

AB Thymoma with Atypical Type A Component: What Risks

Abderrahim Elktaibi¹, Massine Elhammoumi², Mohamed Reda Elochi¹, Mustapha Azakhmam¹, Mohamed Amine Essaoudi¹, Mohamed Oukabli¹, Abderrahmane AL Bouzidi¹, Elhassane Kabiri²

¹Department of Pathology, Mohammed V Military Teaching Hospital, Rabat, Morocco

²Department of Thoracic surgery, Mohammed V Military Teaching Hospital, Rabat, Morocco

*Corresponding author: Dr. Abderrahim Elktaibi

| Received: 16.03.2019 | Accepted: 24.03.2019 | Published: 31.03.2019

DOI: [10.21276/sjmps.2019.5.3.17](https://doi.org/10.21276/sjmps.2019.5.3.17)

Abstract

Type AB thymoma is a thymic epithelial neoplasm composed of component spindle cell (type A) and lymphocyte-rich (type B) component. An exceptional atypical type A thymoma variant with uncertain prognosis was added to the WHO classification of thymomas in 2015. We report a case of a 62 years old man presented with clinical myasthenia. The radiological examination revealed a well-circumscribed anterior mediastinal mass. The patient underwent complete thymectomy. Pathological and immunohistochemical analysis revealed an AB thymoma with atypical component of type A. At the first year post surgery follow-up appointment, the patient was alive with no evidence of recurrence or complication. The presence of atypical component of type A thymoma may increase the risk of progression or metastasis, so extended follow-up for patients is required.

Keywords: AB thymoma, Atypical type A thymoma, Pathology, Thymectomy.

Copyright @ 2019: This is an open-access article distributed under the terms of the Creative Commons Attribution license which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use (NonCommercial, or CC-BY-NC) provided the original author and source are credited.

INTRODUCTION

Type AB thymoma is a thymic epithelial neoplasm composed of component spindle cell (type A) and lymphocyte-rich (type B) component. The relative proportion is highly variable. An exceptional atypical type A thymoma variant with uncertain prognosis was added to the WHO classification of thymomas in 2015. To our knowledge, this is the second description of atypical A component in AB thymoma that may increase the risk of recurrence or metastasis.

Observation

A 62 years old man with a 12 years history of diabetes with insulino therapy, presented with clinical myasthenia with no fever, no shortness of breath no weight loss and chest pain. No therapy was taken before admitted apart from anticholinesterasic. A well-circumscribed anterior mediastinal mass, measuring 52 × 42 × 30mm was identified in chest radiograph and CT associated with calcifications (Fig-1A). Laboratory tests revealed positive anti cholinesterasic anticorps. The patient underwent complete thymectomy through total median sternotomy. Peroperatively, the mass showed an invasive behaviour of the right phrenic nerve

making mandatory nerve section (Fig-1B). Macroscopically, the tumor was encapsulated. The cut surface showed multiple nodules with areas of necrosis separated by white fibrous bands. Histological examination showed that the tumor was surrounded by a fibrous capsule and the majority of the neoplasm was composed of lobulated growth pattern, demonstrated a highly variable mixture of a lymphocyte-poor type A component and a more lymphocyte-rich type B-like component. These components formed separated nodules. Some nodules were characterized by some degree of atypia, hypercellularity, focal necrosis and high mitotic activity (12 mitoses per 2 mm²) (Fig-1C). Immunohistological analyses showed that epithelial cells were positive in cytokeratin19 and CKAE1-AE3. CD3, CD5 and TDT expression was detected in immature lymphocytes T cells. The Ki-67 index was 70 % (Fig-1D). On the basis of these findings, the tumor was finally diagnosed to be mixed AB thymoma with atypical type A component, stage I of Masaoka. The patient was discharged at the third day. At the first year post surgery follow-up appointment, the patient was alive with no evidence of recurrence or metastasis. We continue to conduct careful postoperative follow-up.

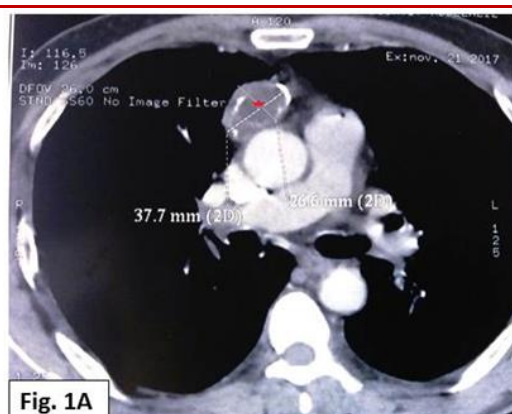


Fig-1A: Chest computed tomography revealing an anterior mediastinal tumor with heterogeneous enhancement (maximum diameter 37, 7 mm)

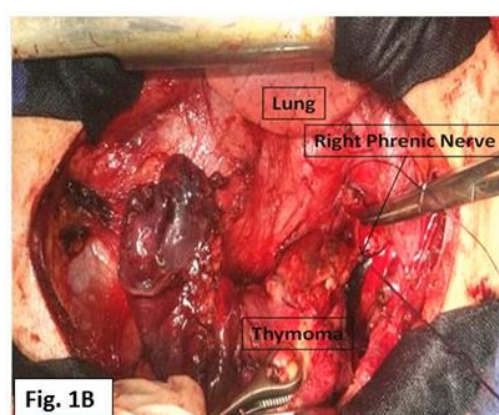


Fig-1B: Intraoperative image showing a mass of the thymic lodge adhering to the right phrenic nerve

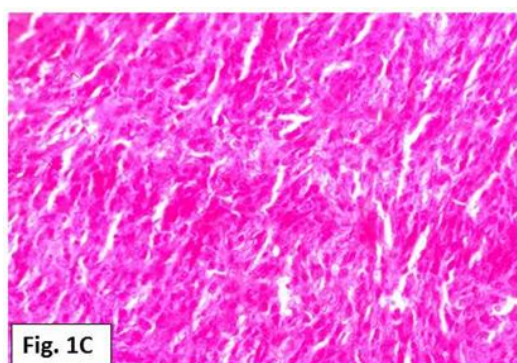


Fig-1C: Microscopic findings showing the spindle- and oval-shaped tumor cells with hypercellularity and moderate atypia. Hematoxylin–eosin staining×250

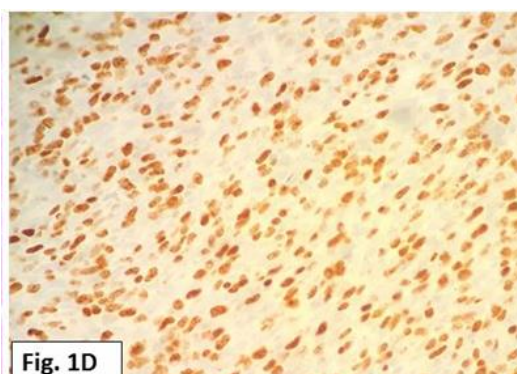


Fig-1D: Microscopic findings on immunohistochemical staining: Ki-67 labeling index was 70 % × 400

DISCUSSION

Thymomas are rare tumors that arise from the thymus gland. They are the most frequent neoplasms of the anterior mediastinum in adults with an approximate incidence ranged from 2, 2 to 2, 6 per million per year. The World Health Organization (WHO) classification of these tumors recognizes types A and AB and B1, B2, and B3 subtypes of thymomas in addition to rarer variants such as micronodular thymoma, metaplastic thymoma, or sclerosing thymoma [1]. Most of the type AB thymoma show almost benign behavior and considered benign tumor because of their excellent prognosis. It is now accepted that a few cases of A thymoma can display some degree of atypia and showing increased mitotic activity, nuclear crowding and/or a scattered foci of necrosis [2]. An atypical type A thymoma variant was added to the WHO classification of type A thymoma in 2015 based on the presence of type A thymoma cases with local aggressive behavior and tumor recurrence [3]. This variant was first described by Nonaka *et al.*, in 2012. Grajkowska *et al.*, reported a case of AB type thymoma with an atypical type A component diagnosed with negative surgical margin and low stage of Masaoka. The patient developed pulmonary and cerebral metastases after 10 and 15 years respectively. The metastases showed a similar histological feature of the atypical type A component [4]. In a study of 23 patients who diagnosed with atypical type A thymoma, 43% developed metastases and recurrences during the follow-up period (average 49 months). The authors reported that necrosis was the only histological factor associated with a poor prognosis of relapse regardless margin resection [5]. Bürger *et al.*, have examined five metastatic type A thymomas and did not find any differences of genetic alterations between the conventional and atypical variants of type A thymoma [4].

CONCLUSION

Our case illustrates an exceptional case of an AB thymoma associated with atypical type A component. The latter impacts the prognosis and can increase the risk of recurrences and metastases. A wide sample of tumor resection coupled with a rigorous pathological analysis is recommended for the detection of an atypical component. When, the diagnosis of atypical type A thymoma variant was made, a careful postoperative systemic follow-up should be conducted.

Conflicting Interests

The Authors declares that there is no conflict of interest.

Consent for Publication

Written informed consent was obtained from the patient for publication and any accompanying images.

REFERENCES

1. Ströbel, P., Kurrer, M. O., Marx, A., Marchevsky, A. M., Badve, S., Marom, E. M., ... & Nicholson, A. (2015). Tumours of the thymus. *Travis WD, Brambilla E, Burke AP, Marx A, Nicholson AG. WHO Classification of Tumours of the Lung, Pleura, Thymus and Heart. 4th ed. Lyon: IARC, 183-298.*
2. Kawakita, N., Kondo, K., Toba, H., Yoneda, A., Takizawa, H., & Tangoku, A. (2018). A case of atypical type A thymoma with vascular invasion and lung metastasis. *General thoracic and cardiovascular surgery*, 1-4.
3. Hashimoto, M., Shimizu, S., Takuwa, T., Tsukamoto, Y., Tsujimura, T., & Hasegawa, S. (2016). A case of atypical type A thymoma variant. *Surgical case reports*, 2(1), 116-119.
4. Grajkowska, W., Matyja, E., Kunicki, J., Szymanska, S., Marx, A., Weis, C. A., ... & Szolkowska, M. (2017). AB thymoma with atypical type A component with delayed multiple lung and brain metastases. *Journal of thoracic disease*, 9(9), 808-814.
5. Hashimoto, M., Tsukamoto, Y., Matsuo, S., Nakamichi, T., Kondo, N., & Hasegawa, S. (2017). Lung metastases in an atypical type A thymoma variant. *Journal of thoracic disease*, 9(9), 805-807.