

DISORDERS OF THE THYMUS AND SPLEEN

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DISORDERS OF THE THYMUS

As is well known, the thymus has a crucial role in T cell differentiation.-----→ lymphomas, particularly those of T cell lineage.

The focus here is on the two most frequent (albeit still uncommon) disorders of the thymus:

- 1- Thymic hyperplasia.
- 2- Thymoma.

Thymic Hyperplasia

- ▶ Thymic enlargement often is associated with the presence of lymphoid follicles, or germinal centers, within the medulla.
- ▶ These germinal centers contain reactive B cells, which are only present in small numbers in normal thymuses.
- ▶ Thymic follicular hyperplasia is found in most patients with *myasthenia gravis* and sometimes also occurs in other autoimmune diseases, such as SLE and rheumatoid arthritis.

Thymoma

- ▶ Thymomas are tumors of thymic epithelial cells.
- ▶ Several classification systems for thymoma based on cytologic and biologic criteria have been proposed.
- ▶ One simple and clinically useful classification is as follows:
 - Benign or encapsulated thymoma: cytologically and biologically benign
 - Malignant thymoma
 - * *Type I*: cytologically benign but infiltrative and locally aggressive
 - * *Type II* (thymic carcinoma): cytologically and biologically Malignant

MORPHOLOGY

- ▶ Macroscopically, thymomas are lobulated, firm, gray-white masses up to 15 to 20 cm in dimension. Most appear encapsulated, but 20% to 25% penetrate the capsule and infiltrate perithymic tissues.
- ▶ Microscopically, virtually all thymomas are composed of a mixture of epithelial tumor cells and nonneoplastic thymocytes (immature T cells).
- ▶ In benign thymomas, the epithelial cells are spindle or elongated and resemble those that normally populate the medulla. As a result, these are sometimes referred to as medullary thymomas. In other tumors, there is an admixture of the plumper, rounder, cortical-type epithelial cells; this pattern is sometimes referred to as a mixed thymoma. The medullary and mixed patterns account for 60% to 70% of all thymomas.

MORPHOLOGY

Malignant thymoma type I is cytologically bland but locally invasive; it accounts for 20% to 25% of all thymomas. These tumors also occasionally (and unpredictably) metastasize.

- They are composed of varying proportions of epithelial cells and reactive thymocytes. The epithelial cells usually have abundant cytoplasm and rounded vesicular nuclei, an appearance similar to normal thymic cortical epithelial cells; spindled epithelial cells are sometimes present as well. The epithelial cells often palisade around blood vessels. The critical distinguishing feature is the penetration of the capsule with the invasion of surrounding structures.
- ▶ **Malignant thymoma type II** is perhaps better thought of as a form of thymic carcinoma. These tumors account for about 5% of thymomas. Macroscopically, they usually are fleshy, obviously invasive masses that often metastasize to such sites as the lungs. Microscopically, most resemble squamous cell carcinoma.

Clinical Features

Thymomas are rare.

- They may arise at any age, but most occur in middle-aged adults. -- In a large series about 30% were asymptomatic; 30% to 40% produced local manifestations such as cough, dyspnea, and superior vena cava syndrome.

- The remainder were associated with a systemic disease,
INCLUDING:

* Most commonly myasthenia gravis.

* Thymomas may be associated with several other paraneoplastic syndromes. These include in rough order of frequency , pure red cell aplasia, hypogammaglobulinemia, and multi organ autoimmunity. The latter bears resemblance with graft versus host disease.]

Disorders of the Spleen

Congenital anomalies

Cysts

Inflammation

Hypersplenism

Thrombocytopenic purpuras

Hemolytic anemia

Other non-neoplastic disorders

Hematolymphoid tumors and tumorlike conditions, (Non-Hodgkin lymphoma, Hodgkin lymphoma, Leukemias, Myeloproliferative neoplasms, Mastocytosis, Other hematolymphoid conditions)

Vascular tumors.

Metastatic tumors

Blunt trauma

- ▶ Blunt trauma to the abdomen and surgical intervention within the abdominal cavity are the two most common factors responsible for rupture of the normal spleen.
- ▶ In most instances, hemoperitoneum is an immediate consequence, leading to an emergency splenectomy.
- ▶ In about 15% of the cases, the rupture is “delayed” anywhere from 48 hours to several months.

Disorders of the Spleen

SPLENOMEGALY

The spleen is frequently involved in a wide variety of systemic diseases. In virtually all instances the spleen responds by enlarging (splenomegaly), an alteration that produces a set of stereotypical signs and symptoms.

11 Disorders may be grouped according to the degree of splenomegaly that they characteristically produce:

• 1- *Massive splenomegaly* (weight > 1000 g)

- 1- Myeloproliferative neoplasms (CML, primary myelofibrosis);
- 2- certain indolent leukemias (CLL and hairy cell leukemia);
many lymphomas;
- 3- infectious diseases (e.g., malaria);
- 4- Gaucher disease.

• *Moderate splenomegaly (weight 500–1000 g)*

- 1-Chronic congestive splenomegaly (portal hypertension or splenic vein obstruction);
- 2-acute leukemias;
- 3-disorders with extravascular hemolysis (hereditary spherocytosis, thalassemia major, autoimmune hemolytic anemia);
- 4- amyloidosis;
- 5-Niemann-Pick disease;
- 6- many infections, including infective endocarditis, tuberculosis, and typhoid;
- 7-sarcoidosis;
- 8-metastatic carcinoma or sarcoma

Mild splenomegaly (weight < 500 g)

1-Acute splenitis;

2-acute splenic congestion;

3- infectious mononucleosis;

3- miscellaneous disorders, including septicemia, systemic lupus erythematosus.

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- ▶ A chronically enlarged spleen often removes excessive numbers of one or more of the formed elements of blood, resulting in anemia, leukopenia, or thrombocytopenia. This is referred to as *hypersplenism*.
- ▶ In addition, platelets are particularly susceptible to sequestration in the interstices of the red pulp; as a result, *thrombocytopenia* is more prevalent and severe in persons with splenomegaly than is anemia or neutropenia.

THANK
YOU!