

Papillary Carcinoma of Thyroid with Rhabdoid Phenotype Transformation: Case report with Review of the Literature

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Abstract: *A rare case of anaplastically transformed papillary thyroid carcinoma with a rhabdoid phenotype in recurrence. A 45-year-old women had nodular goiter since 20 years. She had a known case of thyroid carcinoma, right thyroid lobectomy done. After 2 years later,total thyroidectomy specimen received.Histopathologic examination showed anaplastic carcinoma with papillary carcinoma with capsular and vascular invasion. The focus of papillary carcinoma was immunopositive for thyroglobin, TTF1, pancytokeratin with the area exhibiting rhabdoid features being immunopositive for vimentin. A rhabdoid phenotype is a pathological hallmark indicating the aggressive nature not only in the neck region, but also in other organs.*

Keywords: immunopositive, papillary carcinoma, rhabdoid, recurrence, vimentin

1. Introduction

Rhabdoid tumor is a very aggressive neoplasm first described by Beckwith and Palmer¹ in 1978 as a variant of renal Wilms tumor in children. Rhabdoid tumors have been established as a distinct clinicopathologic entity lacking nephrogenic and myogenic differentiation. These are highly aggressive tumors with characteristic histopathologic, immunochemical and ultrastructure features. Morphologic findings of this tumor composed of noncohesive single cells, clusters or sheets of large pleomorphic cells with abundant glassy eosinophilic cytoplasm, typical eosinophilic inclusions, and eccentric vesicular nucleus with extremely large nucleolus. Immunohistochemistry identified vimentin, sarcomeric actin, myoglobin, markers of smooth muscle lineage, neuron-specific markers, S100^{2,3} and cytokeratin expression in the tumor cells; they are negative for desmin, thyroglobin, and calcitonin; exclusion of other tumor types with rhabdoid inclusions (melanoma, other sarcoma, carcinoma). Ultrastructure findings in these cells include the presence of whorled cytoplasmic filaments⁴

Rhabdoid tumors have been described in different organs^{2,3,5,20}; they can occur at any age^{2,4} and always exhibit aggressive behavior with poor prognosis. Recently, they have been considered a morphologic expression of several types of tumors with high-grade outcome.² This paper aims to comprehensively document a rare case of thyroid carcinoma with rhabdoid phenotype and literature review of this disease. Although WHO classification of thyroid tumor histology does not define this disease entity, few cases were reported. To best of our knowledge, only 13 cases^{6,7,8,17,18,19} have been reported. In all cases, foci of follicular or papillary carcinoma were intermingled with rhabdoid component⁶⁻⁸. Most of cases rapidly developed local recurrence with cervical or mediastinal lymph nodes metastasis¹⁷.

2. Report of a Case

A 45-year-old women presented with a large, hard thyroid mass since last 20 years. Past history was right lobectomy done last 2 years back and diagnosed as carcinoma thyroid. CECT neck findings were ill-defined heterogenous lesion in remaining part of the lobe of thyroid of size 10 x 10.5 cm with multiple calcified foci of lymph nodes. Few lymph nodes were noted in the submental region. The patient had no history of chemotherapy or radiotherapy.

We received total thyroidectomy specimen contains right thyroid mass and left thyroid with isthmus along with right MRND specimens in three separate containers in histopathology sections. Right thyroid mass of size 13x13x5 cm. covered with skin, on section tumor was hard with gritty sensation, gray-white solid area of size 7x6x1.5cm, cystic areas of size 7 cm, with papillary like projection identified. Left thyroid with isthmus was unremarkable. 17 lymph nodes and a small part of parathyroid gland were identified. Histologic examination showed as majority of area of anaplastic carcinoma along with papillary carcinoma with capsular and vascular invasion of right thyroid, but lymphocytic thyroiditis of left thyroid and isthmus. Parathyroid gland and all lymph nodes, overlying skin were free from tumor. Immunohistochemical staining showed the focus of papillary carcinoma was immunopositive for thyroglobulin, TTF-1, pancytokeratin with area exhibiting rhabdoid features being immunopositive for vimentin. The area exhibiting rhabdoid features was immunonegative for pancytokeratin, thyroglobulin, TTF-1, Pax8, CK19, p53, calcitonin, chromogranin, LCA, CD138, MUM1, S100 protein. Finally we diagnosed as papillary carcinoma with rhabdoid phenotype transformation of thyroid gland. It could not be determined by histology basis only.

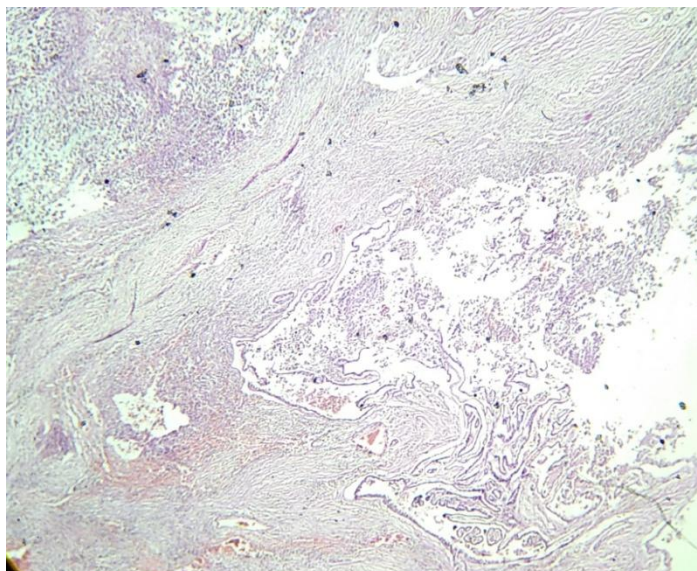
3. Discussion

Rhabdoid tumors are high-grade tumors that arise in many different sites. Tumors with rhabdoid features are considered of neoplasms derived from multiple cell types.^{2,3,5} Thyroid tumors with rhabdoid features are very rare⁶⁻⁸, our case being, to our knowledge, only few cases reported in the recent literature. Majority of the cases founded in women. The patients ages ranged from 42 to 77 years (mean age, 56.7 years). All had extrathyroidal extension of all tumor at the time of diagnosis, and all had local and distant recurrence, cervical adenopathy and distant metastasis. The disease has been uniformly rapidly lethal. These features confirm thyroid rhabdoid tumor to be a highly aggressive neoplasm with a mean survival rate similar to that of anaplastic thyroid carcinoma. Indeed, Rhabdoid tumors and anaplastic carcinoma of the thyroid exhibit similar clinical, morphologic, and immunohistochemical features. Both tumors are wholly, or partially composed of undifferentiated cells that may exhibit immunohistochemical or ultrastructural features of epithelial differentiation¹⁷. Anaplastic carcinoma usually arises by dedifferentiation of a pre-existing differentiated carcinoma.⁹ In our case report, there has been evidence of papillary carcinoma admixed with the rhabdoid component. Rhabdoid tumor of thyroid should be considered a variant of anaplastic carcinoma¹⁷.

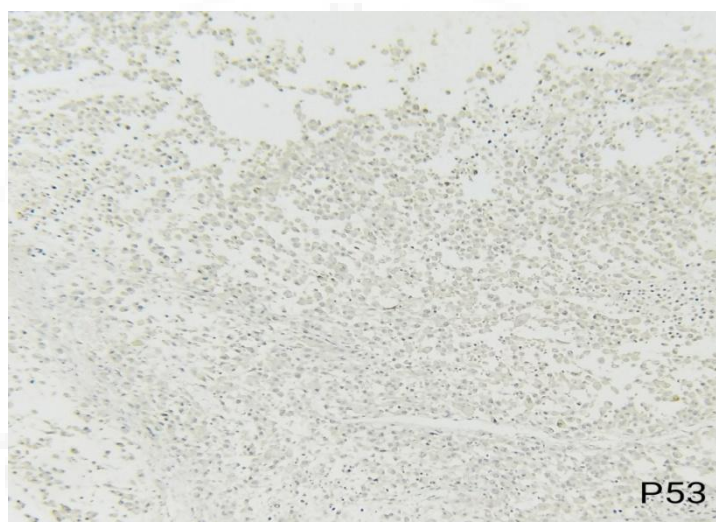
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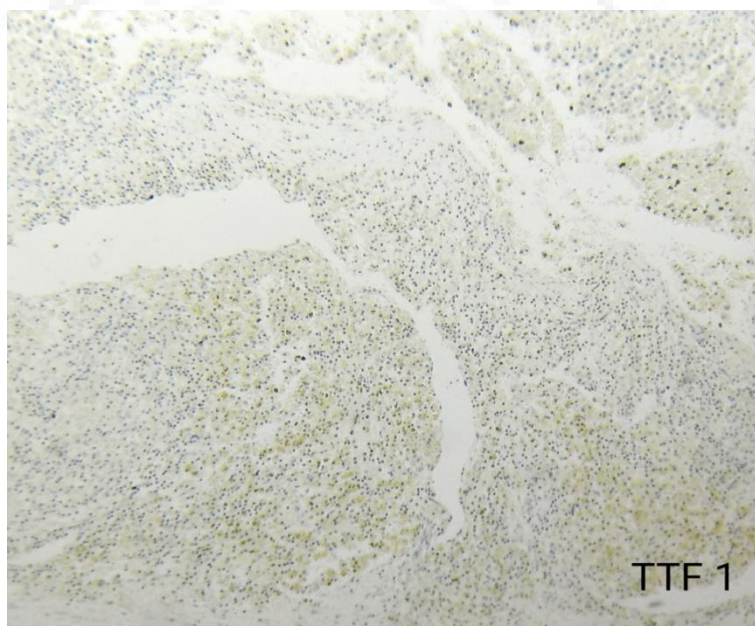
Photomicrograph



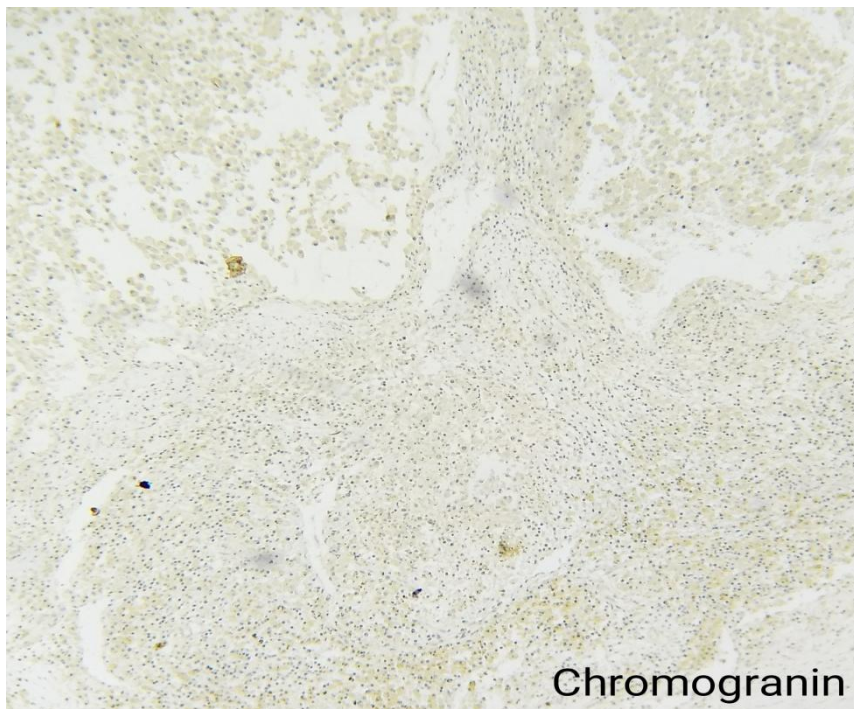
Photomicrograph of papillary carcinoma of thyroid showing rhabdoid phenotype transformation (H & E) x 100



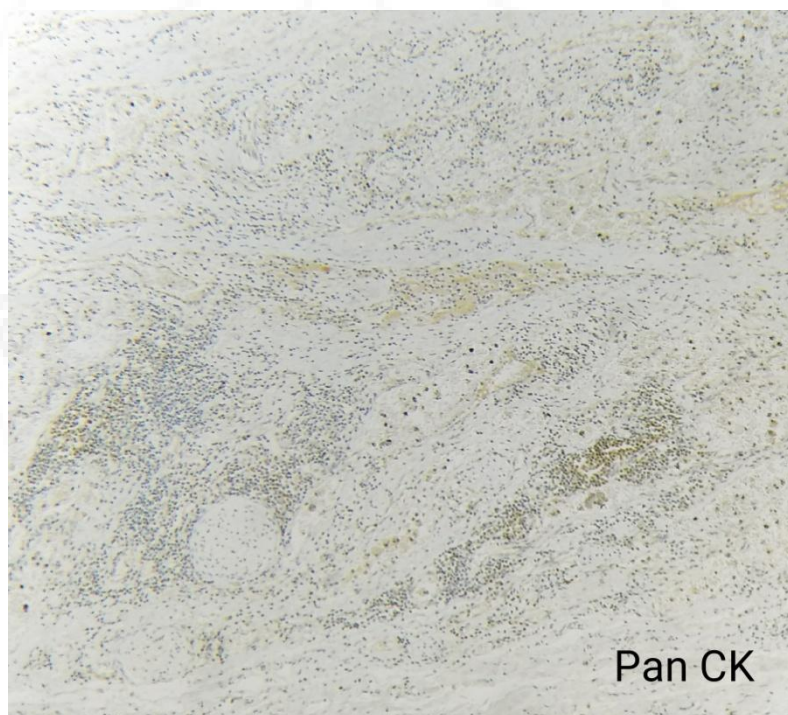
Photomicrograph of IHC showing P53 positivity



Photomicrograph of IHC showing TTF 1 Positivity



Photomicrograph of IHC showing chromogranin positivity



Photomicrograph of IHC showing Pan CK positivity