A HUGE MEDIASTINAL GERM CELL TUMOR

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ABSTRACT

A case of a huge mediastinal endodermal sinus tumor was shown in a 22-year-old male who presented with chronic cough and dyspnea. The mediastinal mass extends from right anterior mediastinum to the right posterior mediastinum. The tumor was a mixture of solid and cystic tissue. The airway was severely compressed. The sternum was destroyed and the anterior chest wall was invaded. The right pleural effusion was massive and multiple pleural nodules were found. The alpha feto-protein was markedly elevated and the beta HCG was zero. The patient passed away due to profound shock immediately after the procedure of intercostal drainage.

INTRODUCTION

Germ cell tumors encompass a group of tumors histologically identical to certain testicular and ovarian neoplasms, all of which are believed to be derived from primitive germ cell elements.[1] It includes benign and malignant teratoma, seminoma, endodermal sinus tumors, choriocarcinoma, and embryonal carcinoma. In the majority of cases, the neoplasm becomes manifest in adolescence or early adulthood, the mean age at diagnosis is around 30-31 years old;[2,3] the age incidence of benign and malignant tumors is similar.[4] For unexplained reasons, benign lesions are more common in females and malignant tumors in males. The vast majority of mediastinal germ cell neoplasms were located in the anterior compartment [4] (94%) and the rest were found in the posterior mediastinum. The most common form of mediastinal germ cell tumor is benign teratoma, particularly the cystic form[5] The most common malignant tumor is seminoma. Many malignant tumors have a mixed histologic appearance.

CASE REPORT

A 22-year-old male patient from Chaiyapum province, was admitted to Ramathibodi Hospital due to dyspnea for 10 days. He had chronic cough for 3 months and was worse for one month. The symptom was aggravated in the supine position. The mass was palpated at the upper chest without tenderness. Weight loss was detected for 5 kgs in 3 months. He had no known chronic illnesses. He smoked regularly for 6 years. At physical exami-nation, the patient had tachycardia and tachypnea, orthopnea, engorged external jugular vein. PMI was at 5th ICS, 4 FB lateral to the MCL. An ill defined, irregular surface mass was noted at the upper midline part of the chest wall, about 3 inches in the diameter. The superficial vein was detected on the mass with intact skin. Hypoalbuminemia was present.

AP chest film showed haziness of the whole right hemithorax with mediastinal shift to the left and depression of the right hemidiaphragm

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PMI = Point of Maximum Intensity

(Fig. 1). Thoracocentesis was performed yielding serosanguinous fluid about 1100 cc. Cytology of the fluid revealed cells that were similar to the adeno Ca. Serum alpha fetoprotein was elevated and measured 10,240 ng/ml. Beta - HCG level = 0. CT scan of the thorax (Fig.2) showed a large mixed density mediastinal mass which was located more at right side, extending from the anterior mediastinum to the posterior mediastinum. The mass contained no calcification and enhanced inhomogeneously. The invasion was seen to the sternum and midline anterior chest wall. Right pleural fluid was noted in massive amount with numerous pleural nodules. The thoracic trachea, carina and main bronchi of both sides were compressed and the lumens were obliterated. Later the bloody fluid was obtained from the ICD and the patient was hypotensive and finally passed away. The necropsy tissue was obtained. Alpha fetoprotein monoclonal antibody was positive; the placenta alkaline phosphatase was negative. Presence of yolk sac tumor or endodermal sinus tumor was noted in the specimen. The seen tumor cells were highly malignant.

DISCUSSION

Mediastinal germ cell tumors are generally considered to arise from cell rests whose journey along the urogenital ridge to the primitive gonad is interrupted in the mediastinum.[1,6-8] Occasionally, clinical or pathological examination of a testicle reveals either viable tumor or focal scarring consistent with regressed tumor [9] indicating that the mediastinal neoplasm represents a metastais. Although careful clinical examination must be performed in every case of mediastinal germ cell tumor to exclude this possibility, the relatively large number of negative pathologic examinations at autopsy[2] and the usual lack of emergence of a gonadal primary during prolonged clinical follow up indicate that this is in fact a rare event. From a diagnostic point of view, metastatic testicular cancer usually involves the retroperitoneal lymph nodes first and can bypass the mediastinum to affect the supraclavicular nodes; when the mediastinum is involved, it is seldom in the anterior compartment.

About 8 per cent showed the association between mediastinal germ cell neoplasms and Klinefelter's syndrome.[10-12] The reason for the increased risk of germ cell neoplasia in these patients is unclear but may be related to the abnormal androgen and gonadotropin secretion that is characteristic of the syndrome or to intrinsically abnormal germ cell tissue in patients with Klinefelter's syndrome (or to both).

Because mediastinal germ cell neoplasms have a varied histologic appearance, pathologic diagnosis can be difficult, especially when the amount of tissue submitted for examination is limited, such as may be obtained by mediastinoscopy or TTNA .[1,13] Although this diagnostic problem may not be very important in relation to specific typing of the tumors, particularly since many of the neoplasms are of mixed histology in any event, current advances in chemotherapy make the distinction from metastatic carcinoma of some importance. It seems reasonable to suggest, therefore, that the possibility of a primary germ cell neoplasm be considered in any young patient with an anterior mediastinal mass in whom a diagnosis of metastatic carcinoma is made in the absence of an obvious primary focus. In this situation, review of histologic material and testing for the presence of serum and tissue alphafetoprotein may alter the diagnosis; if the results of these investigations are equivocal.

Endodermal sinus tumors are highly malignant germ cell neoplasms believed to show differentiation toward yolk sac endoderm. The mediastinal form is rare.[14] The tumor also occurs occasionally in association with other germ cell neoplasms. Histologically, the neoplasm is quite variable in appearance, showing reticular, tubulopapillary, cystic and solid patterns. Perivascular structures resembling the rat endodermal sinus (shiller-Duval bodies) and small intra-and extracellular globules of PAS-positive material are

TTNA = Tran Thoracic Needle Aspiration

characteristic features. Immunohistochemical studies have shown a positive reaction in most cases for alphafetoprotein and alpha antitrypsin;[14] occasionally tumors also show CEA and keratin positivity. Extracellular basement membrane-like material is often present on ultrastructural examination.[1,15] Most endodermal sinus tumors present in young adults, the mean age was 22.6 years; [14] the majority occurs in male. Roentgenographic findings are nonspecific, consisting of an anterior mediastinal mass. Symptoms related to local mediastinal compression or invasion are present in most cases at the time of diagnosis; systemic symptoms (anorexia, weight loss, and fever) are often present as are those related to metastases.[14] Serum levels of alphafetoprotein are elevated in virtually all patients and can be a useful indicator of disease progression or remission with therapy. Although the prognosis is generally poor, prolonged survival and even cure have been documented in some patients treated aggressively.[16-18]

The presence of high serum beta-HCG would suggest seminoma, seminoma with syncytiotrophoblastic giant cells (STGC), embryonal carcinoma with STGC, yolk sac tumor with STGC, and choriocarcinoma. The presence of high serum alphafetoprotein would suggest embryonal carcinoma, embryonal carcinoma with STGC, yolk sac tumor with STGC, and choriocarcinoma. Presence of high serum levels of both alphafetoprotein and beta-HCG were seen in embryonal carcinoma with STGC, chorio Ca and yolk sac tumor with STGC.[19]

The most common malignant germ cell tumor is seminoma, which makes up 40% of the malignant subset. Teratoma with embryonal cell carcinoma (teratocarcinoma) is the next most subtype, with pure endodermal sinus tumor, choriocarcinoma, and embryonal carcinoma being much less common. Most patients with malignant germ cell tumors present with symptoms. Symptoms include chest pain, cough, dyspnea, weight loss, and fever. Primary mediastinal seminoma may be associated with low level of HCG, but significant elevation of AFP indicates that a nonseminomatous component of the tumor exists. In nonseminomatous germ cell tumors, 80% have an elevated AFP and 30% have an elevated HCG.[20]

CT shows seminomas as homogeneous bulky masses with small cystic areas, and nonseminomas contain larger cystic areas, calcifications, and fat plane invasion indicative of invasion. MRI may have a role in imaging these tumors with its ability to display multiplanar images, which may help in tumor surgery. Seminomas are sensitive to radiotherapy, and with this treatment alone, cure rates of 80% are obtained. Because of the success in treating gonadal seminomas, cisplatin-based chemotherapy is being used in patients with mediastinal seminoma. After treatment, there may be a small residual mass, which usually represents scar tissue and rarely harbors seminoma or teratoma if the mass is less than 3 cm. Nonseminomatous mediastinal tumors are treated with cisplatin-based chemotherapy followed by surgery to remove residual tumor. These patients have a worse prognosis than do the seminoma patients with only 50% survival rate.[20]

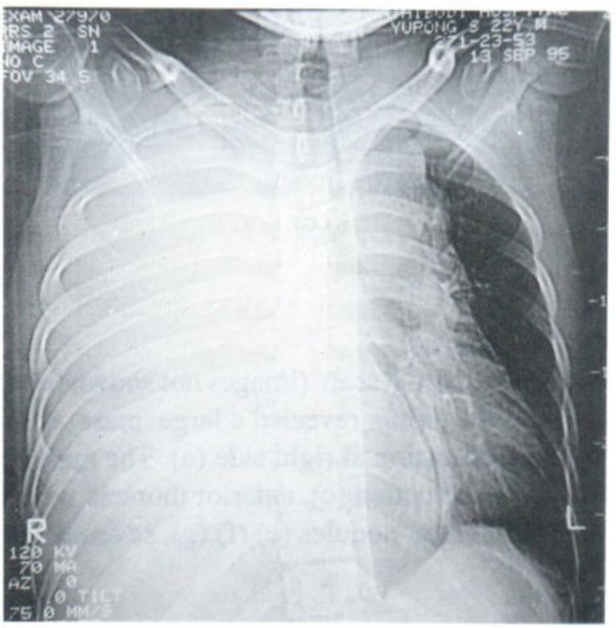
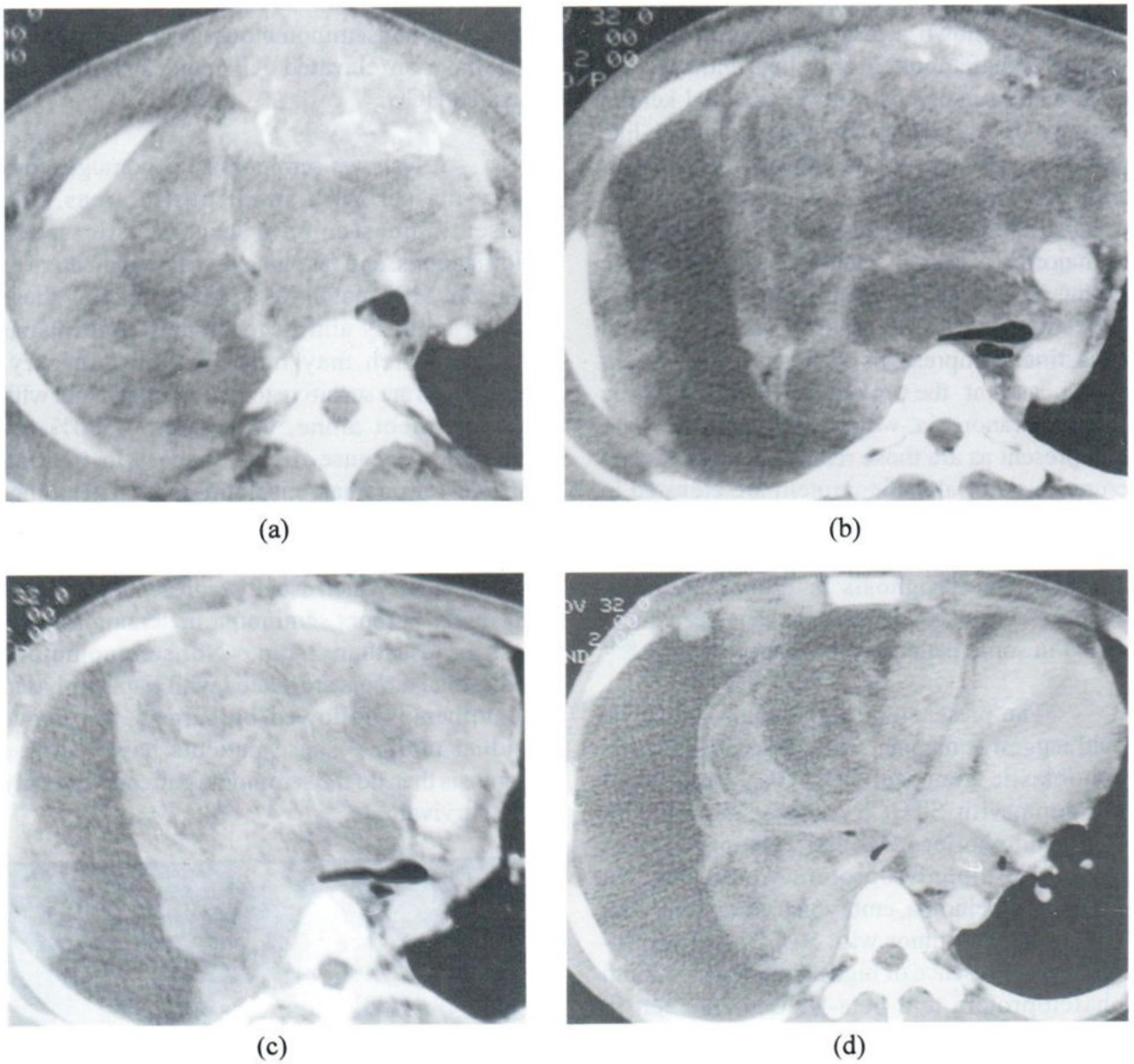


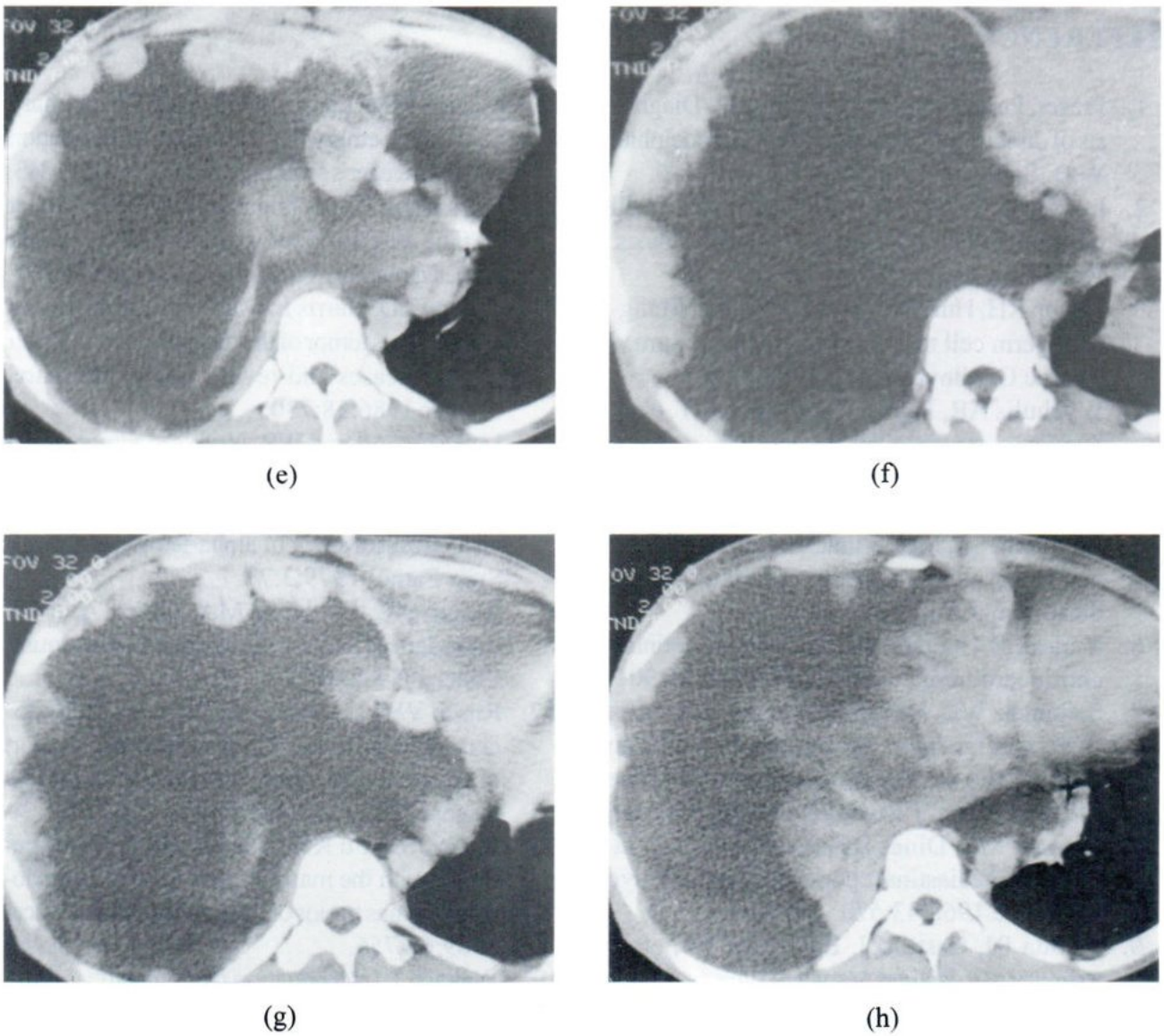
Fig. 1 Plain AP chest film shows total haziness of the right hemithorax with mediastinal shift to the left, inferiorly displaced right hemidiaphragm.



(a) (b)

(c) (d)

Fig. 2: a,b,c,d.
NCE CT scan (images not shown here) showed no calcification in the mass. CE CT scan of the thorax revealed a large mass at the anterior mediastinum with posterior mediastinal extension at right side (a). The mass showed predominantly cystic component (b). Sternal destruction (c), anterior thoracic wall invasion (d), massive right pleural fluid and right pleural nodules (e),(f),(g), severely air way compression were obviously observed (h).



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