

Oncologists Congress 2019: Giant Mediastinal Mixed Germ cell tumor, a rare case report and review of literature - Abdulrahman Hakami - Assistant Professor, Department of Oncology, Jazan University, Saudi Arabia.

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Introduction:

Germ cell tumors are relatively rare, embryologically derived from reproductive cells usually arise in the gonads. Mediastinal germ cell tumor estimated about 1-3 % of all germ cell tumors, generally seen in the anterior mediastinum and the metastatic lesions are mostly seen in the posterior mediastinum. The most antagonistic germ cell tumor subtypes are choriocarcinoma, embryonal carcinoma and yolk-sac tumors. While seminomas only very rarely spread distantly. The presentations vary ranging from accidental findings on routine radiography to life-threatening respiratory and cardiovascular compromise, can also present as gigantic big intrathoracic germ cell tumor like our case. According to a surgeon's point of view, the nature of mass suggests the possible surgical difficulties with regard to the approach and accessibility. A huge intrathoracic mass may compress the contralateral lung during positioning which may obstruct the venous return to the heart and thus poses a challenge to the attending anaesthetist.

We present a case of gigantic intrathoracic germ cell tumour which was resected successfully via a piecemeal surgical approach. The anatomical basis of this huge tumour and the treatment modalities are discussed.

Case report:

30 years old male patient, not known to have any chronic illness, referred from TB hospital center because history of dyspnea, cough and loss of appetite with weight loss for more than 4 months, no history of chest pain or hemoptysis. Chest x-ray done and showed complete obliteration of the right side of thorax, was suspected pleural effusion and diagnosed as case of pleural TB and empyema, started on ant tuberculosis drugs, antibiotics and received chest drain with a little bloody fluid. Patient not improved and referred to our hospital, Computed hospital of chest with contrast revealed a very big mas obliterating the right side of chest, pushing the trachea and mediastinum to the left side with minimal effusion in both sides. Pleural US revealed mass and effusion but no empyema.

Differential diagnosis was mediastina mass, adenocarcinoma, thymic carcinoma, lymphomas, fibroma or fibrosarcoma. US guided transthoracic fine needle biopsy from the right side mass revealed mixed germ cell tumor. The patient's condition had rapidly deteriorated prior the confirming the diagnosis or starting with treatments and died because of difficult airway breathing due to deviated and compressed airway and possible pneumothorax after transthoracic biopsy.

Discussion

Germ cell tumours are embryologically derived from reproductive cells. In majority, they are originated from gonadal organs. It is unusual to find germ cell tumours which are extragonadal in origin, whereby it accounts for 5% of the cases. The most common extragonadal sites include mediastinum, retroperitoneum, vagina, and brain. They have been also reported at sites such as lung, liver, prostate, and omentum. Conventional anatomy textbooks do not highlight the abnormal sites of germ cell tumours, hence giving the case reports as the only source of information. Researchers suggested that there is abnormal cell migration during embryogenesis or profuse distribution of germ cells to organs such as liver, thymus, bone marrow, and brain. These cells act with different regulatory function at sites mentioned above or transmit valuable genetic, hematologic, or immunologic information.

The clinical features vary differently from accidental findings on routine radiography to life-threatening respiratory and cardiovascular compromise. Symptoms arising from such huge tumours are due to compressive effect on the surrounding organs which include cough, shortness of breath, failure symptoms, and chest pain or due to tumour rupture such as pleural effusion and pericardial effusion. The largest ever mixed germ cell tumour of the mediastinum reported was 21 × 20 × 16 cm in size and weighed 3 kg, thus giving this present case a bizarre literature especially originating from South-East Asia.

Preoperative biopsy is indicated in this case in order to guide our treatment strategy. The role of clinical assessment is highly limited. Hence, radiological finding to determine the tumour origin and location in the mediastinum is crucial. Fine needle core biopsy is accepted as the standard procedure for confirmatory histological diagnosis. It should be performed under radiologic guidance by well-trained personals due to extensive vital structures surrounding it. It is essential as lymphoma was one of the differential diagnoses, guided by an elevated LDH level biochemically. Standard treatment for primary mediastinal lymphoma is 6 cycles of chemotherapy alone, namely, R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) protocol, and hence by diagnosing it avoids unnecessary mutilating surgery.

Tumour markers are frequently elevated in germ cell tumours, namely, LDH, AFP, and beta-HCG. Tumour marker measurement is mandatory in assessing the response to chemotherapy especially in chemosensitive GCT. Raised serum AFP levels indicate the presence of yolk sac and embryonal elements in mixed germ cell tumours, as seen in our case. This makes a GCT more likely to be the provisional diagnosis. A rapid decline of tumour marker levels after platin-based chemotherapy is associated with improved overall survival.

Almost 70% of the nonseminomatous germ cell tumours contain more than two germ cell components, so they are termed as mixed germ cell tumours. The main components of the mixed type germ cell tumour are yolk sac tumour and teratoma. In this reported case, the histopathological examination revealed nonseminomatous mixed germ cell tumour with three

germ cell components, namely, teratoma, embryonal carcinoma, and yolk sac. The teratoma component is composed of islands of mature stratified squamous epithelium, keratin cyst formation, mature cartilages, clusters of columnar epithelium with goblet cells, and cystic areas lined by mature ciliated respiratory epithelium. Immunohistochemical studies highlighted the presence of embryonal and yolk sac tumour components as evidenced by positive CD30 and AFP, respectively. However, beta-HCG for choriocarcinoma and PLAP for seminoma were negative.

In the era of minimally invasive surgery, small mediastinal tumours have been surgically removed via laparoscopic procedure, but large masses are best managed using median sternotomy approach. A clamshell incision provides the best exposure for surgical handling in comparison to median sternotomy when it comes to extremely huge tumours. It is not without its shortcomings as it only provides bilateral exposure and does not provide vertical dimension handling. Thus, using a lateral sternal split with anterior thoracotomy at the level of the third intercostal space may provide a better alternative.

Conclusion:

Germ cell tumors are aggressive and rapidly growing cancers, the previous literature reported the nature of the extragonadal mediastinal germ cell tumor can appear as Giant mass occlude whole lung, compressing the great vessels, adherent to chest wall, pericardium, and lung, like our case and this make a worse prognosis, The estimated event-free survival at 10 years after combined treatment is 80.4%. Chemotherapy, debulking and pneumoctomy are the treatment for such cases.