

# Atypical Type A Thymoma – A Rare Presentation: A Case Report

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## Abstract:

**Introduction:** Thymomas are the most common tumor of the mediastinum with varied presentations. Here we present our experience with this tumor in a patient who was treated successfully at our department.

**Case Presentation:** We present a case of 29-year-old male who presented to us with intermittent cough and dull aching chest pain. Biopsy revealed moderate pleomorphism with focal areas of haemorrhage. Immunohistochemistry was suggestive of Type A thymoma. Patient underwent en-mass removal of the thymoma via midline sternotomy. Patient had a smooth post operative recovery.

**Conclusion:** Multidisciplinary approach is needed both for diagnosis as well as successful management of patients with thymoma.

**keywords:** polymastia; supernumerary; breast

## Introduction

Type A Thymoma is low grade malignant tumor. They are included in a family of aggressive tumors (1). We present this rare case which was managed successfully in our department.

## Case Presentation:

A 29-year-old male presented in outpatient department with complaints of intermittent cough with dull aching chest pain. Patient was evaluated at some Pulmonary clinic where CT (Computerized Tomography) guided lung biopsy was taken, which revealed moderate pleomorphism, mitosis present, focal hemorrhage. To differentiate better immunohistochemistry was done, which was positive for Pan CK, LCA and Ki67. It was negative for CK7, CK20, Chromogranin. CECT (Contrast Enhanced Computerized Tomography Chest) suggestive of Anterior mediastinal mass (Figure 1) 6.7x6.0x9.0 cm with SUV max 2.6 and abutting the SVC & RA. PET-CT (Positron Emission Tomography) suggestive of no distant metastasis (Figure 2). Tumor markers CA 19-9, AFP, B-HCG 1.4, CA 125 was within normal limit. Diagnosis of Type A Thymoma was established. He was planned for elective surgery. A Midline Sternotomy was performed and tumor was resected en-mass. It measured approximately 12x8 cm with areas of hemorrhage. It did not invade

pleura, pericardium, heart and great vessels. Postoperative recovery was uneventful (Figure 3). He was asymptomatic at 3 months of follow up.

Macroscopically it was a well encapsulated greyish brown soft tissue mass of size 11.6x 6 cm. microscopic examination showed tumor cells arranged in diffuse solid sheets and vague storiform pattern. Pleomorphism was present with ovoid to spindle shape hyperchromatic nucleus and mitotic figures less than 4 mitosis/mm<sup>2</sup>.

## Discussion:

About 40% of mediastinal tumors are asymptomatic. Symptoms are generally due to stretching or compression on surrounding structures by the tumor. Local tumor growth causing compression to SVC may cause facial puffiness & headache. Impingement to right atrium may cause atrial fibrillation, chest pain and syncope. Pressure on trachea may cause respiratory distress and to esophagus may cause gastrointestinal symptoms such as vomiting. According to histological criteria of WHO Thymoma are classified as:

Type A (Spindle Cell)

Type AB (Mixed)

Type B1 (Lymphocyte Rich)

Type B2 (Cortical)

Type B3 (Epithelial)

Type C (Thymic Carcinoma) (2)

where Type A Thymoma is regarded as benign tumor.

Atypical type A Thymoma was added to WHO classification in 2015 with following pathological finding:

1. Mild to moderate Nuclear Atypia
2. Increase in Mitotic Activity
3. Scattered foci of Necrosis (1)

Literature suggests distant metastasis and tumor recurrence is 43% in case of Atypical Type A Thymoma (1). According to literature, patients with Type A Thymoma survival rate is 100% at 10 years of follow up (3) They are rare in first 2 decades of life. About 50% of the diagnosis are incidental in nature (4). Multidisciplinary involvement is beneficial for the management. We proceeded with midline sternotomy in view of achieving safe resection of the tumor from the vitals structures such as SVC and RA (5). In cases with clear fat planes uni-portal VATS has been advocated. In our patient, the mass was abutting the SVC & RA.

Although CECT provides the detailed anatomical delineation of the tumor, histological diagnosis helps in decision making for further management. Relationship between SUV max and WHO classification suggest that SUV max of 3.43 and 4.43 is low risk and high risk thymomas respectively (6). MRI helps to differentiate between benign and malignant tumors (7). Since facility of MRI was not available at our institute, a PET-CT was performed. Complete radiological and tumor markers assessment is necessary to differentiate it from other mediastinal tumors like Myasthenia Gravis, non seminomatous mediastinal germ cell tumor (NSGCT), lymphoma. Although identifying the nature of the lesion by tissue biopsy remain controversial (8), benefits of recognizing the lesion

before surgery helps to give Neo-adiuvant chemotherapy making lesion amenable for resection.

In our case the patient had already undergone a CT guided biopsy. Differentiating from other pathological condition, remains a challenge for pathologist particularly when there is extensive necrosis (9). In our case since there was minimal necrosis so, Spindle Cell Carcinoid becomes less obvious. Also, the absence of positive neuroendocrine marker makes it more unfavourable. Atypical Thymoma type A is usually well circumscribed. Absence of invasion to adjacent organ with easy resectability is more in line with the diagnosis.

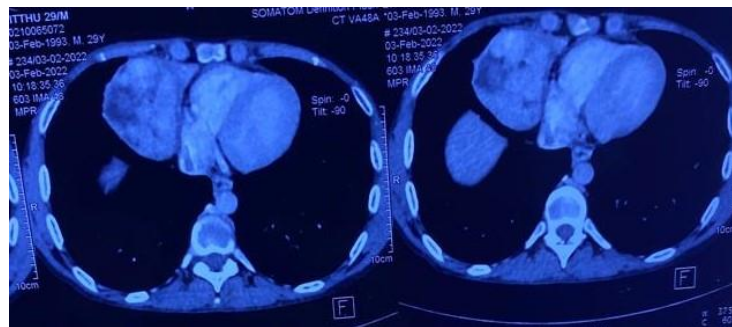
Anesthesiological consideration is of utmost important as any injury to SVC during resection can be fatal thus vascular access in low extremity is important and can be lifesaving. In cases where vascular injury has occurred, Cardiopulmonary bypass and Perfusionist should be readily accessible to provide a rescue. Where SVC is suspected to be involved an extra caution is must as additional monitoring modality like Near Infrared Spectroscopy, Transcranial Doppler is recommended (10).

Post-operative follow up should not be limited to chest imaging but also to extra thoracic. cavity is necessary. So, we have planned the patient to undergo PET CT annually three consecutive years and there by deciding later.

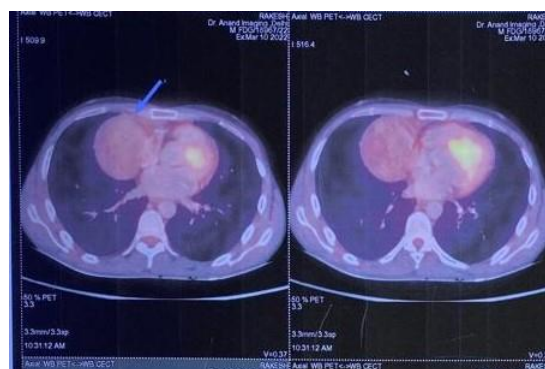
**Conclusion:**

Patients with mediastinal mass should undergo CT guided biopsy followed by surgery for better diagnosis and management. We recommend multidisciplinary approach to provide disease free survival.

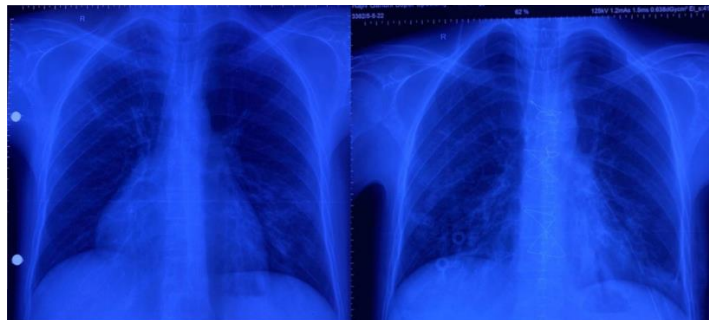
Abbreviations: TB:Tuberculosis, ATT:Anti-Tubercular Drugs, CECT: Contrast Enhanced Computerised Tomography, SVC: Superior Vena Cava, RA: Right Atrium, PETCT: Positron Emission Tomography Computerised Tomography, AFP: Alfa Feto-Protein, CA125: Cancer Antigen125,Beta- Human Chorionic Gonadotropin CK: Cytokeratin LCA: Leukocyte Common Antigen SVC: Superior Vena Cava



**Figure1:** Preoperative CT Scan images showing anterior mediastinal mass, thymoma



**Figure 2:** PET scan of the patient done pre operatively



**Figure 3:** Pre and post operative Chest XRay of the patient

### Declarations:

### Funding:

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### Declaration Of Conflicting Interests:

The author(s) declare no potential conflicts of interest with respect to the research, authorship, and/or publication of this case report.

### Ethical Approval:

Since this is a case report, ethical approval was not required by the Institutional Ethics Committee.

### CONSENT to Participate and Publication:

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

### Availability OF Data and Material:

All relevant data is with the authors and shall be provided as and when asked.

### Code Availability:

Not applicable

### Author contribution:

Study concept: Ankit Jain, Arindam Roy

Data Collection: Ragi Jain, Mona Bargotya

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Revising and critical review: V K Gupta, Rohan Gupta

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