奇美醫學中心胸腔內,外,放射,病理科
臨床病例綜合討論會

1 題目: Thymoma in CXR
2 聯絡人: 李博洧
3 主持人：柯獻欽
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5 地點： 奇美醫學中心 10 樓空橋討論室

摘要:

Thymomas are tumors consisting of an admixture of thymic epithelial cells and reactive lymphocytes, with both circumscribed and invasive patterns of growth.

Cystic change is common and in some cases the majority of the tumor is cystic. Foci of calcification, hemorrhage and necrosis may also be seen.

It is the presence or absence of spread beyond the capsule rather than the histological appearance within the thymus that determines whether a tumor is labeled benign or malignant by the pathologist. For this reason the term "invasive thymoma" is used to describe any tumor that has spread beyond the capsule. 15-40% of thymomas turn out to be invasive.

The average age at diagnosis is approximately 50, or earlier in those who present with myasthenia gravis, with virtually equal frequency in men and women. Thymomas are unusual below the age of 20.

Most patients with noninvasive thymoma have no clinical symptoms due to the tumor itself. When symptoms occur, they are due to local compression causing chest pain, cough, dyspnea, dysphagia, hoarse voice, or superior vena cava syndrome.

Approximately 40% of patients with thymoma have myasthenia gravis, and the incidence of thymoma in patients with myasthenia gravis is 15%.

Thymectomy, regardless of whether or not the thymus harbors a thymoma, improves the myasthenia in some patients. Thymectomy for relieving myasthenia gravis is less effective in patients with thymoma than it is for patients without thymoma. Thymomas should be removed surgically, preferably when still confined to the thymus.

A hematologic cytopenia, notably pure red cell aplasia or aplastic anemia, is seen in 21-50% of patients and thymoma is associated with hypogammaglobulinemia in up to 10% of patients. There is a variety of conditions associated with thymoma.

Most thymomas arise in the upper anterior mediastinum. They are usually found anterior to the ascending aorta, above the right ventricular outflow tract and the main pulmonary artery, and project into one or other hemithorax. They are usually spherical or have a lobulated border. Punctate, curvilinear or ringlike calcification is frequent in both benign and invasive thymomas.

At CT, they are usually of homogeneous density and enhance uniformly, but they may appear cystic with discrete nodular components.

Before age 40, and particularly before 30, diagnosing a small thymoma can be difficult because the normal gland is variable in size and in myasthenia gravis the associated hyperplasia may lead to a bulky gland. Invasive thymomas invade the mediastinal fat and eventually spread to the pericardium and pleura.

Until mediastinal invasion has occurred, it is not possible to distinguish benign from invasive thymoma even with CT. Adjacent mediastinal fat planes are preserved when tumors are confined to the thymus, whereas they may be obliterated by invasive tumors. Preservation of adjacent fat planes does not exclude capsular invasion, but when these fat planes are preserved, extensive

invasive disease is unlikely. Pleural deposits "drop metastasis" may be so extensive that they mimic malignant pleural mesothelioma. In some patients there is no visible mediastinal tumor.

The anterior compartment of the mediastinum includes the aorta and its branches, the great veins, thymus, lymphatic tissue, and lung. While many patients are asymptomatic, some patients may have signs or symptoms of mass effect on the surrounding organs, such as cough, chest pain, shortness of breath, or superior vena cava syndrome. They may also have "B" symptoms of fever, night sweats, or weight loss, or they may have symptoms of conditions that are associated with anterior mediastinal tumors, such as myasthenia gravis (thymoma), pure red cell aplasia (thymoma), thyrotoxicosis (thyroid abnormality), or hyperparathyroidism (parathyroid abnormality).

The differential diagnosis offered above is often referred to as the 4 "T's" - teratoma, thymoma, thyroid abnormality, or terrible lymphoma. This differential can be much extended, to include cardiovascular masses such as aortic aneurysm, cardiac tumor, paricardial cyst, etc., or metastases of other primary tumors, or tumors of phrenic or vagus nerves, or a number of other lesions.

The radiographic appearance of a mediastinal mass can offer some clues to the diagnosis. The radiographs in this case are suggestive of thymoma - the mass is characteristically located between the sternum and heart/great vessels, it is well-circumscribed, and it demonstrates the "sulcus sign." The sulcus sign indicates that the mass is firm, creating a sulcus where the mass meets the sternum or other firm structure, rather than being flattened against the structure and obliterating the sulcus. The well-encapsulated appearance also suggests benign thymoma. Malignancy of thymoma is defined by invasive spread, although thymomas in general are considered slow-growing. Malignant thymomas spread contiguously, and can spread around the pleura and cause multiple pleural masses resembling mesothelioma. Less extensive spread is difficult to rule out. Chest CT may be performed to more sensitively identify invasiveness, but even intraoperatively invasiveness may be difficult to identify. Staging is usually performed with the Masaoka system, but the GETT (French Study Group on Thymic Tumors) is also used. Invasion is the most important prognostic factor with thymoma, with 5-year survival of 70% (50% with local invasion, 75% without invasion), and 10-year survival of 50% (30% with invasion, 60% without invasion).

The age of the patient may be used to predict the character of a mass. Thymoma occur most frequently in patients aged 45-50, rarely in patients less than 20, so this patients age of 28 would not have predicted thymoma as being the most likely lesion. Over all age groups, thymoma is the most common primary tumor of the anterior mediastinum, with an incidence of 0.13 in 100,000. In patients aged 20-40, Hodgkin's and non-Hodgkin's lymphoma and germ cell tumors are most common. These lesions are also most likely to be malignant.

Thymoma has a significant association with myasthenia gravis - 40% of patients with a thymoma have myasthenia gravis, and 15% of patients with myesthenia gravis have a thymoma. Thymectomy usually achieves some attenuation of the severity of myesthenia gravis of these patients. 5-15% of patients with thymoma also have pure red cell aplasia, usually occurring in older women - thymectomy resolves this condition in up to 40% of patients. Immunodeficiency resulting from

hypogammaglobulinemia and white cell aplasia are much less common, occurring in less than 5% of thymoma patients.

Primary treatment for thymoma is surgical resection. Difficulties that may be encountered intraoperatively may include invasion, adhesion of tumor to adjacent structures, or extension of tumor along phrenic nerves which increases the risk of repiratory impairment if these nerves are damaged during dissection. Adjuvant radiation therapy has not been shown to be of benefit after resection of a well-encapsulated, non-invasive tumor. However, adjuvant radiation is often beneficial when used after incomplete resection, after complete resection of invasive tumor, or in unresectable disease. It is sometimes used after total resection in which adhesions were noted. Thymoma is a chemotherapy-sensitive tumor, and chemotherapy may be used in patients with locally invasive or bulky tumors prior to resection. Chemotherapy is also traditionally used for patients with unresectable or metastatic thymoma, or with thymic carcinoma.