Germ cell tumor impersonating a pleuropericardial cyst: a rare phenomenon

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Abstract

A Mixed Germ Cell Tumor (MGCT) is a rare tumor with a low degree of differentiation. The most common sites are the yolk sac tand astrocytoma. Usually, it is gonadal, but in 5% of cases, it can present as extragonadal. The most common extragonadal site is the mediastinum, where the mediastinal mass sometimes mimics thymoma, lymphoma, and pericardial cyst and also occasionally causes pericardial tamponade. The presentations vary, ranging from accidental findings on routine radiography to life-threatening respiratory and cardiovascular compromission. We present an extragonadal MGCT case presenting as a mediastinal mass with symptoms of 1-year duration. The diagnosis was confirmed on the Histopathological Examination (HPE) of the surgically excised specimen. Given a possible life-threatening condition, a timely diagnosis is required.

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Introduction

A Mixed Germ Cell Tumor (MGCT) is a rare tumor form of two or more germ cell components, with a low degree of differentiation.[1] Usually, MGCT occurs in ovary and testis, and is rarely found at extragonadal sites like head and neck, mediastinum, and abdomen. Germ cell tumors usually originate from the gonads and are derived from reproductive cells embryologically. But in approximately 4-5% of all the cases, they have extragonadal origin, with the most common extragonadal site being the mediastinum. Primary mediastinal non-seminomatous germ cell tumors are very rare, approximately 4-5% of all germ cell tumors. The presentations vary, ranging from accidental findings on routine radiography to life-threatening respiratory and cardiovascular compromission.[2] Pericardial cysts are rare, with an incidence rate of 1 in 100000. Usually, they are congenital but, in some cases, can present as acquired pericardial anomalies (e.g., post-inflammatory, hydatid, neoplastic). Pericardial cysts are induced by an incomplete coalescence of fetal lacunae during the development of the pericardium.[3] Pericardial cysts are usually unilocular, well-marginated spherical, or teardrop-shaped and may be attached to the pericardium directly or by a pedicle. Of all pericardial cysts, 70 to 75% are located at the right cardio-phrenic angle, and the rest are on the left side of the mediastinum.[4] The mediastinal mass sometimes mimics thymoma, lymphoma, and pericardial cyst and also occasionally causes pericardial tamponade, which needs proper investigation and timely treatment. Herein, we report a case of mediastinal MGCT. We also reviewed the relevant literature, focusing on the Computed Tomography (CT) and clinical characteristics of mediastinal MGCT.

Case Report

A 42-year-old male came to our Outpatient Department (OPD) in June 2022 with complaints of chronic dry cough, progressive shortness of breath on exertion, and weight loss for the past 1 year.

The patient had visited multiple medical setups for these complaints, and received different treatments, including Anti-Tuberculosis Treatment (ATT), which he stopped on his own after

further decline in his health. Eventually, he came to our OPD. On physical examination, the patient was found to be in good clinical condition, conscious, and oriented to time, place, and person, with Blood Pressure (BP) 120/80 mmHg, Heart Rate (HR) 82/min & SPO_2 97% on room air.

On respiratory examination, diminished breath sounds on the left lower lung area were found on auscultation. Chest X-ray (Figure 1) showed high density, oval-shaped opacity, situated in the right para-cardiac area with anterior projection. Ultrasound of the thorax revealed a pleuro-pericardial cyst located tangentially to the right chest wall.

Contrast Enhanced Computed Tomography (CECT) of the thorax (Figure 2) described a well-defined, non-enhancing hypodense (25-30 HU) lesion (55*55*95mm) in the anterior mediastinum right paracardiac region, at the level of the right atrium, abutting the pericardium with mild indentation. It was also causing smooth indentation on the underlying lung parenchyma. The lesion shows a peripheral intermittent thick calcified rim with non-enhancing hyper-dense content in the inferior portion. Multiple nodules of soft tissue attenuating clustered lesions are seen in the vicinity of the aforementioned lesion in the right paracardiac fat, measuring 51*49*32 mm. Laboratory reports were normal. Two Sputum smears for Acid Fast Bacilli (AFB) were negative. IgG antibodies specific for Echinococcus were negative. A provisional diagnosis was made as a right pleuropericardial cyst, and the patient was advised for surgical intervention.

The patient underwent surgical intervention, and the right para-mediastinal mass was taken out (Figure 3). On Histopathological Examination (HPE), it was diagnosed as an MGCT consisting of seminoma, embryonal carcinoma, and mature teratoma (Figure 4, Figure 5). Hematoxylin and eosin-stained sections showed features of seminoma and embryonal cell carcinoma, respectively. Pus culture taken from para-mediastinal mass shows no growth. Post-procedural chest X-ray and CECT Chest showed no visible space occupying the mass lesion.

Post Procedural chest X-ray (Figure 6), CT chest (Figure 7), and Positron Emission Tomography-Computed Tomography (PET-CT) scan (Figure 8) were done. PET-CT was done mainly to rule out any metastatic possibilities, which showed mildly Fluorodeoxyglucose (FDG) avid soft tissue thickening in the anterior mediastinal region with non-FDG avid nodular densities in upper lobes of both lungs and in the left lung lingula with mild right-sided pleural effusion. The immunoassay done for Alpha Fetoprotein (AFP) (3.65 ng/ml) and Beta Human Chorionic Gonadotropin (HCG) (7.02 IU/L) was found within the normal range.

The patient was advised a reexamination after 6 months, and regular follow up of the case was done till that time.

Discussion

Germ cell tumors develop embryologically from the reproductive cells. They primarily arise from the gonadal organs. Since only 5% of cases of germ cell tumors are extragonadal in origin, it is uncommon to identify these tumors.[5] The most common extragonadal sites include the mediastinum, retro-peritoneum, vagina, and brain.[6] Clinical characteristics range from life-threatening respiratory and cardiovascular impairment, to unintentional discoveries on routine radiography. Such massive tumors can cause symptoms like coughing, shortness of breath, and chest pain, or they can perforate and cause other symptoms, including pleural and pericardial effusion.[7,8] Mediastinal tumors are of different types, *e.g.,* teratoma, seminoma, embryonal carcinoma, choriocarcinoma, and

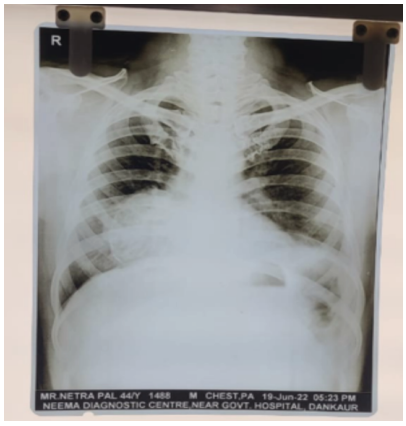

Figure 1. Pre-procedural chest X-ray.

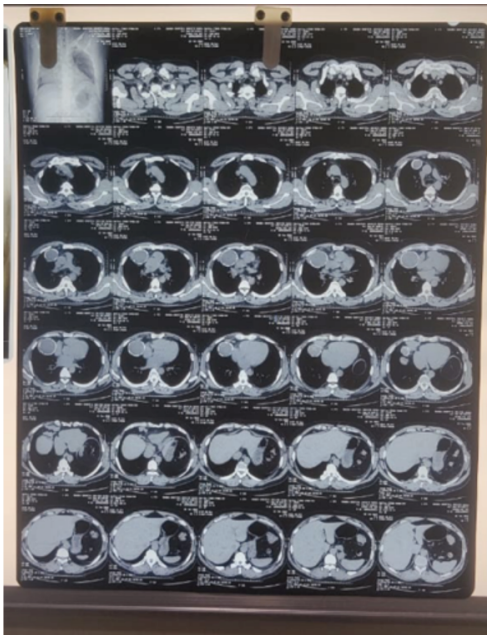

Figure 2. Pre-procedural Computed Tomography (CT) of the chest.

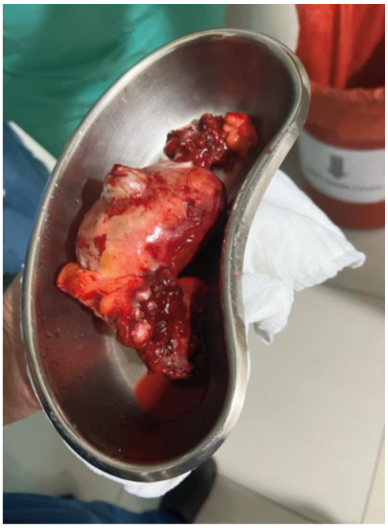

Figure 3. Post-procedural specimen.

MGCT. Teratomas are classified into: i) mature teratoma; ii) immature teratoma – benign; iii) immature teratoma - with a malignant component.

Immature teratoma with malignant component is further classified into: i) Type I, with another Giant Cell Tumor (GCT) [seminoma, Yolk Sac Tumor (YST), Embryonal Carcinoma (EC), choriocarcinoma]; ii) Type II, with a malignant epithelial neoplasm (adenocarcinoma, squamous cell carcinoma, *etc*.); iii) Type III, with a malignant mesenchymal component (rhabdomyosarcoma, angiosarcoma, *etc*.); iv) Type IV, any combination of previous types.[9]

Anterior mediastinal tumors account for 50% of all the mediastinal masses, including thymoma, teratoma, thyroid tumors, and lymphomas.[10]

Thymoma is the most commonly occurring anterior mediastinal mass in the adult population, although it accounts for less than 1% of adult malignancies.[11] Thymomas mainly afflict people over the age of 40; they are uncommon in youngsters and equally affect both males and women. Between 20% and 30% of thymoma patients experience symptoms as a result of pressure.[12]

The anterior mediastinum is where primary mediastinal lymphoma typically develops. Nearly 20% of all mediastinum neoplasms in adults and 50% in children are malignant lymphomas. The most frequent cause of masses in the pediatric mediastinum is lymphomas.[13]

Hodgkin Disease (HD) has a bimodal distribution of incidence, peaking in young adulthood and again after the age of 50 years.[14] T-cell lymphoblastic lymphoma is a form of mediastinal Non-Hodgkin Disease (NHD) that mainly occurs in children and adolescents.[15] A differential diagnosis for mediastinal masses is peri-

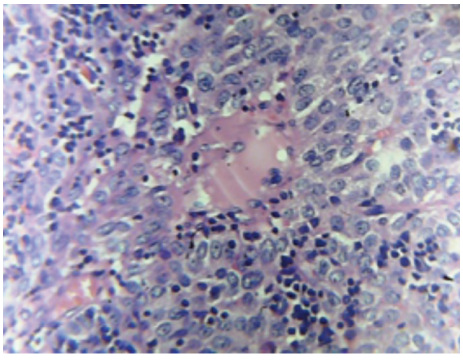

Figure 4. Histopathological Examination (HPE) showing seminoma.

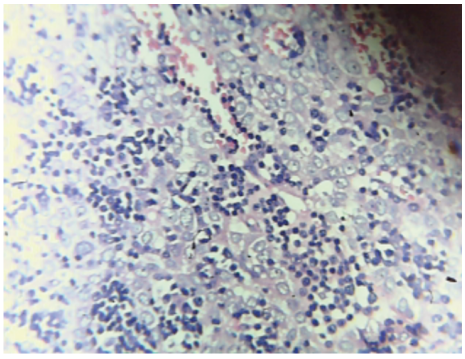

Figure 5. Histopathological Examination (HPE) showing embryonal cell carcinoma.

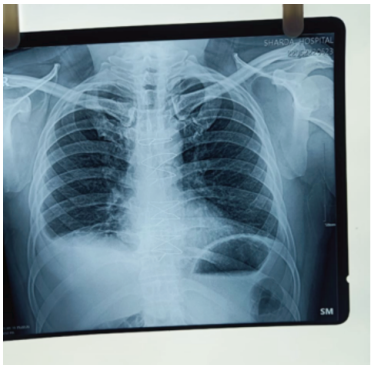

Figure 6. Post-procedural chest X-ray.

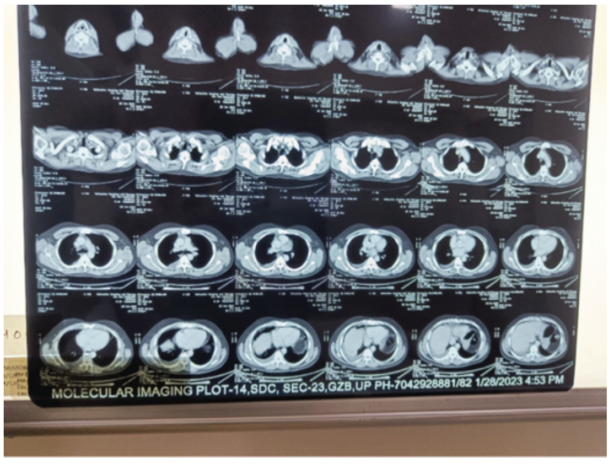

Figure 7. Post-procedural Computed Tomography (CT) of the chest.

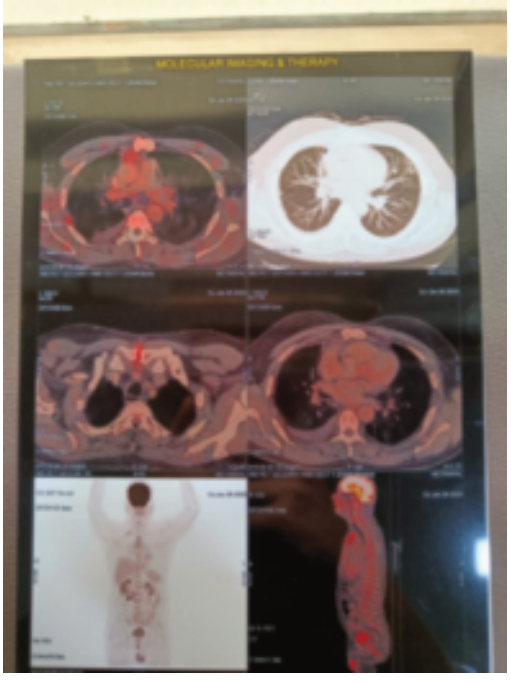

Figure 8. Post-procedural Positron Emission Tomography-Computed Tomography (PET-CT).

cardial cysts, which are often unilocular, well-marginated, spherical, or teardrop-shaped lesions that may be linked to the pericardium directly or by a pedicle. At the time of diagnosis, 85% of patients with mediastinal Non Seminomatous Germ Cell Tumors (NSGCTs) are symptomatic. The most frequent presenting symptoms are chest discomfort, hemoptysis, cough, fever, or weight loss. There may occasionally be pericardial tamponade or superior vena cava syndrome.[4,16] Anterior mediastinal NSGCT presenting as early cardiac tamponade is rarely seen in clinical practice.[17]

At the time of presentation, cough, chest discomfort, fever/chills, and dyspnea are common symptoms. Systemic symptoms are often brought on by the production of too many hormones, antibodies, or cytokines, whereas localizing symptoms are secondary to tumor invasion (respiratory compromise, paralysis of the limbs, diaphragm, and vocal cords, Horner syndrome, superior vena cava syndrome).[18]

In our case, the presenting complaints of chronic dry cough, progressive shortness of breath on exertion, and weight loss for the past year were either due to tumor-induced compression or the total atelectasis of the lung following recurrent pulmonary infections. Benign mediastinal teratomas may be secondarily infected following rupture into nearby structures. Chest X-ray and CECT scan are the preferred investigations for screening and diagnosis.

But, in this situation, a preoperative biopsy would have been useful, although it was not recommended due to the location of the tumor. The acknowledged standard practice for confirming a histological diagnosis is fine needle core biopsy.[19] In germ cell tumors, tumor markers such as beta-HCG, AFP, and Lactate Dehydrogenase (LDH) are typically increased. When evaluating the effectiveness of chemotherapy, especially in chemo-sensitive GCT, tumor marker testing is essential. Raised serum AFP levels suggest that mixed germ cell tumors contain yolk sacs and embryonal components.[20]

The preferred surgical intervention is a thoracotomy, which is done from the side of the tumor where it has more extension. The median sternotomy can also be used when the tumor is tiny, and there are few adhesions to nearby important organs.[21] Surgical excision was done, and on HPE, a diagnosis of mixed germ cell tumor having components of seminoma, embryonal cell carcinoma, and teratoma was made.

Conclusions

Although benign mediastinal teratomas are rare, they may present as a chest wall mass-producing bulging from the mediastinum towards the chest wall. This slow-growing tumor is usually asymptomatic and is often detected incidentally on chest X-ray. Echocardiography and chest CT are helpful in evaluating the compression of adjacent structures. In cases of cystic masses containing fat and calcific densities presenting in unusual locations, the diagnosis of teratoma should be considered. The treatment is surgical excision, along with proper investigations to be done to make the final diagnosis. Though they cannot be removed totally, their recurrence rate is very low.

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