It’s in the chest, but it is not lung!

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Disclosure of Relevant Financial Relationships

Dr. Kalhor declares she has no conflict of interest to disclose.

Case #1

• 38-year-old man presents with chest pain, shortness of breath, and cough for the last two months. Radiographic evaluation shows the presence of an anterior mediastinal mass. FNA aspirate material was interpreted as epithelial neoplasm. Surgical resection of the mass was performed.

Gross Features

• The surgical resection of the mass showed a partially cystic tumor measuring approximately 5.5 cm in greatest dimension. The cut surface was solid and cystic with focal areas of hemorrhage. Necrosis was not present. The tumor was light tan in color.
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p40 immunostain

MAML2 FISH
Diagnosis
Thymic Mucoepidermoid Carcinoma, Low Grade (T-MEC)

T-MEC
- It is a rare salivary gland type tumor of the thymus.
- Numerous case reports and only a couple of medium size series.
- It is part of the spectrum of salivary gland type tumors of the thymus – mixed tumor, adenoid cystic carcinoma, Epithelial-Myoepithelial Ca.

T-MEC
- It represents a rare subset of thymic carcinomas.
  - Thymic carcinomas account for 15-20% of anterior mediastinal epithelial neoplasms.
  - Most low grade tumors present as cystic lesions.
  - While most of the cases reported have been of the low grade type; rare cases of high grade T-MEC may occur.
### Clinical presentation

- Age range from 8 to 84 years (mean 49 years)
- The initial manifestations include respiratory symptoms and weight loss; while some patients are asymptomatic.
- No common association with paraneoplastic syndrome
- Imaging shows an anterior mediastinal mass

### Gross Findings

- Large solid and cystic mass
- May be well circumscribed or adhered to adjacent structures such as pleura and pericardium
- Greatest dimension: 5->15 cm
- Association with multilocular cystic lesions has been documented in a subset of cases

### Histopathologic Features

- There is no evidence of keratinization, or squamous cell Ca.
  
  **Low-grade**
  - Often contain cystic areas
  - A mixture of
    - Epidermoid cells
    - Mucocytes
    - Intermediate cells

  **High-grade**
  - Often solid
  - Areas of necrosis
  - Marked cellular atypia and frequent mitoses

- There is no evidence of keratinization, or squamous cell Ca.
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High grade T-MEC
Immunohistochemical Features

- The tumor may show positive staining for:
  - Keratin 7
  - Keratin 20
  - Keratin 5/6
  - p40

Molecular Pathology

- (11;19)(q21;p13) translocation and mastermind like transcriptional coactivator 2 (MAML2)
- Fluorescence in situ hybridization
- Roden et al showed 2 of 2 primary thymic MECs were positive; while Prieto-Granada et al reported a case of thymic MEC that was negative for MAML2 rearrangement

MAML-2 Rearrangement

- The diagnosis of T-MEC should not be dependent on MAML2
- The diagnosis can be established based on morphology in great majority of the cases in resected specimens.
  - The diagnosis of MEC in biopsy material may be extremely difficult
  - The diagnosis of high grade MEC is more challenging

Differential Diagnosis

- The possibility of primary tumor elsewhere should be excluded.
- Atypical Thymoma (WHO-B3)
- Thymic cystic squamous cell carcinoma
  - Usually it is a keratinizing tumor, contrary to T-MEC
- Adenosquamous carcinoma
  - By definition, it must show an adenocarcinomatous component, which is not present in T-MEC
Atypical Thymoma

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Prognosis and Treatment

- Prognosis depends on the Histologic Grade (low grade vs. high grade) and Staging
- Local invasion and distant metastasis to bone and subcutaneous tissue have been described and are associated with poor outcome
- Surgery
- Chemotherapy
- Radiation

Conclusions

- T-MEC is an unusual primary tumor of the thymus, that may share similar IHC with other thymic epithelial tumors.
- It is important to separate T-MEC into low and high grade tumors.
- It is highly important to separate T-MEC from other thymic epithelial tumors that may follow a different clinical behavior or that may require different treatment.
- The diagnosis of T-MEC does not require positive MAML2.

THANK YOU