Sarcomatoid Carcinoma of The Mediastinal: A Rare Case of Giant Mass Thymic Carcinoma Subtype

Agus Andreas Santoso[1], Ngakan Putu Persama Putra[1], Ungky Agus Setiawan[1], Artono Isharanto[2], Dini Rachma Erawati[3], Hendy Setyo Yudhanto[4]

[1] Department of Pulmonology and Respiratory Medicine, Brawijaya University
[2] Supervisor of Thoracic and Cardiovascular Surgery Department, Saiful Anwar General Hospital, Brawijaya University
[3] Supervisor of Radiology Department, Saiful Anwar General Hospital, Brawijaya University
[4] Supervisor of Pathology Anatomy Department, Saiful Anwar General Hospital, Brawijaya University

Abstract

Introduction: Sarcomatoid carcinoma is a type of thymus carcinoma that contains partial or complete spindle cells. Incidence of Sarcomatoid Carcinoma 5-10% of all patients with thymic carcinoma. Carcinoma Sarcomatoid is a high progressive tumor, most patients will die after 3 years of diagnosis despite aggressive multi modality therapy.

Case Description: A 47-year-old woman with chest pain, shortness of breath, coughing, weight loss, enlarging lump in the neck and chest was felt for 1 month. Physical and supporting examination showed anterior superior mediastinal tumors and soft mass tissue in the right region of the colli. Based on the Trans Thoracal FNAB Guiding ultrasound and FNAB the anterior colli region concluded that Thymoma, thyroid oncocytic adenoma and Nodular colloid goiter with azkanasy cell proliferation. The patient had Partial Sternotomy (Hemiclamshell), thymectomy with the final diagnosis of Sarcomatoid Carsinoma.

Discussion: Sarcomatoid carcinoma is a type of thymic carcinoma that has both malignant epithelium (carcinomatous) and spindle cells (sarcomatose / sarcomatoid), generally with a transition between the two. This case was interesting because of the rare occurrence of Sarcomatoid Carcinoma plus a large size of tumor mass in this patient.

Keywords : Mediastinal Tumor, Sarcomatoid Carcinoma.

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1. Background

Thymus is a lymphatic organ that plays a vital role in the development and maturity of the immune system during childhood, especially T cells, which are instruments for the regulation of cellular immunity and B cells, which are instruments for the regulation of humoral immunity.[6]

As a child, it is disproportionate in size and will gradually be replaced by fat tissue and completely involuntarily in adulthood. Despite this, the thymus still functions at all times and throughout life. The word thymus comes from Greek, thymos which means "warty excrescence", and also means soul / spirit or spirit. The thymus is located in the anterosuperior mediastinum, which is a compartment bounded anteriorly by the sternum, laterally by the pleura and posterior by the vertebrae.[7] The mediastinum is the cavity between the right and left lungs. The

*Corresponding Author:
Agus Andreas Santoso (agusandreas@student.ub.ac.id)
RSUD Dr. Saiful Anwar Malang – Jl. Jaksa Agung Suprapto No. 2., Malang Indonesia

mediastinum contains the heart, arteries, veins, trachea, thymus gland, nerves, connective tissue, lymph nodes and ducts.[8]

Mediastinal tumors are found in patients aged 30 to 50 years. The signs and symptoms of mediastinal tumors vary widely and are non-specific. Although more than 60% showed symptoms, those asymptomatic were not detected at routine examination. Two-thirds of all mediastinal tumors are generally benign, and more than three-quarters of patients with asymptomatic mediastinal tumors have benign lesions. However, the majority of patients who are symptomatic have a malignant underlying process. Symptoms that arise vary according to the anatomical location. Although anterior mediastinal tumors can cause symptoms such as coughing, chest pain or shortness of breath, the onset of systemic symptoms such as fever or night sweats adds to the suspicion of lymphoma, while the onset of new myasthenia symptoms is suspected to be due to thymoma. Posterior mediastinal tumors, which are generally benign and neurogenic, usually show no symptoms.[9]

Diagnosis of mediastinal tumor often need a multidisciplinary team because of its complexities. Sarcomatoid carcinoma is a type of thymic carcinoma which partial or complete spindle cells. The incidence of sarcomatoid carcinoma is 5-10% in people with thymic carcinoma.Sarcomatoid carcinoma is a type of tumor that has high progressiveness, most patients will die after 3 years of diagnosis despite aggressive multi-therapy[1,2,3].

2. Case Report

Ms S, 47 yo, Javanese, farmer. Complaint : Right chest pain 2 months, shortness of breath, cough, loss of body weight in 1 month. Enlargment of node in the right anterior neck for 17 years. Physical examination: difuse enlargment of thyroid (Ø 10 cm), right chest: dullness, decrease of stem fremitus and decrease lung sound. Chest x-rays PA and Lateral D: mediastinal tumor with right pleural effusion (Fig. 1a,b). Thorax-CT: Mediastinum mass (20x20x9cm) expantion to the right side, right thyroid nodular mass with the calcification component(Fig.1c). Initial diagnosis: Anterosuperior mediastinal tumor, invasive thymoma stage III and Oncocytic adenoma dd. nodular colloid goiter with askanazy cell proliferation and then patient undergo partial sternotomy (hemiclamshell) and thymectomy. Transthoracic FNAB January 4th 2018 with the result Thymoma. FNAB Mass in Right Colli January 10th 2018 with the result Oncocytic adenoma dd nodular colloid goiter with askanazy cell proliferation. Partial sternotomy (hemiclamshell) dan thymectomy February 5th 2018 with the result Sarcomatoid carsinoma. Final diagnosis: spindle cell thymic carcinoma subtype sarcomatoid carcinoma with right lung invasion stage IIIA.

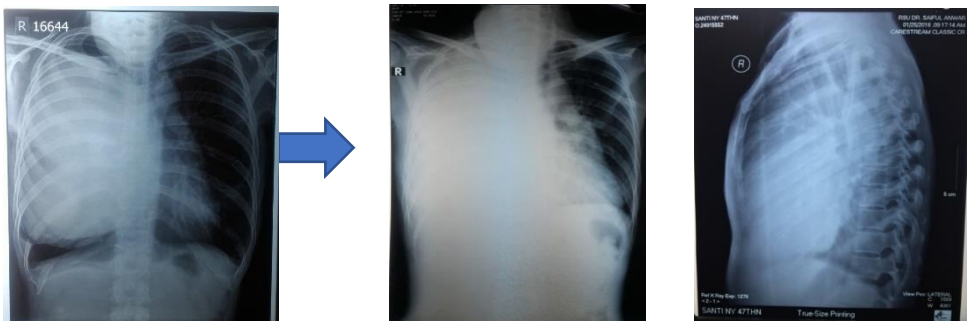

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Figure1. Comparison of Chest x-ray before surgery, getting worse.

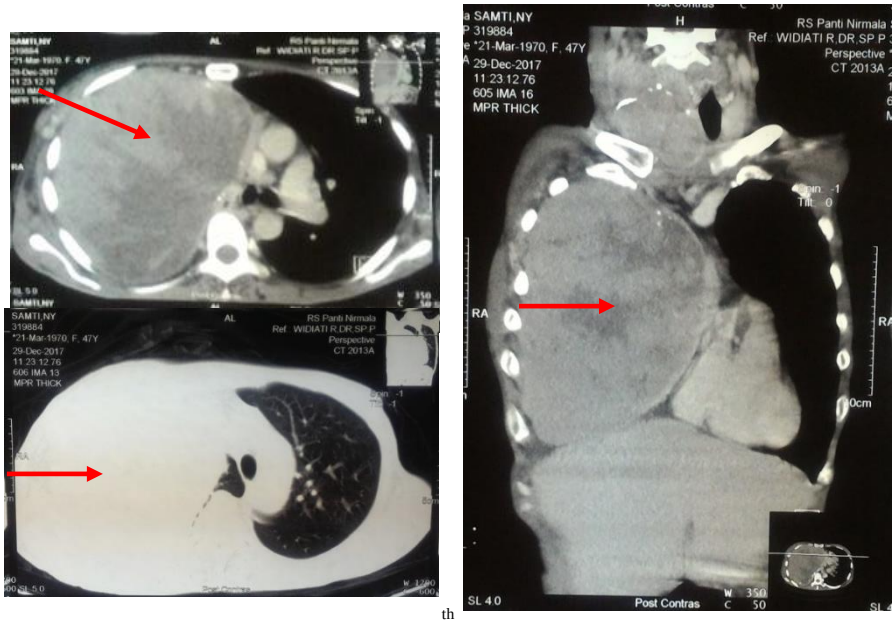

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Figure 2. Ct Scan Thorax mediastinal mass Dextra : 20x20x9 cm.

Bronchoscopy

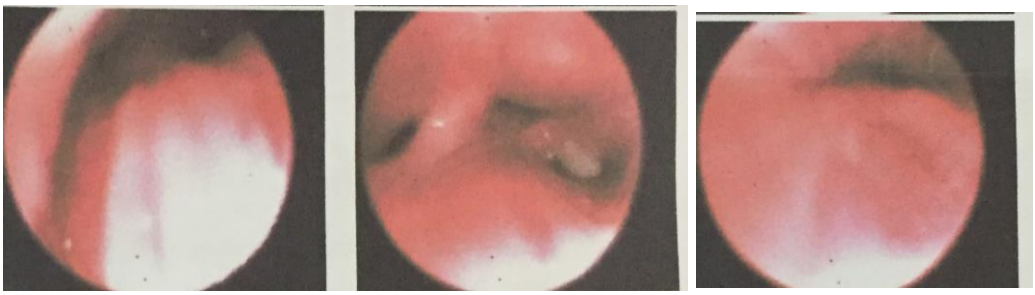

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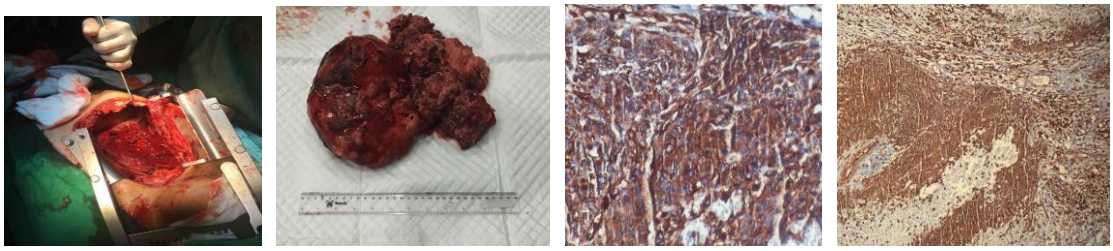
Figure 2. Partially stenoting compression in trachea, right main bronchus and intermedius trunch.

Surgery Process & Pathology Anatomy



A B C D

Figure 3. Surgery Process & Pathology Anatomy; (A) sternotomy: tumor mediastinum, (B) Tumor tissue (total tymectomy), (C) Vimentin (spindle cell) 450X enlargement is diffuse positive, (D) Vimentin 100X enlargement is diffuse positive

3. Discussion

In this case, it was reported that a 47-year-old woman came with complaints of chest pain, shortness of breath, cough, fever, anorexia and weight loss. This is in accordance with the literature which states that the incidence of thymoma increases with increasing age with a mean age of 40-50 years with almost the same predilection for men and women (1: 1,2). Symptoms that can be found in thymoma are cough (60%), chest pain (30%), fever (20%) and shortness of breath (16%). As with other high-grade carcinoma of the mediastinum, in sarcomatoid carcinoma the most common symptoms include chest pain, cough and shortness of breath. Fever, anorexia, weight loss and other systemic symptoms are nonspecific features of malignancy.[4,5]

In this patient, a chest X-ray was examined, and the chest X-ray showed a right lung mass. Then a chest ultrasound examination was performed. The chest ultrasound showed a solid mass in the right pulmonary area. On chest CT scan with contrast, there was a large mass tumor with firm and flat borders covering the entire right hemithorax, calcification spots on the mass tumor, benign impression of the tumor on the right hemithorax DD Adenoma. In the literature, it is stated that on chest CT scans, the presence of a thymoma is usually a well-defined round or oval mass in the thymus without enlarged lymph nodes, in radiologic thymic carcinoma a heterogeneous density mass due to necrosis or bleeding with irregular borders, often local invasion and metastases. far (55-65%), in sarcomatoid carcinoma the imaging image is the same as that found in squamous cell carcinoma, the tumor tends to be large and lobulated, with a low attenuation area on the chest CT scan associated with tissue necrosis, on X-ray intrathoracic gioter. Chest-ray may show superior mediastinal radiopacity causing tracheal deviation to a place opposite to the

superior border of radio-opacity / undetermined mass (cervicochothoracic sign).[9]

The closest diagnosis is thymoma, laboratory tests that support the diagnosis of thymoma are blood AFP and β HCG levels that do not increase. The literature states that Alpha-fetoprotein (AFP) levels and beta-human chorionic gonadotropin (beta-hCG) levels can be measured to exclude germ cell tumors, thymus epithelial tumors are likely to occur if these are found: 1) mediastinal mass defined by both in the area of the thymus which is not adjacent to the thyroid gland; 2) The tumor marker for AFP or beta-hCG is negative; and 3) no other adenopathy.[10]

The result of the histopathological examination after surgery was Spindle Cell Thymic Carcinoma. In the literature, it is stated that the WHO histological classification system, thymoma is classified into 6 subtypes histologically (type A, AB, B1, B2, B3 and C) based on neoplastic epithelial cells and together with the lymphocyte-epithelial cell ratio. Type A consists of spindle cells, type AB is a mixture of spindle cells and lymphocytes, type B1 if there are more lymphocytes than epithelial cells, type B2 is a mixture of lymphocytes and epithelial cells, type B3 if predominant epithelial cells and type C is a thymic carcinoma. Furthermore, an immunohistochemical examination was carried out, namely an examination of the immunohistochemical streak of CK and Vimentin, and the conclusion was that the immunophenotype was more in accordance with Sarcomatoid Carsinoma. In the literature, it is stated that sarcomatoid carcinoma is a type of thymic carcinoma which contains partial or complete spindle cells. These tumors contain both malignant epithelium (carcinomatous) and spindle cells (sarcomatosa / sarcomatoid), generally with a transition between the two, experienced by 5-10% of all thymic carcinoma sufferers. This disease is generally experienced in late adulthood, age four to eight decades, with a mean patient age of 47 years. Sarcomatoid carcinoma is a type of tumor that has high aggressiveness, most patients will die after 3 years of diagnosis despite aggressive multi-therapy. [10]

4. Conclusion

Woman, 47 years old with complaints of chest pain, shortness of breath, cough, weight loss, right neck & chest lumps enlarged since 1 month. Physical and supporting examination showed anterior superior mediastinal tumors and soft mass tissue in the right region of the colli. Based on the Trans Thoracal Guiding FNAB USG and FNAB the anterior colli region

concluded that Thymoma, thyroid adenoma and oncocytic nodular colloid goiter with azkanasy cell proliferation. Patients was performed Partial Sternotomy (Hemiclamshell), thymectomy with final diagnosis of Sarcomatoid Carsinoma. Furthermore, this patient is planned to be given chemotherapy with the ADOC combination regimen.

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