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*Corresponding author

*Nihal Seden Boyoğlu, Department of Otorhinolaryngology - Head and Neck Surgery, University of Health Sciences, Istanbul Training and Research Hospital, İstanbul, Turkey.

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Case Report

A Rare Case of Supraclavıcular Regıon: Branchıoma (Ectopıc Cervıcal Thymoma)

Nihal Seden Boyoğlu*, Ezgi Keskin, Kürşat İlyas Çil, Özgür Yiğit

Department of Otorhinolaryngology - Head and Neck Surgery, University of Health Sciences, Istanbul Training and Research Hospital, İstanbul, Turkey.

Abstract

Background: In the past, it was believed that ectopic thymoma was a rare hamartomatous tumor found in the cervical region, caused by the presence of ectopic thymic tissue due to disrupted thymus migration during fetal development. However, recent literature indicates that these lesions do not originate from the thymus and should be referred to as branchiomas. Despite being uncommon, it is crucial to consider these lesions in the differential diagnosis of cervical masses.

Case presentation: A 53-year-old male patient presented to our clinic with a one-year history of swelling on the left side of the neck. Upon examination, a 6 cm mass was found in the left supraclavicular region. An MRI of the neck revealed an oval mass with mixed components of soft tissue and fat density. A fine needle aspiration confirmed the diagnosis of ectopic thymoma. The patient then underwent a complete surgical resection of the tumor under general anesthesia, with no significant residual tumor. The final histopathological diagnosis was branchioma.

Conclusion: Although ectopic thymomas in the cervical region are rare; thymomas, currently known as branchioma, may be considered a differential diagnosis for supraclavicular masses.

Introduction

Thymic tumors are rare but account for about 47% of anterior mediastinal tumors. They can also be found ectopically in the middle or posterior mediastinum, lung, and pleura. Ectopic thymomas are believed to originate from dispersed thymic tissue that cannot migrate into the anterior-superior mediastinum [1].

Although in previous years, neck lesions with histological features resembling thymoma were categorized as ectopic thymomas, recent immunohistochemical studies have revealed that there is no resemblance between ectopic thymomas in the mediastinum or lung and neck lesions. As a result, this entity has been excluded from the ectopic thymoma classification.

Different names have been previously used to refer to this entity (such as ectopic thymoma of the lung, bronchial mixed tumor, thymic tumor, and branching phenotype), but currently, the term ‘‘branchioma’’; is widely accepted. Branchioma consists of epidermal and mesodermal lineage derivatives, including epithelial islands, spindle cells, without preexisting thymic tissue or evidence of thymic differentiation [2].

In this case, the clinicopathological features of a branchioma in the left supraclavicular region are described and the literature regarding this topic is reviewed, and the rationale for abandoning the term ectopic thymoma is summarized.

Case presentation

A 53-year-old male patient presented with a 1-year history of a slow-

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growing painless mass in the left supraclavicular region with mild tenderness on palpation. He was diagnosed with FMF and type 1 diabetes and was taking oral antidiabetics and colchicine. He had a history of appendectomy 20 years ago and an operation for inguinal hernia 1 year ago. His older sister had leukemia. He did not consume alcohol or tobacco. Routine laboratory data were all within normal ranges. Physical examination revealed a 6.0 cm oval, mobile mass in the left supraclavicular region. The hard mass was slightly tender and had clear boundaries on palpation. Neck MRI also revealed an oval heterogeneous mass containing mixed components of soft tissue and fat density, enhanced with contrast (Figure 1). There was no abnormality on the chest X-ray. A fine needle aspiration was performed, and a condition with undetermined cytopathology was reported. A second fine needle aspiration was performed, and a diagnosis of ectopic hamartomatous thymoma was made. The patient was consulted preoperatively to pulmonology and neurology. EMG and chest CT were performed. No additional pathology or myasthenia gravis was detected. There was no obstacle to the operation. The patient then underwent a major total surgical resection of the tumor under field block anesthesia, without any large residual tumor. During the surgery, a well-circumscribed oval tumor measuring 7.0 cm x 5 cm x 2.5 cm, without invasion, was detected under the SCM muscle in the left supraclavicular region. The tumor caused moderate dorsal displacement of the left carotid artery due to its pressure. Additionally, there was no evidence of tumor invasion into adjacent tissues. The tumor was not connected to the thyroid, clavicle, or mediastinum (Figure 2) . The patient's postoperative period was uneventful.

Pathological tissue presented as a nodular lesion, measuring 7 x 5 x 3.5 cm, with a yellow-brown color and a shiny, smooth surface. On cross sections, there were large yellow areas with a regular appearance, as well as scattered cream-colored, fibrotic-looking areas and occasional bleeding areas.

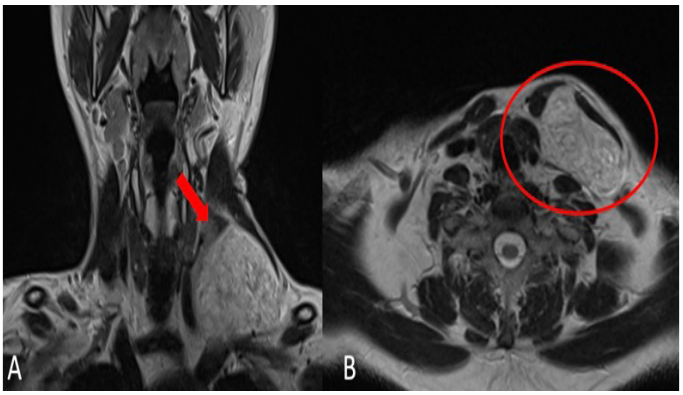

Figure 1:
A. *Cervical mass detected in the left side of supraclavicular region by coronal T2 weighted image (T2WI) (red arrow).*

B. *Enhanced neck MRI axial section showed a solitary, well-defined nodule in the left supraclavicular region.*

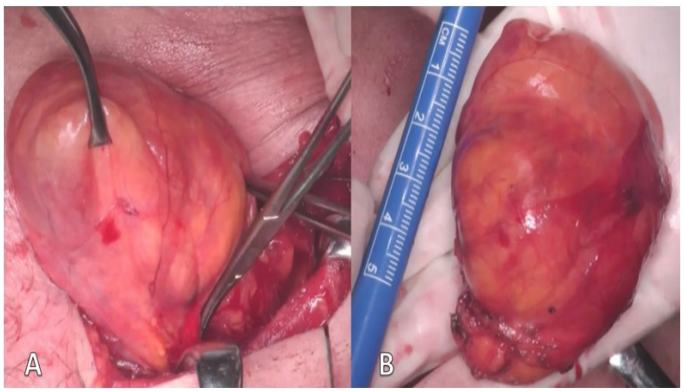

Figure 2:
A. *A well-defined, oval tumor was found beneath the SCM muscle in the left supraclavicular region. Furthermore, there was no indication of tumor infiltration into surrounding tissues.*
B*. Specimen appeared as a nodular lesion, measuring 7 x 5 x 3.5 cm, with a yellow-brown color and a shiny, smooth surface.*

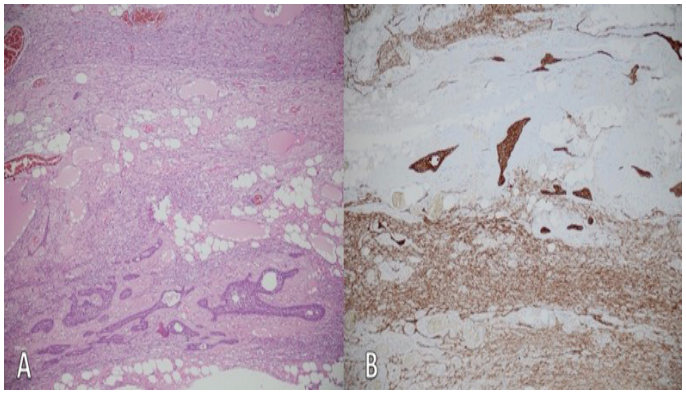

Figure 3:
A. *Polygonal epitheloid cells island and uniform spindle cells which.*
B. *Cytokeratin 19 positivity at both epitheloid and spindle cell component.*

At the histopathological examination, uniform bland spindle cells are intermingled with islands of polygonal epithelioid cells. Immunohistochemical staining revealed nuclear positivity for p63, along with diffuse and intense reactivity for CK5/6, CK7, and CK19 in both spindle cells and epithelial components. Smooth muscle actin, desmin, and S-100 protein were negative in both spindle cells and epithelial components (Figure 3). The tumor originated from a lipofibroadenoma. These clinicopathological features meet the diagnostic criteria for ectopic thymoma. The final histopathological diagnosis was Type A ectopic thymoma, with no evidence of microinvasion, and negative surgical margins.

Discussion

Cervical thymoma was believed to be originated from

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ectopic thymic tissue that forms due to disruption in the migration of the thymus during the fetal period. It was first described in the 2002 edition of WHO in the group of soft tissue tumors. It is a rare tumor consisting of epithelial areas, spindle cells, and mature fat tissue. It is most commonly seen in middle-aged men and is most commonly seen in the supraclavicular, suprasternal, and sternoclavicular areas.

There are various neck structures and organs that originate from the branchial pouches, clefts, and arches, respectively. It is important to highlight that this branch apparatus could potentially be linked to the formation of this tumor. These apparatus give rise to fat, myoepithelial and epithelial cells, including squamous and glandular epithelium, and thus have "branchial" origin. The etymology of "branchioma" encompass the likely origin of the neoplasm (branchi). The lesion is not ectopic, as the components of the tumor are normal for the location embryologically (eutopic). PAX8, typically positive in all thymus-origin lesions, tested negative in this case, indicating a non-thymus origin for the lesion. [3].

There have been 83 cases of the ectopic cervical thymoma reported in English literature up to now [4].The fact that most of the tumors called cervical thymoma occur in the supraclavicular area, as in our case, suggests that they are a result of developmental abnormality in the third or fourth branchial sacs, the cervical sinus of His, and the ultimobranchial body. In a case series, it has been noted that the prevalence of this tumor is higher on the left side compared to the right side. This observation indicates that the left supraclavicular space contains the thoracic duct and jugular lymphatic trunk, suggesting that anatomical variations may contribute to the development of the tumor [5].

It is well known that nearly all of ectopic cervical thymoma show no evidence of atypical or malignant features and pursue a benign clinical course, and complete local excision is the first treatment of choice. Residual thymic structures have never been described in association with ectopic cervical thymoma and a mediastinal counterpart was absent in all cases of cervical thymoma reported to date [6] .

Although there is confusion in taxonomy, we believe that a correct definition will allow for a more active role in patient management. The current naming of ectopic thymoma leads to further examinations for thymomas located in the mediastinum or lung, resulting in additional cost and loss of time. Surgical excision is the treatment method for cervical thymomas, recommended to be called branchiomas, while complementary thymectomy, the treatment approach for ectopic thymomas seen in the thorax, is not considered necessary in these lesions.

Conclusion

In this instance, taking into account the patient's age and current clinical findings, diagnoses that are more frequently seen as a mass in the neck were prioritized. After a fine needle aspiration biopsy and imaging, a diagnosis of ectopic cervical thymoma, a rare condition, was made. However, as a result of the literature review and additional examinations, it was concluded that the name cervical thymoma does not fully cover the characteristics of this lesion and that the name branchioma is more appropriate to describe this lesion.

Additional Information

Author contributions: All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Nihal Seden Boyoğlu, Ezgi Keskin Acquisition, analysis, or interpretation of data: Nihal Seden Boyoğlu, Ezgi Keskin , Kürşat İlyas Çil Drafting of the manuscript: Nihal Seden Boyoğlu, Ezgi Keskin, Kürşat İlyas Çil , Özgür Yiğit Critical review of the manuscript for important intellectual content: Nihal Seden Boyoğlu, Ezgi Keskin , Kürşat İlyas Çil, Özgür Yiğit

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