

interconnecting processes. Whorls of medullary epithelial cells create *Hassall corpuscles*, with their characteristic keratinized cores.

As you know from the earlier consideration of the thymus in relation to immunity, progenitor cells migrate from the marrow to the thymus and mature into T cells, which are exported to the periphery, but only after they have been educated in the “thymic university” to distinguish between self and non-self antigens. During adulthood the thymic production of T cells slowly declines as the organ atrophies.

Macrophages, dendritic cells, a minor population of B lymphocytes, rare neutrophils and eosinophils, and scattered myoid (muscle-like) cells are also found within the thymus. The myoid cells are of particular interest because of the suspicion that they play some role in the development of myasthenia gravis, a musculoskeletal disorder of immune origin.

Pathologic changes within the thymus are limited and will be described here. The changes associated with myasthenia gravis are considered in Chapter 27.

## Developmental Disorders

*Thymic hypoplasia* or *aplasia* is seen in DiGeorge syndrome, which is marked by severe defects in cell-mediated immunity and variable abnormalities of parathyroid development associated with hypoparathyroidism. As discussed in Chapter 5, DiGeorge syndrome is often associated with other developmental defects as part of the 22q11 deletion syndrome.

Isolated *thymic cysts* are uncommon lesions that are usually discovered incidentally postmortem or during surgery. They rarely exceed 4 cm in diameter, can be spherical or arborizing, and are lined by stratified to columnar epithelium. The fluid contents can be serous or mucinous and are often modified by hemorrhage.

While isolated cysts are not clinically significant, neoplastic thymic masses (whatever their origin) compress and distort adjacent normal thymus and sometimes cause

cysts to form. Therefore, the presence of a cystic thymic lesion in a symptomatic patient should provoke a thorough search for a neoplasm, particularly a lymphoma or a thymoma.

## Thymic Hyperplasia

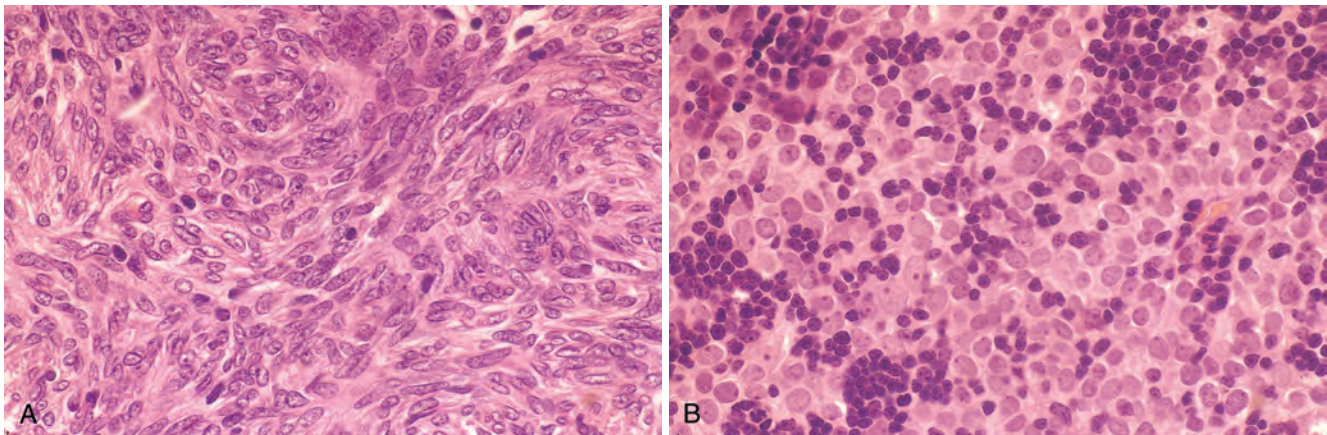
The term *thymic hyperplasia* is a bit misleading, since it usually applies to the appearance of B-cell germinal centers within the thymus, a finding that is referred to as *thymic follicular hyperplasia*. Such B-cell follicles are present in only small numbers in the normal thymus. Although follicular hyperplasia can occur in a number of chronic inflammatory and immunologic states, it is most frequently encountered in myasthenia gravis, in which it is found in 65% to 75% of cases (Chapter 27). Similar thymic changes are sometimes encountered in Graves disease, systemic lupus erythematosus, scleroderma, rheumatoid arthritis, and other autoimmune disorders.

In other instances, a morphologically normal thymus is simply large for the age of the patient. As mentioned, the size of the thymus varies widely, and whether this constitutes a true hyperplasia or is merely a variant of normal is unclear. The main significance of this form of thymic “hyperplasia” is that it may be mistaken radiologically for a thymoma, leading to unnecessary surgical procedures.

## Thymomas

A diversity of neoplasms may arise in the thymus—germ cell tumors, lymphomas, carcinoids, and others—but the designation “**thymoma**” is restricted to tumors of thymic epithelial cells. Such tumors typically also contain benign immature T cells (thymocytes).

The WHO has created a classification system based on histology for thymomas, but its clinical utility remains uncertain. We will instead use a classification that relies on the most important prognostic features, the surgical stage and the presence or absence of overt cytologic features of



**Figure 13-41** Thymoma. **A**, Benign thymoma (medullary type). The neoplastic epithelial cells are arranged in a swirling pattern and have bland, oval to elongated nuclei with inconspicuous nucleoli. Only a few small, reactive lymphoid cells are interspersed. **B**, Malignant thymoma, type I. The neoplastic epithelial cells are polygonal and have round to oval, bland nuclei with inconspicuous nucleoli. Numerous small, reactive lymphoid cells are interspersed. The morphologic appearance of this tumor is identical to that of benign thymomas of the cortical type. In this case, however, the tumor was locally aggressive, invading adjacent lung and pericardium.