A Case of Rhabdomyosarcoma Presenting a Rapidly Growing Thyroid Mass Showing Cytological Features Mimic Anaplastic Thyroid Carcinoma

Young Ju Jin, Hyung Gu Kim and Seong Keun Kwon
Department of Otorhinolaryngology-Head and Neck Surgery, Seoul National University College of Medicine, Seoul, Korea

This is a case of a 30-year-old man who was referred to our hospital for evaluation of a rapidly growing tumor in the left thyroid gland. It was palpated 2 weeks ago. But it was growing fast. A 4×3 cm mass was palpated in the left thyroid gland. Our impression was anaplastic thyroid carcinoma. Computed tomography finding indicated a thyroid malignant tumor and cytology result suggested spindle cell neoplasm. A lower anterior neck mass was resected and final histopathological result turns out to be a rhabdomyosarcoma (RMS). Although cervical RMS is very rare in adults, it has to be included for the differential diagnosis for the rapidly growing mass at thyroid gland.

Key Words: Rhabdomyosarcoma, Anaplastic thyroid carcinoma

Introduction

Soft tissue sarcoma is estimated less than 1% of all adult malignancy. Among them, only 3% was diagnosed with rhabdomyosarcoma (RMS), although for children, it accounts for more than 50% of soft tissue sarcoma. This neoplasm tends to occur in the head and neck (34%), genitourinary tract (14%) and chest/abdomen/pelvis (21%) of children. In adults, the proximal part of the extremities (26%) and the genitourinary site are the most common site, whereas it is affected small portion for the head and neck region (19%). Among head and neck region, RMS is mostly the embryonal type occurring in the orbit.

We experience a case of primary head and neck RMS who presented as a rapidly growing anterior neck mass which cytological differential diagnosis included an anaplastic thyroid carcinoma with rhabdomyoblastic differentiation mimicking an anaplastic thyroid malignancy. Here we report this case with a review of literatures about head and neck RMS.

Case Report

A 30-year-old man visited thyroid clinic for evaluation of anterior neck mass which was detected 2 weeks ago, and had been growing rapidly. The patient denied pain, hoarseness, dyspnea and dysphagia, He had no familial disease. There was about 3 cm palpable left thyroid soft mass without lymph node enlargement. An ultrasonography of the neck with needle biopsy was performed. The result suggested spindle cell neoplasm, reasonable finding for
RMS. But it was not included for evaluation of thyroid tissue. So, fine needle aspiration for thyroid was performed again. The result said there was a possibility of anaplastic thyroid carcinoma. He was admitted to Department of Otolaryngology to undergo an operation. A computed tomography (CT) scan of the neck demonstrated a 4 cm mass in the left thyroid without lymph node metastasis (Fig. 1). PET of the whole body revealed a left thyroid hypermetabolic lesion (SUV 3.8).

The patient underwent total thyroidectomy because the result of intraoperative frozen biopsy was hard to distinguish between RMS and anaplastic thyroid carcinoma. A 4.5×2.5×2.4 cm mass was resected (Fig. 2). The mass was adhered to the left thyroid without invasion into tracheal cartilage. After total thyroidectomy, the mass was separated easily from thyroid parenchyma. During dissection, the recurrent laryngeal nerves were saved without tumor invasion. There was no gross residual mass after thyroidectomy.

Histologic examination showed a malignant lesion composed by proliferation of spindle cells with rhabdomyoblastic differentiation (Fig. 3). On immunohistochemical study, myogenin and desmin were clearly seen in tumor cells, smooth muscle actin was focal positive (Fig. 4). This case was diagnosed histopathologically and immunohistochemically as embryonal rhabdomyosarcoma.

After surgery, the patient has replaced thyroid hor-

---

Fig. 1. CT scan showed a tumor occupying the left thyroid. The left thyroid was compressed by the tumor.

Fig. 2. Intraoperative exposure. Mass (white arrow) was very closely attached to superior and posterior portion of left thyroid gland. However, it was separated well without invasion into adjacent tissue.

Fig. 3. Pathological examination showing rhabdomyosarcoma. Numerous rhabdomyoblasts containing abundant eosinophilic cytoplasm and occasional strap cells (Hematoxylin & Eosin stain, ×400).

Fig. 4. Immunohistochemical staining showing positivity for myogenin (×200).
A Case of Rapidly Growing Thyroid Mass

mone and received 5 sessions of chemotherapy with actinomycin D and vincristine. Until now there was no evidence of recurrence.

The study was approved by the Institutional Review Board at the Seoul National University Hospital (IRB No., H-1406-011-585).

Discussion

RMS is very rare tumor for adult, presumed between 2-5% of soft tissue tumor. Five years overall survival were reported 40%.1) Histologically, RMS is classified as embryonal, alveolar and pleomorphic types. The embryonal and alveolar types occur mainly in children while pleomorphic type occurs almost exclusively in adults. The embryonal type is the most common subtype and consists of two variants, botryoid and spindle cell. The botryoid variant is defined by the presence of subepithelial aggregates of tumor cells while the spindle cell variant is characterized by spindle-shaped cells with a stroma-rich appearance. The embryonal type is mainly appearing in children under 5 years of age and the prognosis is worse in adults than in children. The alveolar type has the similar histological appearance with pulmonary alveoli, This type mainly affects children who are over 5 year of age, adolescents and young adult with poor prognosis. The pleomorphic form is rare, mainly affects adults and shows poor prognosis.3)

Adult RMS is an aggressive malignancy and is similar to other high grade soft tissue sarcomas. The favorable prognostic characteristics for survival were age under 20 years at the diagnosis, tumor size below 5 cm, absence of regional or distant disease and surgical resection with negative margins.4)

The gold standard for treatment recommended by the Intergroup Rhabdomyosarcoma Study IV (IRS-IV) is a complete surgical excision followed by chemotherapy with a combination of vincristine actinomycin-D, and cyclophosphamide. Local radiation therapy is optional.5) Radiation therapy is used as an adjuvant for incomplete tumor resection, suspicion of residual disease or tumor recurrence. Radiation is often avoided in children because it can cause severe growth abnormalities and increases the risk for a second malignancy.6)

Furze et al.7) reported a pediatric case of anterior neck RMS. There was no left thyroid tissue in the neck except mass at the preoperative CT, MRI (magnetic resonance imaging) and intraoperative finding. For that reason, the tumor regarded as malignancy could have originated from the thyroid gland.7)

For adult case, Tsuchiya et al.8) reported a huge anterior neck mass patient with lung metastasis which was recognized as thyroid tumor before operation. However, this patient was diagnosed as RMS originated near to thyroid gland after operation. He died from disease progression 1 year after the operation.

This is the second case report for adult mistaking RMS for thyroid cancer. The patient is expected to have better prognosis than previous adult case due to young age (30 vs. 61 years old), small size (4.5 vs. 9 cm) and without metastasis.

As above mentioned, age and mass size are the most important factors to predict the prognosis. For this reason, when patients visit our clinic with a rapidly growing thyroid tumor, we have to think about the possibility of RMS and operate immediately, if surgical approach is feasible.

References

6) Daya H, Chan HS, Sirkin W, Fette V. Pediatric rhabdomyosarcoma of the head and neck: is there a place for surgical