Mature teratoma with somatic-type solid malignancy in the mediastinum: A case report of rare and poor prognosis tumour

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ABSTRACT

Background: Germ cell tumor with somatic-type solid malignancy is a germ cell tumor accompanied by a non-germ cell, the somatic-type malignant component in the form of sarcomas or carcinomas. These germ cell tumors are rare, accounting for only about 2% of all germ cell tumors in men. About 25-30% of these tumors occur in the mediastinum. These tumors are often located in the anterior mediastinum and occur in adult males with a peak incidence between 20-40 years.

Case description: A 19-year-old female patient came with chief complaints of chest pain and shortness of breath about three months ago. Chest CT scan images showed a right lung reactive pneumonia with suspicious atelectasis at the second and third segments of the right lung. The image also showed a tumor mass with heterogeneous density at the mediastinum. Then thoracotomy surgery and tumor resection were performed. Macroscopically, there was a multicystic tumor mass measuring 14.0x11.0x6.5 cm, partly solid with 7.0x5.0x5.0 cm. Microscopic examination of the tumor mass showed three components of the germ layer, namely ectodermal, mesodermal and endodermal. No immature component appears. The tumor also contains neoplastic epithelial cells that form a solid, cribiform, acinic and papillary structure in the extracellular mucinous pool.

Conclusion: Based on the clinical finding, radiologic, and routine histopathological examination, this case was concluded as mature teratoma with somatic-type solid malignancy in the form of mucinous adenocarcinoma.

Keywords: mature teratoma, mucinous adenocarcinoma, anterior mediastinum, somatic-type.


INTRODUCTION

Germ cell tumors account for approximately 15% of mediastinal tumors in adults, with teratoma being the most common type. Teratomas may undergo malignant transformation to a non-germ cell or malignant somatic component in the form of sarcomas or carcinomas. Many of the germ cell tumors with somatic-type malignancies are mixed type or teratoma, but these tumors are also found in pure yolk sac tumors and seminomas.1,2

Somatic-type solid malignancies are more common in mediastinum than primary gonadal or retroperitoneal tumors. Overall, germ cell tumors with somatic-type malignancy are extremely rare,3,4,5,6 accounting for only about 2% of all germ cell tumors in men.5,6 However, about 25-30% of these cases occur in the mediastinum, and somatic-type malignancies appear to occur more frequently in mediastinal tumors than in primary gonadal or retroperitoneal tumors. With a few exceptions, these tumors occur more frequently in male adults, more frequently in older patients with a peak incidence between the ages of 20-40 years, and usually with a long-standing mature teratoma. However, some reports also occur in females, although very few. Somatic-type solid malignancies can occur in primary germ cells tumor or only in metastases.5,6,7 The epithelial malignancies associated with germ cells tumor are mostly colonic type adenocarcinomas, adenosquamous carcinoma, and squamous cell carcinoma.7 This paper reported the case of a 19 years old female diagnosed with a mediastinal tumor, suspicious of teratoma.

CASE REPORT

A woman, 19 years old, came to the Emergency Unit of the Sanglah General Hospital in Denpasar, bringing a referral from a private hospital with a suspicious diagnosis of a mediastinal tumor. The patient complained of chest pain three months before hospital admission, which worsened for about a week. The pain was felt in the middle of the chest. The patient also complained of shortness of breath that came and went, worsened when lying down and less when sitting and standing. History of cough and night sweats was refuted. On physical examination, it was found that the chest wall was asymmetric, and the sound of the right lung breath was absent. On blood laboratory tests, the results obtained β-HCG blood <0.10 m IU/ ml, AFP 4.8 µL.

Chest X-rays showed a blunt right pleural sinus. At the same time, the left side was sharp, visible enlargement of the right pleural space accompanied by minor fissure thickening, the heart was enlarged, and the lungs appeared to have an increased bronchovascular color. These features suggest cardiomegaly, right pleural effusion, and pulmonary edema. Chest CT scan showed a tumor mass with heterogeneous density in the pericardium.
area of the right atrium with a differential diagnosis of mediastinal mass. It showed right lung reactive pneumonia with suspicion of right lung atelectasis at the second and third segments (Figure 1).

The patient underwent thoracotomy surgery for tumor resection and chest tube insertion. A tumor mass was found in the anterior mediastinal region that compressed against the lung and filled 4/5 of the right hemithoracic cavity. The mass was released from the surrounding tissue along with the tumor capsule.

The tissue is then sent to the Anatomical Pathology Laboratory for histopathological examination. Tumor mass measured 14.0x11.0x6.5 cm, with grey, white, and brownish color and a hilted outer surface. The cut-section showed a blackish-brown discharge and multiple cysts with a 0.5-4.0 cm diameter. The specimen also contains a dermal plaque measuring 1.5x0.7 cm and a solid area measuring 7.5x5.0 cm.

Microscopic examination of the tumor mass showed three components of the germ layer, namely ectodermal, mesodermal and endodermal. The ectodermal component consists of the skin and its adnexa (Figure 2A). The mesodermal component consists of mature cartilage, mature fat tissue and smooth muscle (Figure 2B). The endodermal component consists of the gastrointestinal epithelium, pseudostratified respiratory epithelium and pancreatic tissue (Figure 2C and 2D). Another focus showed the presence of neoplastic epithelial cells that form papillary structure, cribiform, acinic, and solid pattern between the extracellular mucinous pool, and mild infiltration of lymphoplasmacytic inflammatory cells (Figure 3). Based on this histopathological examination, the patient was diagnosed as mature teratoma with somatic-type solid malignancy in the form of mucinous adenocarcinoma.

**DISCUSSION**

Mature teratomas show well-differentiated somatic elements, such as nerves, fat, skin, cartilage, and other mature tissue. A non-germ cell malignant tumor from a teratoma has been described as a teratoma with malignant transformation or a teratoma with somatic-type malignancies. These lesions, especially in the mediastinum, are very rare. The malignancies reported in mature mediastinal teratomas include sarcoma, adenocarcinoma, squamous cell carcinoma, and neuroendocrine neoplasms.1,8,11

Germ cell tumors with somatic-type solid malignancies show the same local symptoms as other mediastinal germ cell tumors, such as exertional dyspnea, chest pain, shortness of breath, and pleural effusion. These tumors are more often symptomatic in about 90% of cases than pure teratomas.7,9,12 In this patient, the symptoms were middle chest pain and shortness of breath that come and go. Complaints worsen when lying down and less when sitting and standing.

Radiologic images generally showed a solid mass representing sarcoma or carcinoma components associated with a cystic teratoma structure or lesions showing heterogeneous attenuation, a dominant area with increased soft tissue elements, calcification and massive necrosis. CT and MRI images show the variable size of the lesion with heterogeneous density and intensity. A cystic mass or lesion associated with malignancy is local thickening, calcification, solid areas, focal disruption with the invasion of nearby structures, or the presence of pleural or pericardial effusions.4,7,13 In this patient a tumor mass with heterogeneous density was the present in the area of the right atrial pericardium with a differential diagnosis at mediastinal, and also showed a right lung reactive pneumonia with suspicion of atelectasis at right lung second and third segment. Bronchoscopy showed normal vocal cords, sharp carina, narrowing of the mediastinum, smooth mucosa, not bleeding easily, and the conclusion was extra bronchial mass.

Germ cell tumors with somatic type malignancy more often occur in the anterior mediastinum.8,9,12 Germ cell tumors range in size from 6 cm to 30 cm.1 These tumors usually show a cystic cut surface and varied areas of focal necrosis. The malignant component of the somatic type appears soft and grayish or bleeding and often adheres to the surrounding mediastinal structures. The tumors are usually encapsulated, lobulated, heterogeneous, and multicystic in mature teratomas.4,7 In this patient, the longest size of the tumor mass was 14 cm, grey white and brownish. Discharge and multiple cysts with a diameter of 0.5-
The somatic-type solid malignancy was in the tumor mass showed mature teratoma and endodermal, without immature tissue. Somatic-type malignancies associated with germ cell tumors are predominantly of adenocarcinoma type. Extragonadal germ cell tumors, including mediastinal, arise due to arrested migration of totipotent primordial germ cells during fetal development. They encompass the entire range of histopathological subtypes of germ cell tumors seen in the gonads, with diverse outcomes, so treatment and clinical outcomes vary according to the histology. Teratoma with somatic-type solid malignancies, especially in the mediastinum, is associated with a poor prognosis with a high frequency of metastases and recurrences. Early resection of tumors has an important role in managing these tumors, whereas the use of adjuvant chemotherapy at any stage is controversial. Adjuvant chemotherapy following resection had no impact on overall survival, whereas histological findings and cancer stage were significant factors affecting survival rates. Only a minority of patients survive after chemotherapy and complete surgical resection of the tumors. Late local infiltration, metastases, and incomplete resection are poor prognostic factors, whereas the type of somatic malignancies at the first biopsy has no major impact on survival. Lin et al. reported a case of a patient with teratoma with malignant transformation in the mediastinum who achieved a good response to chemotherapy. This tumor is considered resistant to chemotherapy and radiotherapy. However, chemotherapy based on transformed histology has recently been advocated, which may help improve patients’ outcome. Early resection for treatment of mature teratomas, and careful inspection of pathological specimens of long-standing teratomas are very important. This patient underwent right posterolateral thoracotomy surgery, tumor resection and chest tube insertion, and four series of chemotherapy. The results of the last CT scan showed no mass at the mediastinal and lung, but there is no further follow-up data for this patient.

Germ cell tumors with solid malignancies of the somatic type may be difficult to distinguish from immature teratomas. If the clear atypia and infiltrative growth were found, it would favor somatic-type malignancies. In chemotherapy-induced, atypia is generally diffusely distributed throughout the tumor, while somatic-type malignancy is a focal process, often forming masses and invading surrounding structures.

CONCLUSION

Extragonadal germ cell tumor with somatic-type solid malignancy is rare and most often located in the anterior mediastinum. Epithelial malignancies associated with germ cell tumors are predominantly of adenocarcinoma type. This tumor is associated with a poor prognosis and a high frequency of metastases and recurrences. Identifying the malignant component within these neoplasms is important, and carefully sampling solid areas in a teratoma is required.

DISCLOSURES

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I Gusti Ayu Sri Mahendra Dewi is responsible for the study from the conceptual framework.

Conflict of interest
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