

Primary leiomyosarcoma of the thyroid

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A 56-year-old male with primary leiomyosarcoma of the thyroid is presented. The paucity of diagnostic maneuvers, including tumor markers, fine needle aspiration, and frozen section biopsy, are stressed, in addition to the fulminate course of the disease.

INTRODUCTION

Primary leiomyosarcoma of the thyroid (PLT) is rare [1]. The differential diagnosis includes medullary thyroid cancer, anaplastic thyroid cancer, and primary versus metastatic sarcoma [2,3]. Histologically, PLT is composed of characteristic spindle cells [4]. It is an aggressive malignancy with poor long-term prognosis [1-4]. We report a 56-year-old male with PLT, who presented with enlargement of the neck, hoarseness and dysphagia.

CASE REPORT

A 56-year-old male presented with 4 months history of a rapidly enlarging left anterior neck mass, hoarseness, and dysphagia. He was referred from another city with an initial diagnosis of thyroid cancer. No comorbidities existed, and the medical history was unremarkable. A history of radiation exposure did not exist, either. Physical examination revealed a hard mass of about 3 cm in the left lobe of the thyroid gland. There were no palpable cervical or supraclavicular lymph nodes. He was clinically euthyroid. Serum thyroid stimulating hormone, serum T3 (triiodothyronine) and T4 (thyroxinne) were within normal levels. Ultrasound of the neck showed a 2.71 cm × 2.59 cm solid nodule arising from the left thyroid lobe, with small contralateral nodules measuring 0.5 and 1.1 cm (Fig. 1). There was no invasion to adjacent tissues.

Fine-needle aspiration (FNA) biopsy of the thyroid mass was then performed. FNA biopsy reported malignant spindle-shaped cells. Computed tomography of the chest and abdomen showed no distant metastases. Serum calcitonin, carcinoembryonic antigen, and thyroglobulin were within normal levels. Initially, the left thyroid lobe was excised and sent to pathology for frozen section examination,

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which reported poorly-differentiated tumor suggesting a 'malignant spindle cell tumor'. Total thyroidectomy and central neck dissection were performed. No complications occurred, and the patient was discharged on the first postoperative day.

On gross examination, the tumor measured 3 cm × 3 cm × 2.5 cm. Histologically, the tumor cells infiltrated the thyroid parenchyma, with no invasion to the thyroid

capsule. Vascular invasion was present. Hematoxylin-eosin staining demonstrated spindle-shaped cell bundles. The tumor cells showed a disordered fascicular growth pattern,

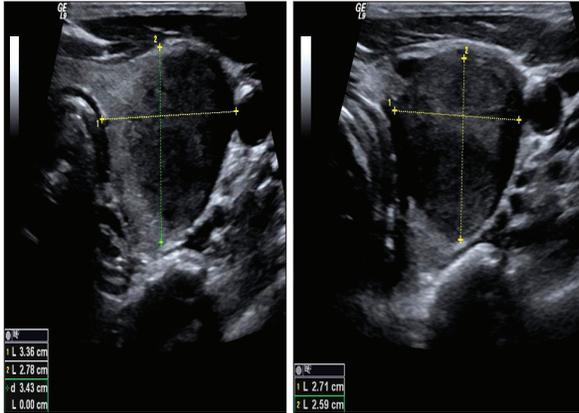


Fig. 1. Ultrasound of neck showed 2.71 cm × 2.59 cm solid nodule arising from left thyroid lobe.

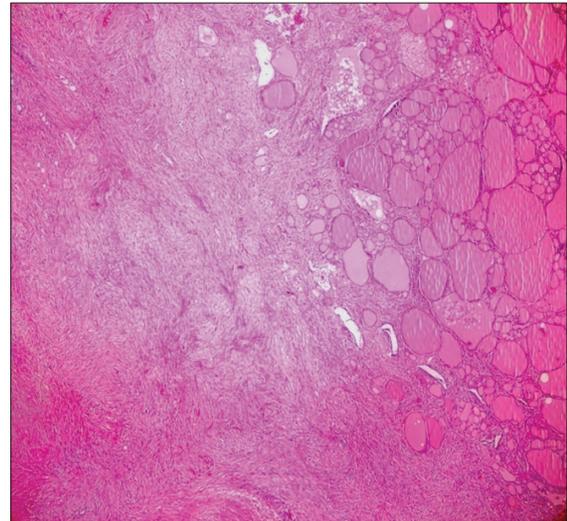


Fig. 2. Hematoxylin-eosin staining showed spindle-shaped cells with hyperchromatic, blunt-ended nuclei and abundant eosinophilic cytoplasm (H&E, ×10).

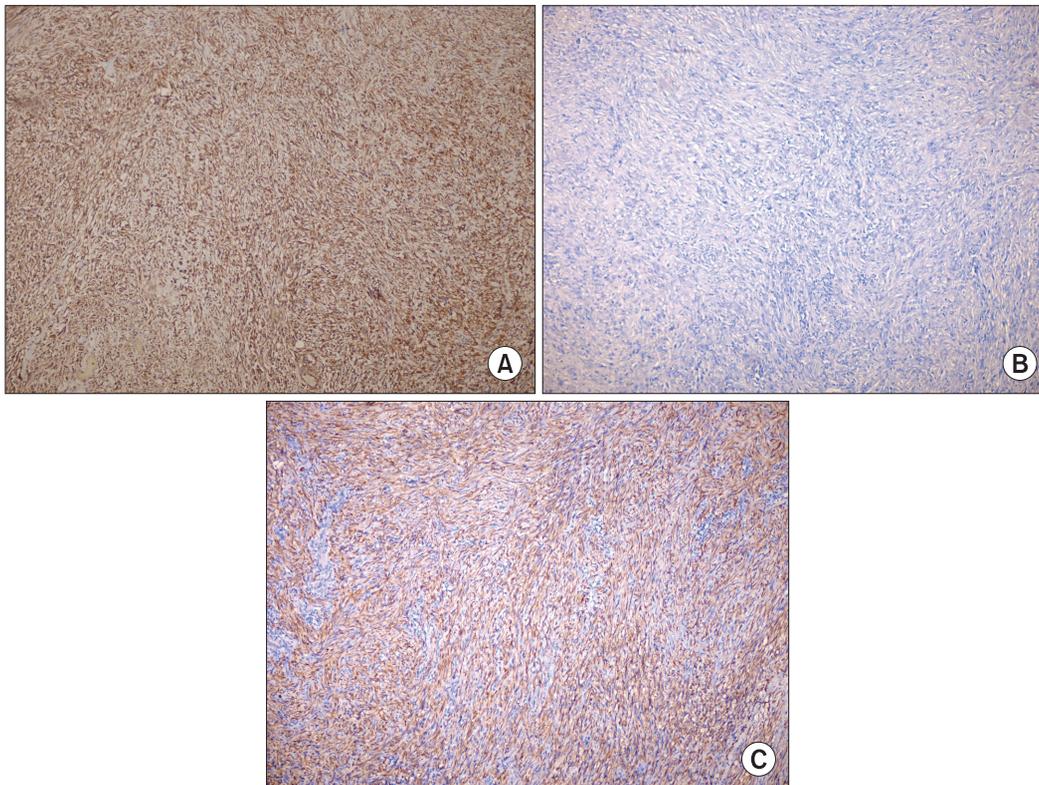


Fig. 3. The tumor was strongly positive for smooth muscle actin, positive for desmin, and negative for pancytokeratin, suggesting primary leiomyosarcoma of thyroid. (A) Positive for desmin (x10). (B) Negative for pancytokeratin (x10). (C) Strongly positive for smooth muscle actin (x10).

round or spindle configuration with hyperchromatic, blunt-ended nuclei and abundant eosinophilic cytoplasm (Fig. 2). Immunohistochemical staining with pancytokeratin, desmin, and smooth muscle actin was performed. The tumor was strongly positive for smooth muscle actin, positive for desmin, and negative for pancytokeratin (Fig. 3). No tumor-bearing lymph nodes were detected.

The oncology department did not suggest any adjuvant modalities. The patient was free of disease at three- and six-month's follow-ups. About eight months after the operation, his family doctor informed us that he had developed sudden respiratory symptoms and died in the intensive care unit with numerous pulmonary metastases.

DISCUSSION

Although the the incidence of thyroid cancer rate is different in the Korea population (the age-standardized incidence rate of thyroid cancer in 2007 was 32.8 per 100,000 [9.9 per 100,000 men and 55.6 per 100,000 women]) [5,6], thyroid cancer represents less than 1% of all malignancies in the world. The 90% to 95% of thyroid cancer cases are categorized as well-differentiated tumors arising from the follicular cells [7,8]. However, PLT gland is extremely rare, and it is the most aggressive form of thyroid cancer [2]. Only a few cases have been reported [1,3].

In our patient, FNA biopsy of the thyroid mass was the initial diagnostic step. The unusual FNA biopsy report of malignant spindle-shaped cells lead us to consider unusual tumors of the thyroid. Computed tomography of the chest and abdomen did not add to the diagnosis, similar to several tumor markers within normal ranges. The frozen section examination also failed to define the exact pathology, although the definition of a poorly differentiated tumor with malignant spindle cells weighed in favor of our suspicion of PLT. Still, possibilities such as anaplastic thyroid cancer, and primary versus metastatic sarcoma existed [1,3]. The final diagnosis could only be done by immunohistochemical staining.

Spindle cell anaplastic carcinomas resemble sarcomas. Focal keratin positivity is common in anaplastic carcinomas. Immunohistochemically, leiomyosarcomas are reactive for desmin or vimentin and actin [9]. In our patient, the diagnosis of leiomyosarcoma was favored because smooth muscle markers and desmin were positive and immunostains for keratins were negative. PLT showed smooth muscle cells with atypia and mitotic figures. Smooth muscle markers were uniformly positive, confirming the diagnosis. Leiomyosarcomas are malignant mesenchymal tumors with

smooth muscle differentiation that can arise in the smooth muscle of any organ; in the thyroid, this includes the muscular vessel walls located in the thyroid capsule [6]. Primary thyroid leiomyosarcomas tend to occur in older populations, equally in both genders. They tend to be larger than their benign counterparts with malignant histologic features characterized by pleomorphism, prominent mitotic activity, necrosis, hemorrhage, and invasive and/or extrathyroidal growth. Thyroid leiomyosarcomas are generally fatal, although follow-up was limited in the published reports [10]. Leiomyosarcomas of the thyroid can also be metastatic in origin, with primary sites in soft tissue, stomach, and pelvis, all being excluded in our patient by preoperative imaging.

The mainstay of treatment for PLT is oncologic resection, as exemplified again in this case. Chemotherapy is of limited benefit. The disease is invariably fatal, and survival rates are reported to be 5% to 10% at 1 year [3]. In spite of the silent and optimistic short-term course, our patient also experienced a fulminating and fatal tumor attack sooner than a year.

CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

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