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Thyroid carcinoma with rhabdoid phenotype: Case report with review of the literature

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ABSTRACT

Objective: This paper aims to comprehensively document a rare case of thyroid carcinoma with rhabdoid phenotype and literature review of this disease.
Methods: A 59-year-old man presented with a rapidly enlarging, painful left lateral cervical mass. CT scan revealed a tumor over the left the thyroid gland with multiple left cervical lymphadenopathy over left level II–IV and level VI. Fine-needle aspiration cytology reported carcinoma, type undetermined. Total thyroidectomy with central compartment and left neck dissection was performed.
Results: Pathology report showed rhabdoid phenotype of thyroid carcinoma. Final staging was pT4N1M1.
Conclusions: Although WHO classification of thyroid tumor histology does not define this disease entity, few cases were reported. In the last 20 years, English literature review revealed only 12 cases about thyroid carcinoma with rhabdoid phenotype. Major treatment of thyroid carcinoma with rhabdoid phenotype is surgery, and the benefit of adjuvant therapies as radiotherapy or systemic chemotherapy is not clear. The prognosis of thyroid carcinoma with rhabdoid phenotype is extremely poor, with mean survival of only 6 months.

1. Introduction

The rhabdoid cell has been first described in kidney in 1978 [1] and then extrarenal origin as epithelial, endocrine, mesenchymal, and melanocytic tumor with rhabdoid phenotype were also noted later [2]. The rhabdoid phenotype is a pathological presentation associated with aggressive nature not only in thyroid gland but also in other organs [3]. In literature review, the median survival is only 6 months in thyroid tumor [4]. Here, we reported a case of poorly differentiated papillary thyroid carcinoma with rhabdoid phenotype. A discussion of rhabdoid phenotype of thyroid carcinoma was also described.

2. Case presentation

A 59-year-old male presented at our clinic with an enlarging painful left neck mass for 2 months. He also

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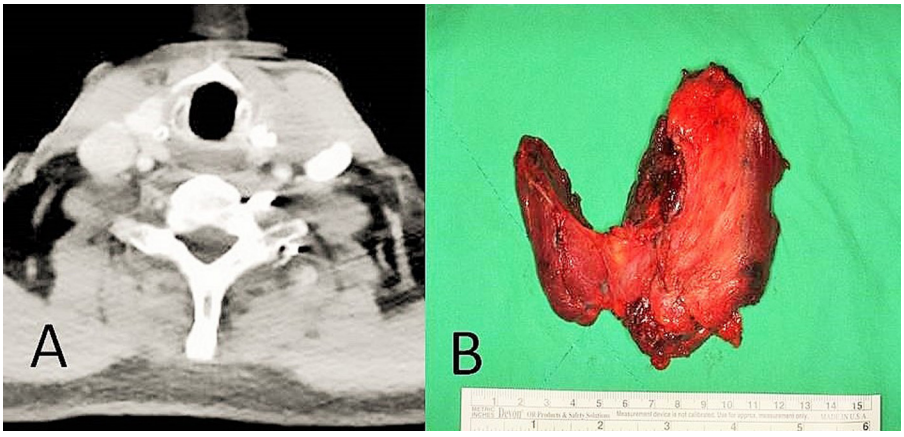

Fig. 1. Cervical CT scans and gross view of thyroid tumors. (A) A mass over left thyroid lobe with lymphadenopathy with extension to sternocleidomastoid muscle. (B) The thyroid tumor measured 8.5 cm × 7 cm × 4 cm with calcification, cystic degeneration, and extra-thyroidal extension.

experienced symptoms as hoarseness, dysphagia, and odynophagia but there was no dyspnea. There was no family history of thyroid disease, and he also denied history of radiation exposure to head and neck region. Physical examination revealed an ill-defined mass over level II–IV of left side neck. Nasopharyngoscopy revealed freely movable bilateral vocal cords without visible lesion over the larynx or pharynx. Neck ultrasonography showed a 3.7 cm × 2.2 cm hypoechoic left thyroid tumor with ipsilateral multiple enlarged lymph nodes, the largest being around 2.6 cm. Serum thyroid hormones and thyroid-stimulating hormone were within normal range. Fine-needle aspiration cytology reported carcinoma type undetermined, while computed tomography (CT) revealed left thyroid hypo-dense tumor with coarse calcification, sternocleidomastoid muscle invasion and left level II–V and VI lymphadenopathies (Fig. 1A). Total thyroidectomy, central compartment and left level II–V neck dissection were performed.

On pathological examination, the tumor measured 8.5 cm × 7 cm × 4 cm with calcification, cystic degeneration, and extra-thyroidal extension. Histologically, the tumor exhibits extensive poorly differentiated cells characterized by large cell size, abundant cytoplasm, eosinophilic inclusion, and eccentric nuclei containing distinct nucleoli compatible as rhabdoid cell (Fig. 2), mixed with focal area of conventional papillary thyroid carcinoma (PTC). Rhabdoid cell component in this case was positive for TTF-1 (Fig. 3A) and vimentin (Fig. 3C), but negative for thyroglobulin (Fig. 3B), chromogranin, HBME-1 (Fig. 3D), or chromogranin A. The DNA sequence analysis showed V600E missense mutation (T1799A) in the BRAF gene.

Post-operative PET-CT was arranged, showing multiple uptake foci over paratracheal area, left pulmonary hilum, body of T-4, right 6th rib and right iliac bone. Final staging of this case turned out to be pT4N1M1. By a multi-disciplinary conference, I-131 ablation with 200 cCi was arranged 6 weeks post-operatively. The post-therapeutic whole body scan, however, revealed no I-131 uptake at the metastatic foci. Then this patient underwent post-operative radiotherapy, and 1 year later, he lost follow-up.

3. Discussion and review of the literature

The rhabdoid cell is a special type of cell characterized by abundant cytoplasm with eosinophilic globular inclusions and eccentric with vesicular nuclei [5]. The initial concept of the rhabdoid cell has been used for a subtype of Wilms' tumor with a rhabdomyosarcomatous pattern first described by Beckwith and Palmer in 1978 [1]. In 1991, Chetty and Govender identified it in a poorly differentiated follicular thyroid carcinoma [5].

Thyroid tumors with rhabdoid phenotype are rare; only 12 cases (include this case) have been reported in recent literature (Table 1). Eight of the 12 cases were female. The patients' age ranged from 29 to 77 years with a mean of 56.4 years. All cases have extra-thyroidal extension at the time of diagnosis and cervical or distant metastases are common clinical presentations. Thyroid tumor with rhabdoid phenotype shares many features of anaplastic carcinomas, including microscopic undifferentiated cells with immunohistochemical

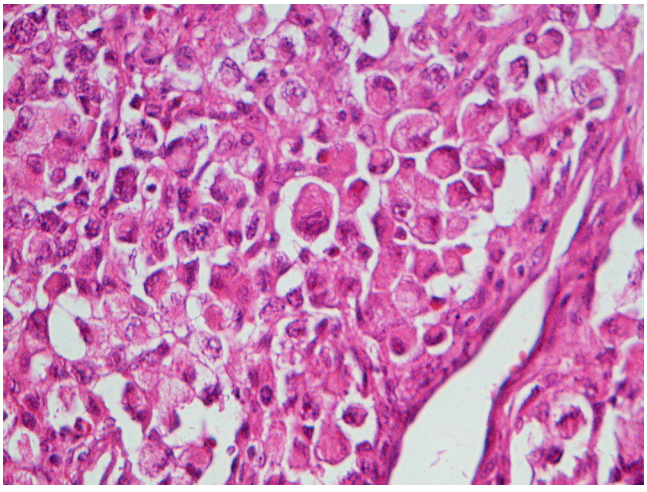

Fig. 2. Histological appearance of rhabdoid cell: large cell size, abundant cytoplasm, eosinophilic inclusion, and eccentric nuclei containing distinct nucleoli (H&E, 400×).

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