A Rare Case of Mediastinal Yolk Sac Tumor

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Abstract: Yolk sac tumor is a germinal cell tumor that commonly causes testicular malignancy. The incidence of these tumors lies in 90-95% of all testicular malignancies and commonly affects children with a median age of 1.5 years. In mixed type in adults, these tumors present in the age group 25-30 years. We reported a case of a 63-year-old man, who suffered from shortness of breath, accompanied by weight loss. Chest x-ray showed a homogeneous consolidation in the mediastinum. With suspicion of a mediastinal tumor, the patient underwent a core biopsy of the mass in the mediastinum. On histopathology examination showed pleomorphic tumor cell form, enlarged nucleus, hyperchromatic, eosinophilic cytoplasm. The tumor cells appear to form Schiller-Duval bodies. This case was diagnosed as a yolk sac tumor and underwent chemotherapy.

1 INTRODUCTION

Germ cell tumor is a group of benign and malignant neoplasm originated from primitive germ cell during early embryogenesis. Germ cell tumor frequently occurred in the gonads, only 5-10 % happened in the extra gonad. Extragonadal germ cell tumor is mostly found in the anterosuperior mediastinal (Bokemeyer *et al.*, 2002). Primary yolk sac tumor in the anterior mediastinum is rare and has a vicious prognosis. Patients often present with advanced stage tumors that are bulky and unresectable. Like other germ cell tumors, yolk sac tumor is predominantly a disease of young adults. However, a few cases of gonadal and extragonadal germ cell tumors have been reported in elderly patients as well (Nakhla and Sundararajan, 2016). We present a very rare case of an elderly 63-year-old male with primary yolk sac tumor of the mediastinum.

2 CASE REPORT

A 63-year-old male was admitted with shortness of breath that he had been suffering for the past 3 months. Shortness of breath was not related to physical activity or weather changes. The patient also complained of cough that he had experienced in the previous 2 months without any sputum production. Systemic complaints experienced by patient included very rapid weight loss during the last 6 months and sub-febrile fever during the last 1 month. His vital signs were as follows: blood pressure 120/80 mmHg, pulse rate of 112 beats per minute, respiratory rate of 28 breaths per minute, and temperature of 37.4^{o}C. Based on the laboratory findings, Hemoglobin was 12.5 gr%, leukocytes were 9100 / mm^3, and platelets were 424,800/mm^3 respectively. Atrial blood gas results were pH 7.45, pCO_2 39.7 mmHg, pO_2 110.2 mmHg, HCO_3 27.9 mmol/L, BE 2,9 SaO_2 98.8%. Chest x-ray showed widening in the mediastinal segment. With suspicion of a mediastinal tumor, the patient underwent a core needle biopsy of the mass in the mediastinum guided by thoracic CT scan (Figure 1-3). Histopathology results showed pleomorphic tumor cell form, enlarged nucleus, hyperchromatic, eosinophilic cytoplasm (Figure 4). The tumor cells appear to form Schiller-Duval bodies. This case was diagnosed as a yolk sac tumor and the patient underwent chemotherapy.

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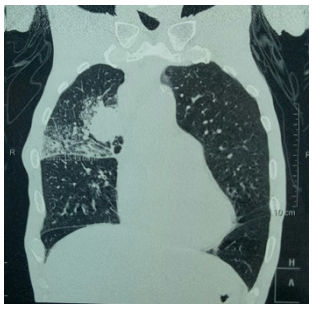

Figure 1: Coronal slice of Thorax CT showed the mass in the right upper lobe.

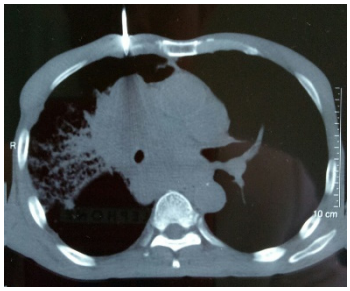

Figure 2: Thorax CT showed needle marker in the insertion phase.

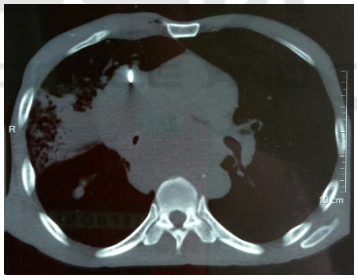

Figure 3: Thorax CT showed needle marker in the tumor site.

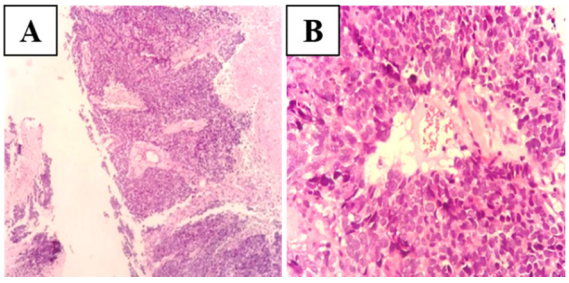

Figure 4: (A) Histopathology examination showed pleomorphic tumor cell form, enlarged nucleus, hyperchromatic, eosinophilic cytoplasm. (B) The tumor cells appear to form Schiller-Duval bodies.

3 DISCUSSIONS

Germ cell tumors mostly occur in the gonads. Extragonadal germ cell tumors are rare and most can arise in the pineal gland, retroperitoneum, and the mediastinum. The mediastinum is the most common site of extragonadal germ cell tumors. Malignant germ cell tumors in the mediastinum account for 1–6% of all mediastinal tumors. Primary extragonadal germ cell tumors, especially primary mediastinal tumors, are considered to have a poor prognosis. Germ cell tumors are histologically categorized into teratomas, teratocarcinomas, seminomas, and nonseminomatous carcinoma, including choriocarcinoma, embryonal carcinoma, yolk sac carcinoma, and mixed type carcinoma. Greater than 90% of malignant extragonadal tumors of the mediastinum occur in men (Bokemeyer *et al.*, 2002; Nakhla and Sundararajan, 2016). In a retrospective study by Sakurai et al. (2004) with 48 patients of extragonadal germ cell tumors, the median age at presentation was 28.8 years.

Yolk sac tumors can occur in both men and women, usually arising from germ cells in testes and ovaries, respectively. Pure yolk sac tumors are usually found in young children and mixed germ cell tumors with yolk sac are found in the adult. Similar to other nonseminomatous germ cell tumors, the latter can be associated with hematologic Klinefelter's syndrome (up to 20%) and other hematological malignancies such as acute leukemia and myelodysplastic syndrome. In an international study by Bokemeyer et al. (2001) with 381 mediastinal germ cell tumors, the most common symptoms on presentation were dyspnea (25%), chest pain (23%), cough (17%), fever (13%), night sweat, or weight loss (11%). Night sweat, fatigue, hemoptysis, and symptoms of superior vena cava compression were seen in <10% of patients with mediastinal germ cell tumors (Nakhla and Sundararajan, 2016).

Histologically, extragonadal germ cell tumors and mediastinal germ cell tumors have many similarities. Schiller-Duval bodies are pathognomonic and are helpful for identification. Yolk sac tumors immunohistochemical testing is positive for AFP, glypican-3, SALL4, and placental alkaline phosphatase (Bokemeyer *et al.*, 2002; Sakurai *et al.*, 2004).

The treatment regimens of extragonadal and gonadal yolk sac tumors are similar since they share histological patterns. Extragonadal nonseminomatous germ cell tumors have a considerably poorer prognosis. Chemotherapeutic schemes based on cisplatin have shown significant results with up to 50% of patients achieving long-

term survival (Sakurai *et al.*, 2004). Bleomycin-Etoposide-Cisplatin (BEP) therapy or etoposide (Vepesid), ifosfamide, and cisplatin (VIP), with at least 4 cycles of chemotherapy, are widely accepted regimens. VIP regimen may be preferred over BEP since patients with mediastinal germ cell tumors might need postchemotherapy thoracotomy for removal of residual tumor and bleomycin-induced pulmonary toxicities can be potentiated by surgery (Giannis *et al.*, 2009).

Surgical resection as the primary treatment modality is not recommended in mediastinal germ cell tumors because of the likelihood of early metastasis. However, there is a definite role for postchemotherapy adjuvant surgery to remove residual lesions and a rising serum tumor marker after completion of chemotherapy is not considered as a contraindication for surgery. Complete resolution of serum AFP marker occurs in less than 5% of patients. Survival rates have increased in patients who have the serum of AFP decreased after chemotherapy and in cases with residual tumor surgical excision (Vuky *et al.*, 2001).

Elderly patients with germ cell tumor generally have worse clinical outcomes compared to younger patients. A Surveillance, Epidemiology, and End Results (SEER) database analysis of 12,811 patients comparing the outcomes of testicular cancers in young adults (age <50) versus older adults (age >50 years) found survival from both localized and metastatic nonseminomatous germ cell tumors to be much better in younger patients compared to elderly (76.9% versus 57.0%) (Bokemeyer *et al.*, 2002; Sakurai *et al.*, 2004). Our patient had an overall poor prognosis due to multiple reasons such as his age, tumor location, and tumor bulk.

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