RADY 416 Case Presentation

Stephanie Stookey, MS4
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Thank you to chief resident Justin Rodriguez, MD
Focused Patient History and Workup

Chief Complaint: “productive cough”

History of Present Illness:
• 25 yo female presented to UNC ED referred by her PCP for abnormal chest xray.
• Chest xray revealed “markedly abnormal contour to the right aspect of the mediastinum including a right para-tracheal stripe... peripheral right thorax favored to represent extraparenchymal lesions”

Past Medical History:
• Generalized Anxiety Disorder- well controlled on home Sertraline

Family History:
• Negative for lung cancer

Review of Symptoms:
• No fever, chills, URI sx
• No changes in weight, night sweats
Focused Patient History and Workup

Physical and Exam:
General: Tearful young female
HEENT: Atraumatic, normocephalic, EOMI, PERRLA, MMM.
CV: Tachycardic, regular S1, S2. No rubs or gallops
Lungs: CTAB, no wheezes, rhonchi, or rales
Abdomen: Normoactive bowel sounds, soft, NTND
Extremities: no clubbing, cyanosis, edema
Skin: No rashes or lesions

Pt was admitted to inpatient and Interventional Pulmonology and Heme/Onc were consulted. Workup included contrasted LP, bronchoscopy w/ infectious panel and mass biopsy, respiratory viral panel, and TB.
List of UNC Imaging Studies

• CTA Chest with contrast
  • Obtained: 03/21/2019

• CT Chest with contrast
  • Obtained: 05/29/2019

• PET Body
  • Obtained: 07/02/2019
Computerized Tomography Chest Axial View (March and May)

Mediastinal mass with pleural and lung metastases
Anterior mediastinal soft tissue mass and extensive **right pleural nodularity** noted.

- Confluent mediastinal adenopathy involving the anterior mediastinum, paratracheal nodes, subcarinal nodes, aortopulmonary window, and right hilar nodes.
- These findings are concerning for invasive mediastinal malignancy
- Diffuse pleural masses on the right with confluent mediastinal adenopathy and diffuse pulmonary nodules. Differential includes metastatic disease, lymphoma, or primary pleural malignancy.
• S/p CAPP (Cisplatin + doxorubicin + cyclophosphamide + prednisone)

• Marked decrease in size of anterior mediastinal mass (compatible with invasive thymoma), and right pleural metastases, compatible with response to therapy.
PET CT Axial View (note dates)

Mediastinal mass with pleural and lung metastases

07/02/2019
PET CT Axial View (June)

Mediastinal mass with pleural and lung metastases

- Anterior mediastinal mass demonstrating increased FDG uptake (CT 59) corresponding to known invasive thymoma.
- Multiple right-sided pleural-based lesions demonstrated FDG avidity. The lesions are predominantly medial, involving the anterior and posterior medial pleura, and inferior.
- Right-sided pleural-based lesions represent drop metastases from primary invasive thymoma. The most avid metastatic lesion is in the inferior right pleura.
• Mediastinal mass with pleural and lung metastases found on CT were consistent with Stage IV invasive thymoma.

• Patient was followed by Heme Onc and received 4 courses of CAPP (Cisplatin + doxorubicin + cyclophosphamide + prednisone).

• Given tumor burden and lack of surgical options, CT surgery was consulted.

• On 8/5/2019 she underwent a right extrapleural pneumonectomy requiring resection of the right diaphragm, partial resection of the right hepatic lobe, pericardium, and SVC resection with bovine pericardium patch repair.

• She was discharged on 8/15/2019 and will follow up with CT surgery and Heme Onc as outpatient.
Approach to Thymoma

• Thymomas are slow-growing neoplasms that can exhibit aggressive behavior such as invasion of adjacent structures.¹

• Histologic classification primarily distinguishes thymic carcinoma from the different types of thymoma. However, management decisions rest primarily on the stage of disease and the completeness of resection. ¹

• Treatment includes resection and postop radiation therapy.
Approach to Thymoma

• Tumors with a favorable outcome are those that are encapsulated and amenable to complete resection. Invasive and unresectable tumors have a poor prognosis regardless of their histologic characteristics.¹

• A 2001 review showed invasive thymomas were more likely to have lobulated (16/27, 59%) or irregular (6/27, 22%) contours than noninvasive thymomas (8/23, 35% and 1.5/23, 6%, respectively) (p < 0.05).²

• Invasive thymomas had a higher prevalence of low attenuation areas within the tumor (16/27, 60%) than noninvasive thymomas (5/23, 22%) (p < 0.001) as well as foci of calcification (14.5/27, 54% vs. 6/23, 26%; p < 0.01).²
### WHO Histologic Typing of Tumors of the Thymus

#### Epithelial tumors

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
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<tbody>
<tr>
<td>A</td>
<td>Thymoma</td>
</tr>
<tr>
<td>AB</td>
<td>Micronodular thymoma with lymphoid stroma</td>
</tr>
<tr>
<td>B1</td>
<td>Metaplastic thymus</td>
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<tr>
<td>B2</td>
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<td>B3</td>
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#### Thymic carcinoma

- Squamous cell carcinoma
- Basaloid carcinoma
- Mucoepidermoid carcinoma
- Lymphoepithelioma-like carcinoma
- Sarcomatoid carcinoma
- Clear cell carcinoma
- Adenocarcinomas
- NUT carcinoma
- Undifferentiated carcinoma

#### Lymphomas of the mediastinum

<table>
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<th>Type</th>
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<tbody>
<tr>
<td>Primary mediastinal large B-cell lymphoma</td>
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<tr>
<td>Extracapsular marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma)</td>
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<tr>
<td>Other mature B-cell lymphomas</td>
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<tr>
<td>T lymphoblastic lymphoma/leukemia</td>
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<tr>
<td>Anaplastic large cell lymphoma and other rare mature T- and NK-cell lymphomas</td>
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<tr>
<td>Hodgkin lymphoma</td>
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#### Neuroendocrine tumors

- Typical carcinoid tumor
- Atypical carcinoid tumor
- Large cell neuroendocrine carcinoma
- Small cell carcinoma

#### Germ cell tumors of the mediastinum

- Seminoma
- Embryonal carcinoma
- Yolk sac tumour
- Choriocarcinoma
- Mature teratoma
- Immature teratoma
- Mixed germ cell tumour
- Germ cell tumour with somatic type solid malignancy
- Germ cell tumour with associated hematological malignancy

#### Histiocytic and dendritic cell neoplasms of the mediastinum

- Langerhans cell lesions (thymic Langerhans cell histiocytosis, Langerhans cell sarcoma)
- Histiocytic sarcoma
- Follicular dendritic cell sarcoma
- Interdigitating dendritic cell sarcoma
- Fibroblastic reticular cell tumour
- Indeterminate dendritic cell tumour

#### Myeloid sarcoma and extramedullary acute myeloid leukemia

Take Home Points

• Thymomas are the most common primary neoplasms of the anterior mediastinum.\textsuperscript{1-2}

• Preoperative differentiation of invasive from noninvasive thymoma has important implications for treatment planning such as preoperative chemotherapy or radiation therapy and prognosis.\textsuperscript{2}

• The presence of lobulated or irregular contour, areas of low attenuation, and multifocal calcification are suggestive of invasive thymoma.\textsuperscript{2}

• However, CT has a low specificity in assessment of the infiltration of adjacent mediastinal structures, pleura, lung, and chest wall.\textsuperscript{2}
References

