Quick Review: Thymoma

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Citation


Abstract

PRIOR TO 1976:

Any tumor of the thymus gland was referred to as a “thymoma”

ture epithelial tumors
thymic carcinoma
seminoma
lymphoma

- Rosai & Levine then defined the modern classification

- the functional unit of the thymus gland is the Lobule
  - composed of a cortex & medulla
  - epithelial cells
  - lymphocytes

THYMOMAS ARE, BY DEFINITION, PRIMARY TUMORS OF BENIGN-APPEARING THYMIC EPITHELIAL CELLS

- Given this definition, most thymomas are “benign”
  - the majority behave in an indolent fashion
  - however, they can invade and metastasize (uncommon)
  - when a thymoma is contained within the thymic capsule, it is referred to as “benign” or “noninvasive”
  - when it penetrates through the capsule, it is referred to as “malignant” or “invasive”

- Thymic tumors composed of truly-malignant-appearing epithelial cells are now referred to as thymic carcinoma
  - rare event
  - accounts for less than 10% of all primary thymic tumors
  - locally aggressive & metastasize hematogenously
  - poor overall prognosis

IN 1985:

A revised histologic classification system

- based primarily on the relation of tumor histology
- not on the degree of cellular atypia
- confusing and unclear metabolic behavior
- controversial etiology & development
- vague natural history & unknown incidence

SOME POINTS

- Most large series from major referral centers report 100-200 patients over 25 - 30 yrs.
- Patients range from 8 months - 89 yrs of age
- Median age of presentation is 52 yrs.
- Thymomas occur equally in men & women
- Racial or ethnic predominance has not been recognized

“A MASS IN THE ANTERIOR MEDIASTINUM”

DIAGNOSTIC STUDIES: CXR & C.T. SCAN

- Thymomas - Lipomas
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- Teratomas - Pericardial Cyst
- Thyroid tissue - Morgagni Hernia
germ cell tumors (seminoma)

**THYMOMA SYMPTOMS**
- Local Signs & Sx's
  - Due to invasion or compression of the mediastinum
  - Hoarseness, cough, chest pain, dyspnea, & SVC-syndrome
  - Usually due to advanced-stage disease
- Systemic Signs & Sx's: the paraneoplastic syndromes
  - Appear early in the course of disease
  - Myasthenia gravis / Red-cell aplasia / Aplastic anemia / Hypogammaglobunemia

The paraneoplastic syndromes are thought to be related to an autoimmune-phenomenon
- ~ 30% of patients with Myasthenia have a thymoma
- role of thymectomy in Myasthenia is clear & convincing

however, this operation has NOT proven effective in treating the other forms

**STAGING**
The most important factor in determining prognosis is the tumor stage at time of operation, which is best described by macro-invasiveness by the surgeon of record
- Stage I, II, & IIb: macroscopically negative
- Stage III, IVa, & IVb: macroscopically-invasive
- Biopsy or “No-Biopsy”
  - 85% of patients will present with resectable disease and should proceed to extended thymectomy regardless of the biopsy result
- these patients should be taken to the operating room without a tissue diagnosis and undergo en-bloc resection
- patients with radiographic-unresectable disease may be candidates for open biopsy or core-biopsy

**SURGICAL RESECTION IS THE MAINSTAY OF TREATMENT FOR THYMOMA**
Following clinical stage, the most important determinant of recurrence is the ability of the operating surgeon to achieve a complete resection at the initial operation
- Extended-resection, 5-yr. Survival: 80 - 98%

**THE EXTENDED THYMECTOMY**
A direct mediastinal approach to allow for the en bloc removal of all cervical & mediastinal soft tissue, including the thymus, mediastinal pleura, adjacent perithymic fat, epiphrenic, & perithyroidal tissue
- removes 95 - 98% of all thymic tissue
- based on Jaretzki’s study of thymic-origin
- Other Surgical Approaches
  - Transcervical Thymectomy
  - Video-assisted Thymectomy
  - Partial mediastinal-debulking
- Operative mortality in most series is 0 - 1%
- Permanent injury to the Phrenic or Recurrent nerves: 1%
- Most common complications: arrythmias, residual ptx, effusions

**OTHER MODALITIES OF TREATMENT**
- Primary Radiotherapy: “unresectable tumor” / SVC-syndrome
- Adjuvant Radiotherapy: the standard of care for thymoma
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- thymomas are well-known to be radiosensitive
- decreases recurrence by 5-fold across the board

Chemotherapy:
- glucocorticoids & platinum-based agents
- controversial / lack of long-term trials

MYASTHENIA GRAVIS

- 25,000 patients in the U.S. population
- 2 - 80 yrs. of age (average age at surgery: 34 yrs)
- 2:1 female predominance
- incidence of thymoma in M.G. patients: 25 - 30 %
- presents as progressive-generalized weakness

  - eyes: ptosis / diplopia
  - bulbar muscles: Dysphagia / dysarthria
  - proximal limbs

“A deficiency of Ach-receptors (due to anti-AchR antibodies) leads to a summarily-weakening of end-plate potentials and thus a progressive failure to generate muscle contractures over time”

- First-line Tx: Medical
  - anticholinesterase agents: slow Ach breakdown
  - immunosuppressive agents: suppress antibody formation
  - plasmapheresis: removal of plasma-related factors

Thymectomy & Myasthenia Gravis

- The treatment of choice for all patients with generalized sx’s
  - Preceded by medical stabilization & optimization
  - Controversial with ocular-pt’s (15 %)
  - The exact technical approach is dependent on preference
  - Modified Osserman Classification
    - Classes I – V
    - Following surgery, up to 96 % of pt’s will improve by 1-2 classes

QUESTION: WHAT IS THE “”?
QUESTION: WHO WAS THE FIRST TO REPORT HIS SURGICAL EXPERIENCE WITH MYASTHENIA GRAVIS?
(clue: he removed a cystic mass from a young woman with myasthenia and she suddenly improved...)

References
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