Mucoepidermoid Carcinoma of the Thymus: A Review

Singh VP¹, Kannan N²

¹Department of Surgical Oncology, Command Hospital (Eastern Command), Kolkata, India
²Department of Surgical Oncology, Command Hospital (Southern Command), Pune, India

Abstract: Mucoepidermoid carcinoma of the thymus is an extremely rare malignant mediastinal neoplasm, and to our knowledge, only 13 cases have been reported. We report a case of mucoepidermoid carcinoma of the thymus that was seen in a 37-yr-old man with right chest pain and non productive cough. Chest CT scan showed a huge, cystic mass having a focal solid portion with direct invasion of the adjacent anterior chest wall and pericardium in the anterior mediastinum. Mucoepidermoid carcinoma of the thymus should be included in the differential diagnosis for masses of the anterior mediastinum associated with extensive cystic changes, although the carcinoma is exceedingly rare.

Keywords: Mediastinal Neoplasms, Thymus Neoplasms, Carcinoma, Mucoepidermoid

1. Introduction

Thymic carcinomas are unusual malignant neoplasms that have wide variety of morphologic appearances. Mucoepidermoid carcinomas of the thymus are extremely rare malignant mediastinal neoplasms and account for 2% of thymic carcinomas (1, 2). Until now, radiologic reports of mucoepidermoid carcinoma of the thymus have been rare, but the described gross findings are multilocular cystic structures with focal areas of induration within the walls of the cyst or well-circumscribed homogeneous tumor masses (3). We report a case of mucoepidermoid carcinoma of the thymus with radiologic and histologic findings.

2. An Interesting Case

A 37-yr-old male was admitted to a peripheral hospital complaining of right chest pain. The physical examination and laboratory studies were unremarkable. He was transferred to this tertiary care centre for further management.

Erect posteroanterior and lateral radiographs of the chest showed a well marginated anterior mediastinal mass in the entire right lung zone, compressing the lower trachea, right bronchus, and heart toward the left side (Fig. 1). CT scan of the chest demonstrated a 20×16×10 cm-sized, cystic mass with focal heterogeneously enhanced solid portion anteromedially in the anterior mediastinum. Direct invasion of the anterior chest wall and pericardium by the mass was suggested, because pericardial fat plane was obliterated and the margin between the mass and the adjacent right anterior chest wall and pericardium was indistinct (Fig. 2). Minimal amount of right pleural effusion and pericardial effusion were associated.

Figure 1

PA radiographs of the chest shows a huge, well marginated, homogeneous mass in the anterior mediastinum displacing the lower trachea, bronchus and heart contralaterally.
At surgery, after aspiration of fluid within the cystic mass, pericardial invasion was noted. Thus, the invaded portion of pericardium was also resected out and a bovine pericardium patch was placed. Histologically, squamoid tumor cells formed cords or solid sheets within the fibrous stroma admixed with mucin-secreting epithelium lining gland-like spaces (Fig. 3). The final pathologic diagnosis was well differentiated mucoepidermoid carcinoma of the thymus. The patient is on follow up and doing well.

Figure 2: (A) Chest CT scan at the level of aortic origin demonstrates a huge, low attenuating, cystic mass in anterior mediastinum with internal septa and heterogeneous solid area anteromedially. (B) Contrast enhanced CT scan shows the focal solid mass area

Postoperative photomicrographs of the lesion (a) The tumor was composed of nests of epidermoid cells with mucin-producing areas (left side of the figure) and lymphocyte-rich areas (right side of figure). They represented the MEC component and thymoma component, respectively. (b) High magnification showed the mucin-producing cells were surrounded by epidermoid cells in MEC component. (c) Invasive nests of well-differentiated epidermoid cells, mucin-producing and intermediate cells were observed in MEC areas. Bands of fibrous connective tissue were observed in the tumor. (d) The epidermoid cells were observed to be positive for CK5/6, but the mucin-producing cells were CK5/6-negative. (e) MEC component was negative for CD5. (f) The Alcian blue-positive material was seen in lumen of gland structure and the mucin-producing cells within the nest of epidermoid cells. (a, H & E staining with original magnification of 200 ×; b-c, H & E staining with original magnification of 400 ×; d-e, immunohistochemical staining with original magnification of 400 ×; f, Alcian blue staining with original magnification ×400).

3. Discussion

The thymic carcinomas are a heterogeneous group of aggressive epithelial malignancies that have a strong propensity for early local invasion and wide spread metastases. Squamous cell carcinoma and lymphoepithelioma-like carcinoma are the most common cell types and usually occur in middle-aged men with a mean age of 46 yr (4-6). Radiologically, thymic carcinomas commonly manifest as large, poorly defined, infiltrative anterior mediastinal masses and associated with areas of necrosis, hemorrhage, calcification, or cyst formation (2, 6, 7).

Mucoepidermoid carcinoma was first recognized as a distinct pathologic entity in salivary glands by Stewart et al. in 1945. Since then, this tumor has been identified in several other organs, including lung, esophagus, anus, cervix, and skin. Although extremely rare, mucoepidermoid carcinomas of the thymus have also been described (3). So far, only thirteen cases have been reported in the English literature (3, 8, 9). The patients ranged in age from 17 to 66 yr, with a mean age of 35 yr on reported cases (3). Clinically, most patients were asymptomatic or had symptoms of chest discomfort, retrosternal pain and dyspnea.

Radiologic reports of mucoepidermoid carcinoma of the thymus were rare. But two distinct gross findings, as described by Moran et al., are multilocular cystic structures varying in size with focal areas of induration within the walls of the cyst and well-circumscribed, homogeneous tumor masses (3). Our case presented as a 20 cm-sized, cystic mass with focal heterogeneously enhanced solid portion anteromedially in the anterior mediastinum. The invasion of anterior chest wall and pericardium was
suspected due to indistinct margin between the mass and adjacent right anterior chest wall and pericardium, and surgically proved.

Histologically, the lesions showed a spectrum of features that ranged from those of well-differentiated, to moderately well-differentiated, to poorly differentiated mucoepidermoid carcinoma, with sheets and solid islands of squamoid cells admixed with mucin-secreting epithelium lining gland-like spaces (3). The histogenesis of these tumors is still unclear; however, the demonstration of transitions between the tumor cells and the epithelium lining the cysts as well as residual non-neoplastic thymic remnants within the walls of the cysts in four of six cases supports the theory that these tumors arise from thymic epithelium (3).

Differential diagnoses of masses of the anterior mediastinum associated with extensive cystic changes include thymic cysts, thymomas, teratomas, seminomas, Hodgkin's disease, and metastasis (10-14). And also mucoepidermoid carcinoma of the thymus should be included in the differential diagnosis, although the carcinoma is an exceedingly rare anterior mediastinal malignant tumor (3). It is very difficult to differentiate from each other by only radiologic findings (10-14). However, mucoepidermoid carcinoma of the thymus should be considered when a large cystic mass with focal induration within the anterior mediastinum on CT scan is noted.

4. Dedication

Dedicated to my teacher Dr N Kannan, who expired in 2018 due to Glioblastoma Multiforme. Conflicts of Interest: NIL

References