

Quick Review: Thymoma

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Citation

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Abstract

PRIOR TO 1976:

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

true 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

THYOMAS 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

- 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: arrhythmias, residual ptx, effusions

OTHER MODALITIES OF TREATMENT

- Primary Radiotherapy: “unresectable tumor” / SVC-syndrome
- Adjuvant Radiotherapy: the standard of care for thymoma

- 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. pateints: 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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