

Primary Pleomorphic Rhabdomyosarcoma of Thyroid Gland in an Adult Patient: A Case Report

Erişkin Bir Hastada Tiroid Bezinin Primer Pleomorfik Rabdomyosarkom Vakası

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Abstract

Thyroid sarcoma is a very rare entity, accounting for less than 1% of all malignant thyroid tumours. Rhabdomyosarcoma (RMS) is a sarcoma subtype, which is more common in children and adolescents. In this case, a 68-year old man, presented with hoarseness and diagnosed with pleomorphic RMS, was explored. No study of primary thyroid pure RMS has been reported in the literature, with the exception of the case reports of differentiated RMS.

Keywords: Pleomorphic rhabdomyosarcoma, adult, thyroid cancer

Öz

Tiroid sarkomu, tüm tiroid malignitelerinin %1'inden azını oluşturan oldukça nadir bir klinik antitedir. Rabdomyosarkom (RMS) çocuk ve adölesanlarda daha yaygın görülen sarkom subtipidir. Diferansiye RMS vaka sunumları hariç, primer saf tiroid RMS vakası rapor edilmemiştir. Biz ses kısıklığı ile şikayeti ile başvuran ve pleomorfik RMS tanısı konulan 68 yaşında erkek hastayı sunduk.

Anahtar Kelimeler: Pleomorfik rabdomyosarkom, erişkin, tiroid kanseri

Introduction

The most common thyroid cancers are papillary and follicular thyroid carcinomas, accounting for 90% of all thyroid malignancies [1]. Thyroid sarcomas are uncommon tumours, accounting for less than 1% of all thyroid malignancies [2]. Rhabdomyosarcoma (RMS) is a tumour of skeletal muscle that originates from embryonic mesenchymal tissue. RMS is the most common connective tissue sarcoma and accounts for 4-6% of the malignancies among children and young adults. In the paediatric population, approximately 40% of the RMSs are seen in the head and neck region [3]. RMS frequency decreases with advancing age. Pleomorphic rhabdomyosarcoma is more common in men and the median age of onset corresponds to the sixth decade of life [4-7]. In this study, we present a case of pure RMS of the thyroid gland in an adult patient.

Case Report

A 68-year old man, presented with hoarseness, referred to our clinic. There was no abnormal feature other than smoking

in his history. On the physical examination, a nodule (5x4 cm in size) was detected in the right lobe of the thyroid gland of the patient, who was otherwise healthy. A hypoechoic, solid mass lesion (47x27 mm in size) with lobulated contours was detected on thyroid sonography. Computed Tomography (CT) scan showed a mass lesion (55x40x30 mm in size) with malignant appearance, which was originated from the inferoposterior region of the right thyroid lobe and spreading to mediastinum, and which had vague demarcation with the cervical esophagus and trachea, was detected (Figure 1); then he underwent bilateral total thyroidectomy. Surgery revealed a mass (5 cm in diameter), which was invading esophagus at the posterior region of right thyroid lobe and recurrent laryngeal nerve. The surgical specimen was reported as pleomorphic rhabdomyosarcoma after pathological evaluation. Immunohistochemical evaluations revealed positive staining for vimentin and desmin, whereas negative staining for CD38, CD34, Bcl-2, thyroglobulin, TTF-1, pan-keratin, pancreatin, LCA, SMA, S100 and CD117. The number of mitoses was 4-5 at 10X magnification and the Ki67 proliferation index was 40% (Figure 2). During surgery, the mass was removed totally and surgical margins were negative. Moreover, there was

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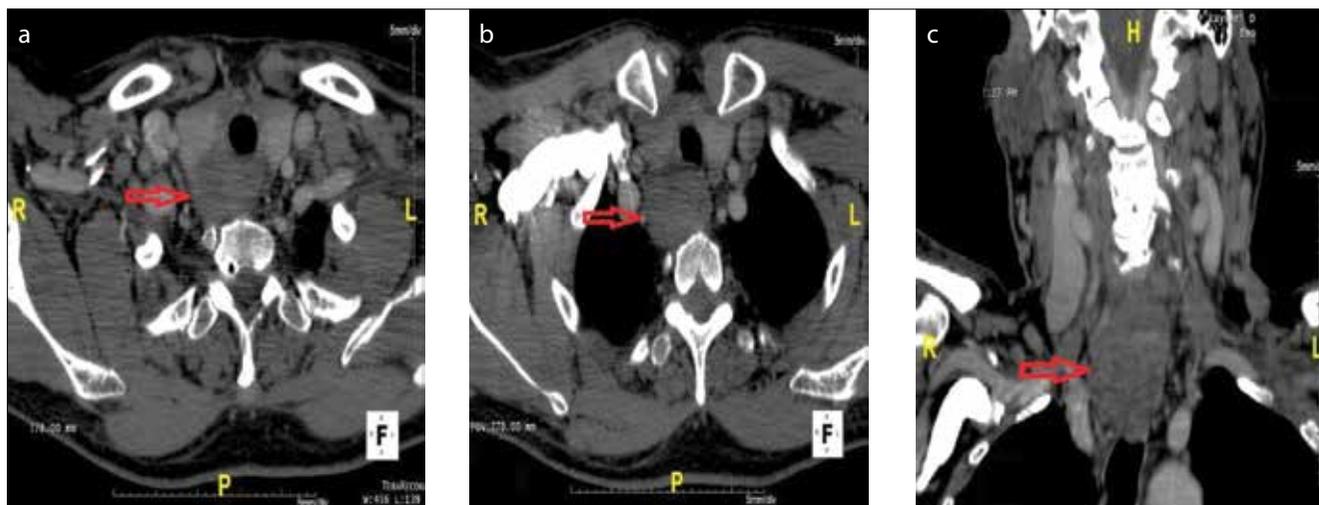


Figure 1. a-c. Tomography Scanning of the case: Axial contrast enhanced CT images (a, b) at the level of cervical trachea show a heterogeneous hypoattenuating mass in the right posterior thyroid gland. Coronal reformatted contrast enhanced CT image (c) it is demonstrated that mass extends the retrosternal space.

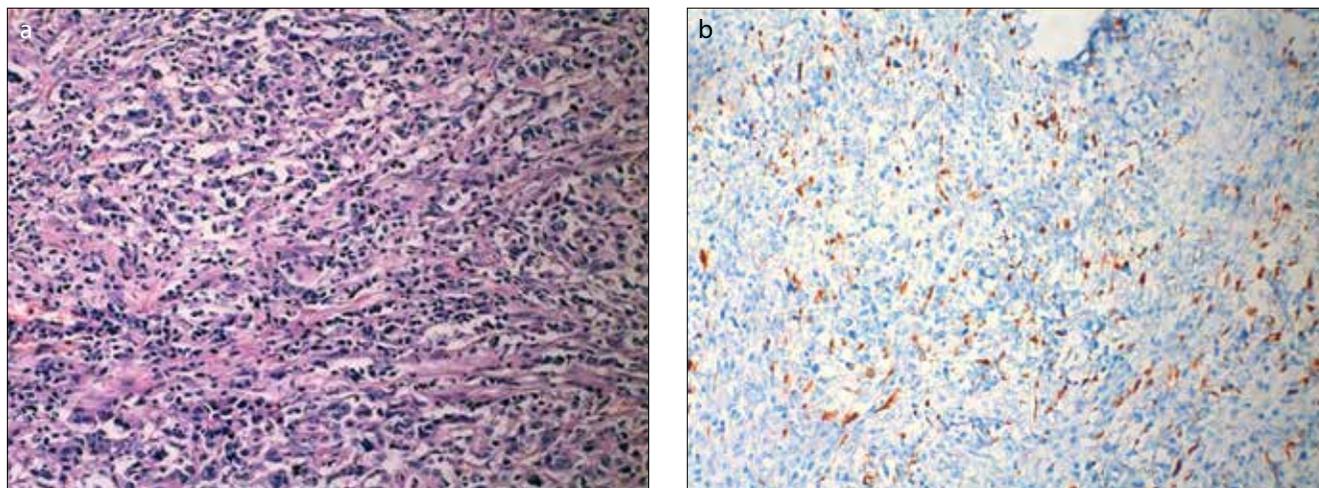


Figure 2. a, b. Histopathological and immunohistochemical imaging of the case: a) Most of the tumour cells are spindled and arranged in interlacing fascicles. The nuclei are large and pleomorphic, and mitotic figures are easily found (10x). A minor component of the tumour displays "rhabdoid" tumour cells with distinct cytoplasmic borders, voluminous pink to red cytoplasm, eccentric nuclei and prominent nucleoli (100x). b) Immunohistochemical positive staining for desmin.

no regional lymph node involvement or distant metastasis, so that, it was decided to follow-up without any additional treatment.

Discussion

RMSs constitute 3% of all soft-tissue sarcomas [8]. Traditionally, RMS has been classified as alveolar, embryonal and pleomorphic. Alveolar and embryonal variants are more commonly seen in paediatric and adolescent populations, while the pleomorphic variant is more common in adults

and in the lower limbs [5]. There is a male predominance for RMSs [6]. The identification of subtype is essential, as there is a significant difference between subtypes regarding prognosis. RMSs localized in the head and neck region are classified into 3 subtypes including orbital, parameningeal and non-parameningeal. Among them, the best prognosis is seen in orbital subtype [9].

Pleomorphic RMSs are seen commonly in middle-aged men with a median age of 54-56 years and most commonly involve the lower limbs. However, they may also involve the abdomen, retroperitoneum, chest wall, spermatic cord/tes-

Table 1. Cases of primary thyroid sarcomas in the literature

Cases	Age	Gender	Type of tumour
Griem et al. [12], 1989	23 years	Male	Radiation-induced sarcoma
Poniecka et al. [10], 2007	45 years	Female	Kaposi sarcoma
Yeo [13], 2009 (9 cases in literature)	6 th -7 th decades	Male (generally)	Fibro sarcoma
Petronella et al. [14] 2012	71 years	Male	Angio sarcoma
Chirila et al. [15] 2013	48 years	Male	Ewing sarcoma
Boudin et al. [16] 2014	55 years	Male	Synovial sarcoma
Conza et al. [11], 2014 (12 cases in literature)	6 th -7 th decades	Female (generally)	Leiomyosarcoma
Alexis et al. [18], 2005	7 months	Male	Rhabdomyosarcoma
Dutta et al. [19], 2013	7 years	Male	Rhabdomyosarcoma
Our case	68 years	Male	Rhabdomyosarcoma

tes, upper limbs, mouth and orbit more rarely. Because of the aggressive behaviour of tumour, prognosis is generally poor in RMSs [5, 6].

Up to date, the reported cases of thyroid sarcomas in the literature are as follows: Kaposi sarcoma, leiomyosarcoma, radiation-induced sarcoma, fibro sarcoma, angio sarcoma, Ewing sarcoma and synovial sarcoma [10-16]. However, they were not large series but only case reports. Primary RMS of the thyroid gland is a truly rare malignancy. Up to now, only 4 cases have been reported, these include 2 cases with anaplastic thyroid carcinoma exhibiting RMS differentiation [17] and 2 cases who were child, in whom thyroid gland tissue was detected within anterior cervical tissue and diagnosed as RMS [18, 19] (Table 1). These tumours also exhibited the characteristics of aggressive tumour by invading the surrounding tissue as in our case. RMS generally presents with bulky mass and compression of the surrounding tissues. In a study, it was reported that RMS was detected in one of 77 children, in whom the etiology of thyroid nodules was evaluated [20]. In addition, Hafez MT et al. have reported a metastasis to the thyroid gland from RMS originated from lower limb as a case [21]. To the best of our knowledge, there is no previous report about primary pleomorphic RMS in an adult patient.

Rhabdomyosarcomas generally have a very high mitotic rate, explaining the aggressive nature and poor prognosis of these tumours. Histologically, small blue cells are seen in pleomorphic RMSs and positive expression of desmin, myogen and myoD1 are most frequently seen [22]. Differential diagnosis includes anaplastic thyroid carcinoma with rhabdoid variant due to the presence in each variant of spindle cell elements. The majority of anaplastic (undifferentiated) thyroid tumours show 'sarcoma-like' features, with spindle-

shaped neoplastic cells arranged in a fascicular or whorled pattern of growth. Immunohistochemical positive staining for keratins, confirm the epithelial nature of the tumour [17]. Immunohistochemical evaluations revealed positive staining for vimentin and desmin, whereas negative staining for pan-keratin thus, rhabdoid variant of anaplastic thyroid carcinoma was excluded as in our case.

The imaging has no specific feature in RMS. On CT scan, it is similar to skeletal muscle and there is increased attenuation. However, imaging is usually helpful in staging as 44% of RMS cases have metastasis at the time of diagnosis. On MRI, it appears as homogenous mass, which has increased the signal intensity, when compared to muscle and fat, on T1-weighted images while iso-intensity or minimal hyperintensity could be detected, when compared to muscle, on T2-weighted images [4].

In conclusion, the thyroid gland can now be added to the list of infrequent involvement sites of pleomorphic RMS such as the upper limb, mouth, orbit, spermatic cord/testes, abdomen, retroperitoneum and chest wall. This could be translated as novel sites of involvement by pleomorphic RMS that will appear in the future.

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