Thymoma

Treatment of Early Stage Thymic Tumors: Surgery and Radiation Therapy

Benjamin T. Gielda, MD
Rick Peng, MD
Joy L. Coleman, MD
Charles R. Thomas, MD*
Robert B. Cameron, MD

Address
*Department of Radiation Oncology, Rush University Medical Center, Chicago, IL, USA.
E-mail: thomasch@ohsu.edu

Opinion statement
Tumors of the thymus are an uncommon entity, constituting 30% and 15% of anterior mediastinal masses in adults and children, respectively. The majority of these tumors are thymomas, with thymic carcinomas less common, and thymic carcinoids exceedingly rare. Recognition of the distinct clinicopathologic behavior of various thymic neoplasms is crucial to providing optimal treatment. Evidence guiding the treatment of early stage thymic tumors is limited secondary to the low incidence and resulting lack of randomized data. Proper management requires a careful analysis of the available literature with particular attention paid to limitations of the existing studies. This article provides a discussion of the presentation, evaluation, diagnosis, surgical techniques, and treatment outcomes relevant to early stage thymomas, thymic carcinomas, and thymic carcinoid tumors. The role of radiation therapy in the management of early stage thymic tumors remains controversial and is discussed in detail.

Thymic neoplasms

Although lymphomas, carcinoid tumors, and germ-cell tumors all may arise within the thymus, only thymomas, thymic carcinomas, and thymolipomas arise from true thymic elements.

Thymic neoplasms constitute 30% and 15% of anterior mediastinal masses in adults and children, respectively, with thymomas being the most common [1, 2]. Ninety percent of thymomas occur in the anterior mediastinum, and the remainder arise in the neck or other areas of the mediastinum including, rarely, the heart [3].

Thymomas grossly are lobulated, firm, tan-pink to gray tumors that may contain cystic spaces, calcification, or hemorrhage. They may be encapsulated, adherent to surrounding structures, or frankly invasive. To unify the pathology of thymic neoplasms, the World Health Organization (WHO) adopted a new classification system for thymic neoplasms (Table 1).
WHO-type A-B2 tumors are more likely to present with loco-regional disease, compared to WHO-type B3-C tumors [7, 8]. Based on the Masaoka thymoma staging system, stage I and II are considered early stages. In stage I, the tumor is macroscopically encapsulated and without microscopic capsular invasion. In stage II, the tumor invades into the surrounding fatty tissue or mediastinal pleura or exhibits microscopic capsular invasion.

### Thymoma

#### Presentation

- Nearly one-half of thymomas are asymptomatic and discovered incidentally. In symptomatic patients, 40% have symptoms of myasthenia gravis whereas others complain of chest pain and symptoms of hemorrhage or compression of mediastinal structures [9].

#### Associated systemic syndromes

- Myasthenia gravis (MG) is the most common autoimmune disorder associated with thymoma, occurring in 30–50% of patients [10, 11]. Symptoms include diplopia, ptosis, dysphagia, and fatigue. Ocular symptoms are the most frequent initial complaint, eventually progressing to generalized weakness in 80%. Some improvement in myasthenic symptoms almost always occurs after thymectomy, but complete remission rates vary from 7% to 63% [12]. Patients with MG and thymomas do not respond as well to thymectomy as do MG patients without thymomas. Other less commonly associated systemic syndromes include red cell aplasia and hypogammaglobulinemia.

#### Radiographic imaging studies

- Radiological studies play a central role in the evaluation of thymoma. Since many patients are asymptomatic at presentation, a widened mediastinum or loss of the normal anterior clear space on the lateral film of a routine chest radiogram may be the first sign of disease. In such a patient, an intravenous contrast-enhanced spiral computed...