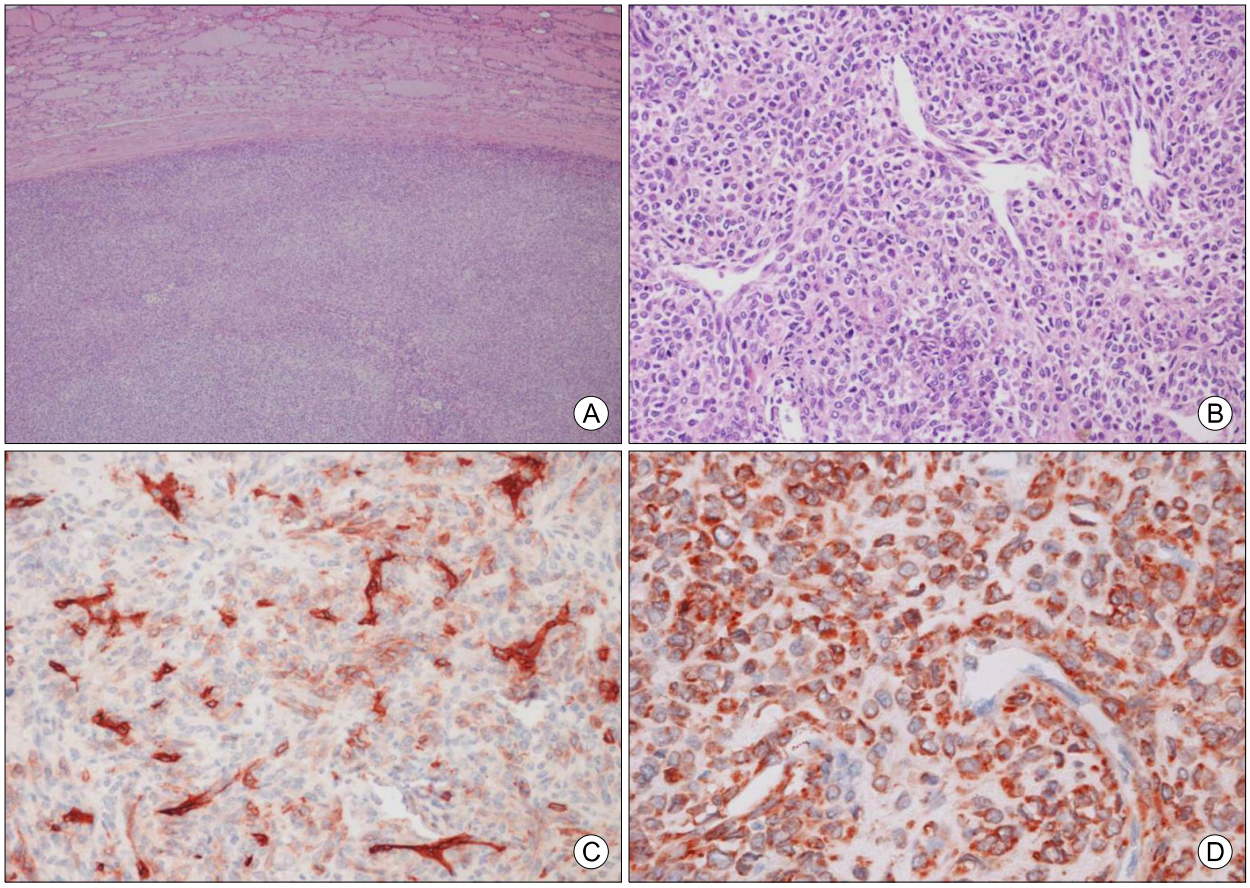

Fig. 3. Microscopic findings of the thyroid mass. (A) Low-power view of the thyroid revealed a well-marginated, encapsulated mass with evenly distributed cellularity (H & E, ×40). (B) Tumor cells were tightly packed and arranged around numerous thin-walled vessels. The cells were spindle-shaped to round and of uniform size, with small amounts of pale or eosinophilic cytoplasm with indistinct margins (H & E, ×200). (C) Tumor cells revealed cytoplasmic immunoreactivity for CD34 (CD34, ×200). Delicate, branching vessels were accentuated. (D) Tumor cells revealed cytoplasmic immunoreactivity for CD99 (CD99, ×200).

Discussion

HPC is a rare tumor where the majority of patients are adults, with a female : male ratio of 3 : 1.[5,6] The mean age of onset is 45 years. The most common site of occurrence is the lower extremities, followed by the retroperitoneum and the head and neck area.[3] In a study by Espat et al.[7], extremity tumors were mostly of the axilla and thigh, whereas the head and neck area was mainly found in the meninges and the cheek. The thyroid is a very rare site of occurrence. Cumulative data shows that it is extremely rare, with only 10 cases being reported thus far.[1] When involving the thyroid, median age of diagnosis is 46 years, with predominance in females by 3 : 2. Higher incidence is observed in the left lobe. The patient of our case was a 40-year-old female diagnosed with metastatic HPC of the left lobe. Primary sites were the head and neck area.

Clinical symptoms are not characteristic, although there may be symptoms such as pain, which occurs lately as the tumor size increases, or a palpable mass when involved superficially. Specific symptoms may arise according to the location of the tumor.

HPCs appear in variable forms, depending on the proportion of cells and fibrous stroma. Classical HPC is characterized by tightly packed round to fusiform cells arranged around thin, branching vascular structures. These vascular structures form a conspicuous "staghorn" configuration. Commonly, the vessels are wrapped by a thick coat of collagen that extends

within the interstitium.[1] When involving the thyroid, diagnosis through FNAC is very difficult, although the histopathological appearance of HPC itself is characteristic and easily diagnosed. Immunohistochemically, HPCs usually express CD34, CD99, and bcl-2, while absent for desmin and cytokeratin.[4] Microscopic findings of our patient were consistent with HPC, showing spindle-shaped to round cells of uniform size tightly packed and arranged around delicate, branching vessels. The tumor cells showed immunoreactivity for CD34, CD99, and bcl-2.

In 1976, Enzinger and Smith[8] proposed a criterion for malignancy and they suggested that large tumor size (>5 cm), increased mitotic rate (≥4 MF/10 HPF), high cellularity, nuclear pleomorphism, and focal hemorrhage and necrosis were predictors of a malignant course. In our case, the lesion showed an increased mitotic rate (>10/10 HPF) and high cellularity.

The most common sites of metastasis are the lung, bones, and liver.[7,9] Frequently, patients are present with metastases at the point of diagnosis. Previous studies reported that 11-12% of patients already had metastatic disease at the time of primary diagnosis. The sites of metastasis in our case were the lung, bones, liver and thyroid.

Despite characteristic histopathological features and easy diagnosis, there are still no universally accepted treatment criteria or prognostic criteria. The general opinion on treatment is that complete surgical resection may be helpful in gaining a favorable outcome. In previous studies, patients undergoing complete surgical excision showed a 100% median survival after 5 years.[7] Especially, lower local recurrence rates and higher disease-free survival were observed when lesions were associated with the extremities.[7,9] Other treatment modalities such as radiotherapy and chemotherapy may be considered. Our patient was treated with complete surgical resection fifteen years ago, but the disease recurred twelve years later with combined multiple metastases. She was treated with several regimens of combined chemotherapy and despite effort, the disease progressed each time. The patient is currently being treated with an oral protein kinase inhibitor, and the disease is stable thus far.

The prognosis of HPC is generally good, although local recurrence and metastatic lesions arise in some patients.[3] Espat et al.[7] reported a 5-year survival rate of 80%, with a local failure rate of 4% and metastatic rate of 20%. In a study by Spitz et al.[9], the overall 5-year survival rate was 71%, with local recurrence and metastasis occurring in 32% and 36% of patients, respectively. In a more recent, long-term follow-up study on meningeal HPCs by Shiariti et al.[10], mean overall survival was 213 months. Local recurrence rates were 90% and 32% were affected by metastasis. They proposed poor prognostic factors such as low-grade tumor, incomplete excision without postoperative radiation therapy, and metastases, although the majority was not statistically significant. In thyroidal HPC, however, prognostic factors are not established due to very rare incidence.

We have presented a rare case of metastatic HPC of the thyroid gland. Although generally considered benign, the behavior of this uncommon neoplasm is still not fully understood. Frustration may arise when clinicians encounter this unpredictable tumor, especially when multiple metastases are involved as well. Definite criteria for treatment and prognosis are necessary to ensure a disease-free interval and favorable outcome.

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