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Case Report

Non-Alpine Epithelioid Angiosarcoma of Thyroid Gland: A Rare Clinical Scenario

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Abstract

Malignant sarcomatous tumors of thyroid gland of vascular endothelial origin are uncommon. Moreover, epithelioid variety of angiosarcoma of thyroid gland is even rarer and exhibit relatively aggressive oncological behavior especially in Non-Alpine world. Here we report such a case of the thyroid neoplasm which proved to be angiosarcoma of thyroid gland on final immunohistological analysis and was treated with total thyroidectomy followed by adriamycin based chemoradiation. This tumor accours very rarely in Non-alpine region and to the best of our knowledge only 19 cases of such a variety have been previously reported in the literature in form of case reports. We also give a brief review of the literature.

Keywords: Angiosarcoma; Thyroid Gland; Thyroidectomy

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Introduction

Angiosarcoma of thyroid gland is a rare but aggressive malignant neoplasm which is infrequently diagnosed in 2-10% of Austrian, Swiss and Northern Italian thyroid malignancies [1-3]. The exact etiology of this demographic distribution is unknown however iodine deficiency induced long-standing goiter may be one plausible cause [4]. Epithelioid variety of angiosarcoma in non-Alpine areas is even rarer incidence [1-5]. Older females are more likely to be affected [1-6]. Cytological analysis is usually non-diagnostic [7]. Even its histological differentiation from poorly differentiated cancers can be challenging and immunohistochemical analysis could only be diagnostic. These tumors are poorly encapsulated and tend to involve surrounding structures more readily and have higher recurrence rates. Distant metastasis remains a poor prognostic indicator [1, 2]. Regional lymph nodes and lungs are the most commonly involved distant sites [2]. Adriamycin based chemotherapy following en-block resection is the only treatment of described. Role of radiotherapy still remains undetermined [1, 2].

Here we describe such a case with Stage IV metastatic epithelioid angiosarcoma of thyroid gland with brief review of the literature.

Case Presentation

49 year old patient with maturity onset diabetes mellitus presented with solitary nodule of left lobe the thyroid gland for last 5 months. Ultrasound documented a 55 mm x 50mm x35 mm nodule in the left lobe of the thyroid gland. An additional nodule sizing 12mmx20mm was noted in Rt. Lobe of thyroid gland. Thyroid scan showed reduced tracer uptake in the left lobe nodular region. FNAC of the cold nodule showed follicular cells with high grade cellular atypia suggesting a follicular lesion. The patient 2 months earlier also developed hemoptysis associated with right sided pleural effusion for which he was worked up by cytology which was not conclusive. CT scan chest showed multiple metastatic nodules on right side and few small nodules on left side. CT guided biopsy of the pulmonary nodules showed malignant cells but did not show any definitive site of primary lesion. Taking into account of the all above total thyroidectomy was performed (Figure 1) with clinical impression of metastatic follicular carcinoma. Final histopathology showed tumor composed of vascular elements arranged in sheets with cellular atypia without any significant cellular differentiation. Diagnosis of epithelioid angiosarcoma of the thyroid gland was made with Thyroid Transcription Factor-1 (TTF-1) negative and CD-34 & ERG positive immunoreactivity (Figure 2). Patient is currently having chemotherapy with Adriamycin and is fine 3 months postoperative with resolution of the pulmonary metastasis.

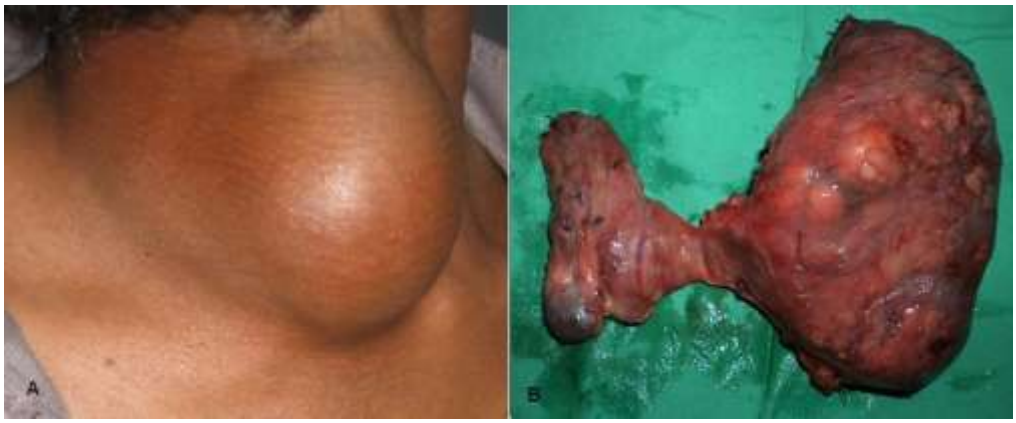

Figure 1 A, Preoperative Neck Image, **B,** Thyroid Specimen

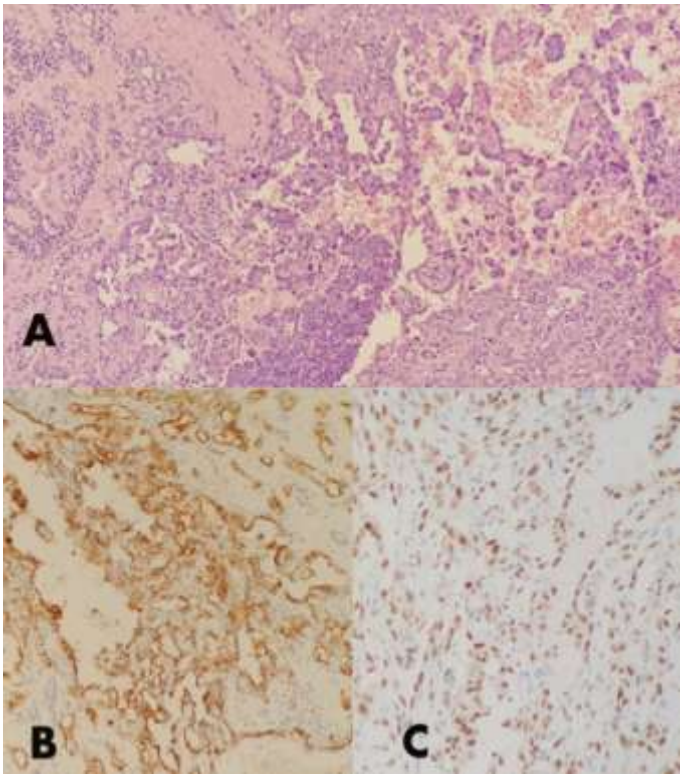

Figure 2 A, shows the undifferentiated pattern of the tumor cells on Eiosin and Hemotoxyciline staining. B, shows CD-34 and C shows ERG immunoreactivity.

Discussion

Epithelioid Angiosarcoma is a very rare tumor of thyroid gland with poor prognostic indicators without any well known etiological risk factors. Considering the higher incidence of this tumor in Alpine region, it has been postulated that long-standing goiters in hilly areas due to iodine deficiency may be one plausible risk factor [2,4]. Females are preferentially affected over the males like any other thyroid problem [3]. Preoperative diagnosis based on FNAC is unlikely and all diagnoses are exclusively made on histological and immunohistochemcial basis [1, 2].

Epithelioid Angiosarcoma of thyroid was first recognized by Eusebi et al in 1990 as a distinct variety which was previously reported as angiomatoid variant of anaplastic carcinoma [8, 11]. This controversy has been partially settled in 2004 when Epithelioid Angiosarcoma was formally included as a distinct pathology in WHO thyroid malignancy classification [8, 11]. Histologically it is challenging to differentiate the angiomatoid anaplastic cancer from the Epithelioid angiosarcoma however modern immunological staining techniques has been useful in this differentiation [9, 10]. Here in our case we used three important markers namely, Thyroid Transcription Factor-1 (TTF-1), CD-34 and Erythroblast Transcription specific related gene (ERG) to reach to a diagnosis. Negative Immunostain with TTF-1 virtually excludes the follicular cell origin and makes the possibility of undifferentiated follicular variant of anaplastic carcinoma least likely. Moreover, immunoreactivity with CD-34 and ERG strongly supports the epithelioid sarcomatous features of vascular origin [1, 2].

Management of Angiosarcoma of thyroid is primarily surgical if permissible. En-block resection followed by chemoradiation is imperative considering aggressive behavior of this tumor [12]. Adriamycin is the drug of choice though we do not have any controlled data to support it. Radiation therapy has also been used for the local control [1, 2]. Prognosis primarily depends on extracapsular extension of the tumor and distant metastasis. Goh et al have estimated a 5 year survival for this tumor to be 33% [13]. Patients with Stage IV disease have been reported to die within 6 months [13].

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