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Anterior Mediastinal Tumor

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－Abstract－

Primary anterior mediastinal neoplasms comprise a diverse group of tumors and account for 50% of all mediastinal masses. Thymic epithelial neoplasm are most common and classified into thymoma, invasive thymoma, and thymic carcinoma. Neuroendocrine differentiation of thymic epithelial neoplasm are rare malignancies. Germ cell tumor (GCT) is second most common anterior mediastinal tumor and most of them are mature cystic teratoma. Malignant mediastinal GCT are rare than benign. Primary thoracic lymphoma is rare than thoracic involvement of systemic lymphoma and most common location of primary thoracic lymphoma is anterior mediastinum. The clinical and radiologic appearance of the most common masses are reviewed.

Key Words: Mediastinum, Tumor, CT

Introduction

The mediastinum is located in the central portion of the thorax, between the two pleural cavities, the diaphragm and the thoracic inlet.[1] It is usually divided into anterior, middle, and posterior compartments to help categorize tumors and diseases according to their site of origin and location. The anterior mediastinum is defined as the region posterior to the sternum and anterior to the heart and brachiocephalic vessels. It extends from the thoracic inlet to the diaphragm and contains the thymus gland, fat, and lymph nodes.[2] Primary mediastinal tumors are a heterogeneous group of neoplastic, congenital, and inflammatory conditions.[3] This article will review primary

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anterior mediastinal tumors, which comprise approximately one half of all mediastinal masses including thymoma, thymic carcinoma, thymic neuroendocrine neoplasm including thymic carcinoid and small cell carcinoma, germ cell tumors, and lymphoma

Thymoma

Thymomas are benign or low-grade malignant tumors arising from the thymic epithelium and are characterized by the presence of a variable number of immature, nonneoplastic T cells.[4] Thymomas represent 20% of all mediastinal neoplasms in adults; they are the most common anterior mediastinal primary neoplasm in adults but account for less than 5% of mediastinal tumors in children.[5] The peak prevalence of thymoma is during the fifth and sixth decades of life. Thymomas have no sex predilection. Patients with thymoma are frequently asymptomatic; however, 20%~30% of patients have pressure-induced symptoms such as cough, chest pain, dyspnea, dysphagia, hoarseness, or superior vena cava syndrome. One-third to one-half of thymoma patients develop myasthenia gravis.[6, 7]

At radiography, thymomas typically appear as sharply marginated retrosternal areas of increased opacity with smooth or lobulated borders. Thymomas may project to either side of the mediastinum and obscure the heart border. On CT scans, thymomas

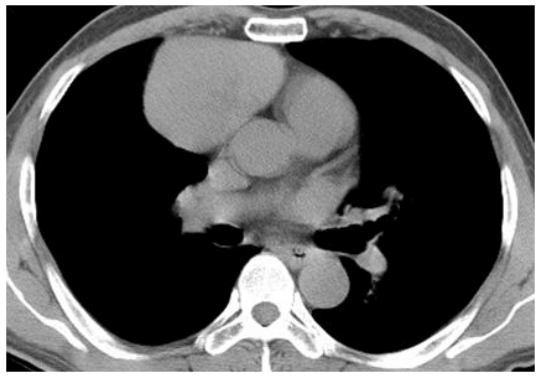

Fig. 1. 52 years old male with incidentally found mass. About 8cm sized well circumscribed soft tissue mass is seen at anterior to ascending aorta.

usually appear as homogeneous solid masses with soft-tissue attenuation and well-demarcated borders (Fig. 1). Thymomas may be oval, round, or lobulated and usually do not conform to the shape of the thymus. Large thymomas may show areas of cystic or necrotic degeneration. Calcification may be present in the capsule or throughout the mass (Fig. 2). Well-defined fat planes between

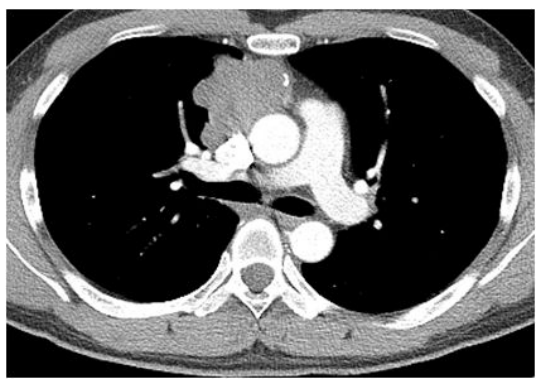

Fig. 2. 34 years old male with fatigue for a month. Lobulating mass with focal calcification. The margin between mass and mediastinal structure is unclear, but on surgery, no tissue invasion was proved.

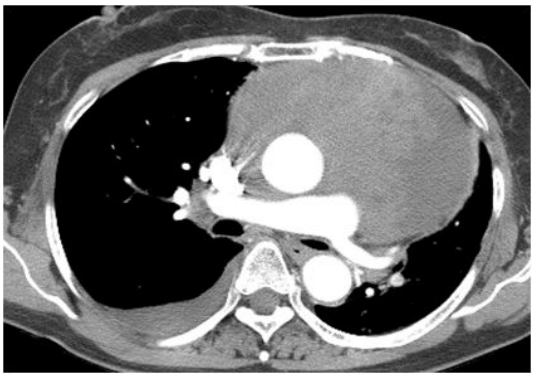

Fig. 3. 40 years old female with chest pain and discomfort. More than 10cm sized mass with inner necrosis is demonstrated at anterior mediastinum. Tissue plane between mass and thoracic great vessels is unclear. Right side effusion also represent tumor spreading and proved by pathology.

the thymoma and adjacent structures generally indicate absence of extensive local invasion. However, minimal invasion may escape detection at imaging. Certain findings, such as encasement of mediastinal structures, infiltration of fat planes, and an irregular interface between the mass and lung parenchyma, are highly suggestive of invasion (Fig. 3). Pleural thickening, nodularity, or effusion generally indicates pleural invasion by the thymoma.

Thymic carcinoma

Thymic carcinomas account for about 20% of thymic epithelial tumors.[8] Thymic carcinomas behave more aggressively than invasive thymomas and are more likely to metastasize to distant sites.[7] Unlike thymomas,

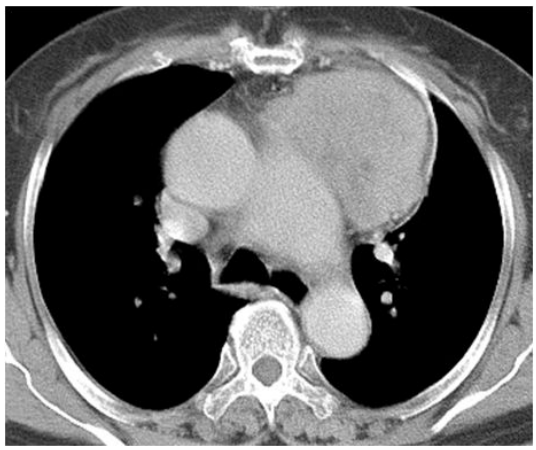

Fig. 4. 66 years old female with cough for several months. About 8cm sized lobulating mass is seen left anterior to ascending aorta. Tissue plane between mass and ascending aorta is unclear. Needle biopsy proved thymic carcinoma, squamous cell type.

in which neoplastic cells show morphologic and immunohistochemical features characteristic of thymic epithelial cells, the epithelial cells of thymic carcinomas show overt atypia.[9] Thymic carcinoma is uncommon in adults and even rarer among children. The mean age of patients with thymic carcinomas is 50 years.[7] Thymic carcinomas freely invade adjacent structures and often cause compressive symptoms. Unlike thymomas, thymic carcinomas rarely cause paraneoplastic syndromes such as myasthenia gravis.

On CT scans, thymic carcinomas typically appear as large, multilobulated masses (Fig. 4) that may contain areas of low attenuation or calcification. It is difficult to distinguish thymic carcinomas from thymomas solely on the basis of imaging findings. Nevertheless,

some features such as distant metastasis or mediastinal lymphadenopathy suggest thymic carcinoma.[7]

Neuroendocrine tumors of the thymus

Neuroendocrine tumors are uncommon tumors of variable malignant potential. Histologically, these range from relatively benign(thymic carcinoid) to highly malignant tumors(small cell carcinoma of the thymus).[8] Thymic carcinoid tumor is the most common of this group of tumors.[10] They occur over a wide age range (mean age, 43 years) and occur three times as frequently in men as in women.[11, 12] Patients with thymic carcinoids often present with endocrine disorders such as Cushing syndrome (25%~40% of patients) or multiple endocrine neoplasia types Ⅰ and Ⅱ (about 20% of patients).[7]

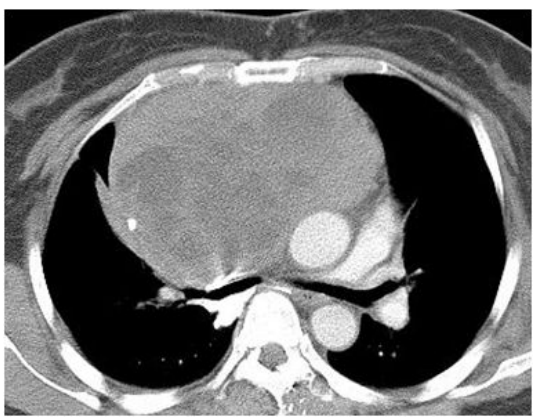

Fig. 5. 42 years old female with chest pain for 2 months. Well marginated heterogeneous enhancement of mass and eccentric calcification are seen. Thymic small cell carcinoma was proved on pathology.

Neuroendocrine tumors of the thymus manifest on chest radiographs as well- or ill-defined anterior mediastinal mass and may contain calcification (Fig. 5).[13]

Thymic lymphoma

Thymic involvement of lymphoma is more common in clinical setting. Hodgkin's disease accounts for the majority of cases of thymic lymphoma. The overall incidence of thymic involvement by Hodgkin's disease is difficult to determine; estimates have varied from 30 to 56%. In most cases, both mediastinal nodes and the thymus are affected; isolated thymic disease is quite rare.[14, 15]

Thymic lymphoma manifests on chest radiography as a unilateral or bilateral, frequently well-circumscribed, mediastinal

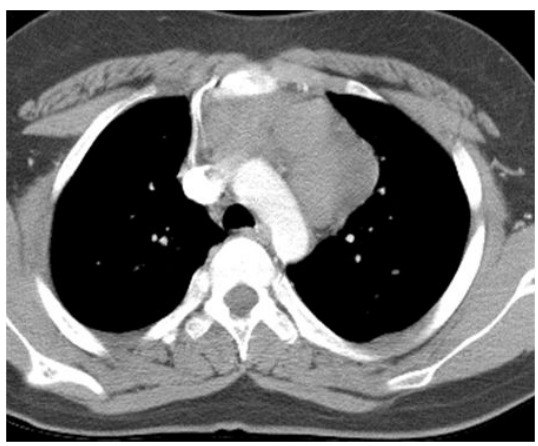

Fig. 6. 14 years old male with chest pain for several months. More than 8cm sized anterior mediastinal mass with containing multiple small masses. Low density area represent tumor necrosis. Hodgkin's disease was proved on pathology.

mass. On CT scan, thymic lymphoma is seen as either diffuse thymic enlargement or as solitary or multiple thymic masses (Fig. 6). Homogeneous enlargement of the thymus in the presence of mediastinal or hilar lymphadenopathy usually suggests lymphoma. Cystic changes in the thymus with or without calcification are seen at CT in about 20% of patients before they receive therapy.[7, 15)]

Thymic germ cell tumors

Germ cell tumors usually arise along the midline, from the pineal region to the sacrococcygeal region. Both pure and mixed germ cell tumors can develop within or near the thymus. The anterior mediastinum is the most common site of extragonadal germ cell tumors.[6)] More than 80% of mediastinal germ cell tumors are benign, the most common being benign teratoma.[7)]

Germ cell tumors account for 1%~15% of mediastinal tumors in adults and about 25% of mediastinal tumors in children.[12, 15)] Although benign germ cell tumors have no sex predilection, malignant germ cell tumors tend to occur in men, with a peak prevalence in the third decade of life.[7, 16)]

Mature teratomas are frequently found incidentally at conventional radiography. Although most mature teratomas are asymptomatic, the larger ones may cause compressive symptoms and even erode through the tracheobronchial tree. In contrast, patients with malignant germ cell tumors are usually symptomatic.[6, 16)]

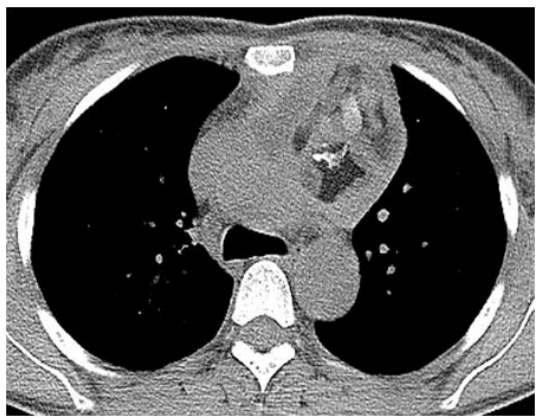

Fig. 7. 45 years old female with acute onset chest pain. More than 7cm sized well marginated mass is seen anterior mediastinum which contain fat and bone tissue. Focal capsular rupture was detected by surgery.

Chest radiography of teratomas typically reveals large, round, lobulated masses. Since teratomas contain derivatives of all three germinal layers, the presence of teeth, bone, or calcification (seen in 20% of cases) is diagnostic.[6, 16)] On CT images, teratomas typically appear as a combination of fluid or fat cysts, soft tissue, calcification, and bone or teeth (Fig. 7). About 90% of mature teratomas contain fluid and about 75% contain fat. A fat-fluid level within the mass is diagnostic of teratoma but is seen in only about 10% of cases. Teratomas are usually clearly demarcated and surrounded by a capsule, which may show rim enhancement. Benign teratomas are typically smooth, clearly defined, and cystic; 90% of them contain fat. In contrast, malignant teratomas

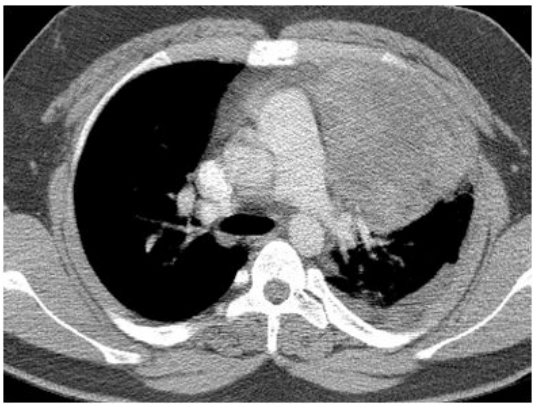

Fig. 8. 16 years old male with chest pain for several months. More than 8cm sized heterogenous enhancing mass at left hemithorax. Left side effusion is also demonstrated. Needle biopsy proved yolk sac tumor.

are nodular and poorly defined and have more solid components than do benign teratomas; only 40% of malignant teratomas contain fat. Compression of adjacent structures, a thick enhancing capsule, and areas of necrosis or hemorrhage are other features of malignant teratomas (Fig. 8).[6, 7]

Conclusion

Many mediastinal masses are discovered on chest radiographs obtained for other reasons. Some patients will come to clinical attention with vague chest complaints.

The most common primary anterior mediastinal tumors are thymoma, teratoma, and lymphoma. All other lesions are extremely rare.

Discovery of a mediastinal mass is usually alarming. Radiologists play a major role in differentiating normal variants, ectopic thymic tissue, and nonneoplastic conditions such as rebound hyperplasia from neoplastic conditions. A thorough knowledge of the embryology and anatomy of the thymus, normal variations and ectopic locations of the thymus, and its dynamic changes are essential to prevent performance of unnecessary invasive procedures.

한글초록

원발성 전 종격동 종양은 종격동 종양의 반 이상을 차지하고 그 종류도 다양하다. 흉선 상피 종양이 가장 흔하고 악성 흉선종은 드물다. 생식세포종은 두 번째로 흔한 전 종격동 종양으로 보다 어린나이에 발생하며 대부분 양성이다. 임파종은 대부분 전신성 질환의 흉부 침습 형태로 나타나나 원발성 흉부 임파종은 Hodgkin씨 병이 많다. 저자는 대표적인 전종격동 종양의 임상증상과 영상의학적 소견을 소개한다.

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