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CASE REPORT

Primary Mediastinal Tumour – (Germ Cell Type) A Rare Case Report

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Abstract

The prevalence of mediastinal lesions found to range from 0.73% to 0.9% with mediastinal tumours occurring in <1% of the population.A19 year old male presented with back ache, chest discomfort and generalized weakness .On CECT Chest, diagnosis of mediastinal Lymphoma was made. Gross followed by routine microscopic examination was done and diagnosis of GCT was made supported by raised S.AFP level .We emphasise the need of high index of suspicion to diagnose Primary Mediastinal extragonadal GCT as they have poorer prognosis and survival outcome.

Key Words

Germ Cell Tumour, Alpha Foeto Protein, Yolk Sac Tumour

Introduction

The mediastinal lesions have a prevalence ranging from 0.73% to 0.9% with tumours in mediastinum being seen in only <1% of population[1] Germ cell tumors arise predominantly seen within testis but approximately 5 % to 10% of all germ cell cancers arise in extragonadal sites as in pineal gland, central nervous system, coccyx, cervix, vulva, pelvis, liver, prostatemediastinum and retroperitoneum**.** Malignant mediastinal germ cell tumors account for only 1% to 3% of all Primary Extragonadal germ cell tumor[2, 3]. Extragonadal germ cell tumourdevelop from primordial germ cell remanants.Majority of primary mediastinal germ cell tumours are seminomas with non seminomatous tumours being less commom. yolk sac tumor belongs to non seminomatous G CT category with grave prognosis. It is the most common germ cell tumour in infants and children (Prepubertal) whereas it is seen in only 2.4% of adult patients (post pubertal) in pure form[4].

We here highlight rare entity of Primary mediastinal tumor – (yolk sac type) diagnosed in our hospital.

.Case Report

A 19 year old male presented with back ache, chest discomfort and generalized weakness from past 20 days to a tertiary health care center where a routine blood examination was found to be normal.USG Chest showed a right sided moderate pleural effusion 300-350 cc in volume, left sided minimal pleural effusion, heterogenous mass 8 x 7.5 cm noted in left lobe, showing vascularity. CECT Chest revealed a large 90 x 150 x 210 mm well defined solid enhancing mass lesion with large areas of necrosis in Anterior Mediastinum in Midline and towards Left Side-likely a Lymph Nodal Mass - Lymphoma. Superiorly the Mass was extending up to the level of sternal notch, inferiorly it was extending up to infracardiac level. Medially it was abutting the pericardium and causing significant compression on Pulmonary Arteries (50% Narrowing noted in MPA, RPA & LPA due to compression. However, there was no true Invasion). Mild Pericardial effusion was noted. Left Sided Mild Pleural effusion with fissural extension and subsegmental lung collapse of underlying segments were seen (Fig. 1).

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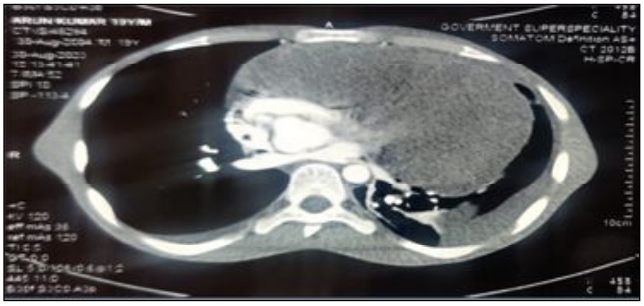

Fig. 1 : CECT Chest revealed a Large 90x150x210mm well defined solid enahncing mass lesion (arrow) with large areas of necrosis noted in anterior mediastinum in midline and towards left Side-likely a Lymph Nodal Mass s/o a Lymphoma. Superiorly the Mass was seen extending up to the Level of Sternal Notch.

Fig. 2 : Gross: Mediastinal mass measuring 35x12cm as multiple tissue pieces greyish white to grey brown tissue extremely friable.

During further workup, Computer Tomographic Aortic Angio scan revealed mass effect on aortic arch and its branches, SVC, Pulmonary Artery and Vein causing their mild luminal narrowing with posterior displacement and 90 – 120 degrees contact however no true invasion noted. On the basis of radiological findings patient was planned for thoracic surgery for resection of mediastinal mass, subsequently tumor was removed from SVC, thymic area, pericardium. Tumor tissue infiltrating into left area of lung was left as such.

Pathological Examination

Gross: Mediastinal mass measuring 35 x 12 cm (multiple tissue pieces greyish white to grey brown tissue extremely friable, areas of haemorrhage and necrosis) along with left upper pole Thymus, tissue from pericardium and lymph node was received in Pathology department. (Fig. 2)

Frozen Section: showed a tumor with reticular pattern as well as microcystic andmacrocystic areas lined by round to oval pleomorphic cells with moderate to abundant cytoplasm. Diagnnosis of Neoplastic pathology with the possibility of germ cell neoplasm was extended. Radiological diagnosis of lymphoma and clinical diagnosis of thymoma were ruled (Fig. a). **Microscopic Examination** of the sections revealed a tumor with abundant necrosis, hemorrhage and lymphoplasmacytic inflammatory cell infiltrate. The tumor cells showed

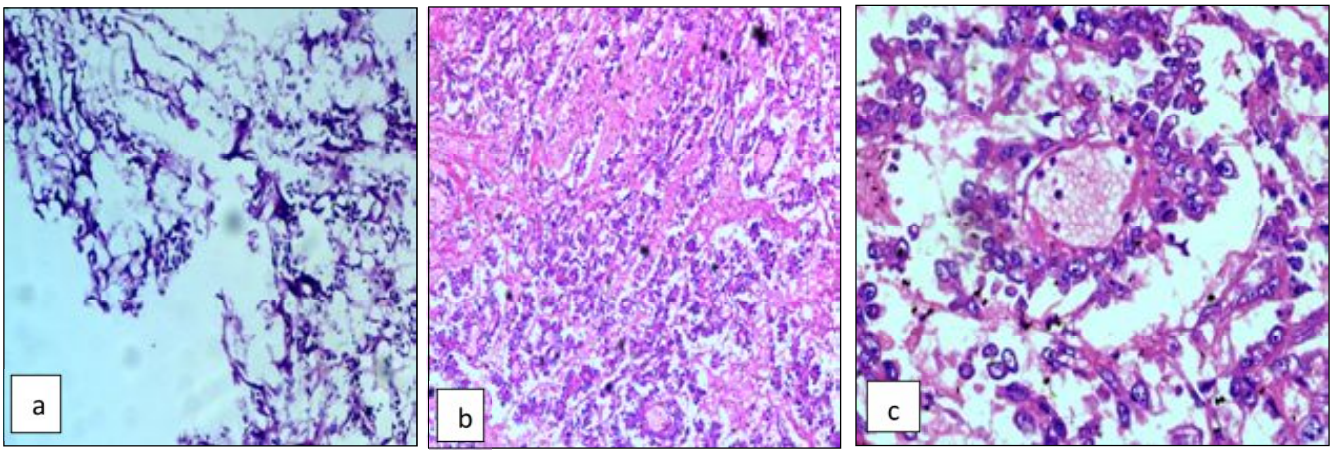

Fig. (a) Photomicrograph shows a tumor with predominantly having micro and macrocystic areas (arrow) lined by round to oval pleomorphic cells suggestive of malignant pathology. (Frozen, H&E 200X). Fig. (b) Photomicrograph shows a tumor with abundant necrosis, hemorrhage and lymphoplasmacytic inflammatory cell infiltrate. The tumor cells showed variable pattern such as reticular, microcystic, macrocystic lined by tumor cells have variable morphology cuboidal to columnar to polygonal in shape with mild to moderate eosinophilic cytoplasm .(H&E200x) Fig. (c) Photomicrograph shows a schillerduval bodies. (arrow) (H&E400x)

variable pattern such as reticular, microcystic, macrocystic lined by tumor cells having variable morphology with cuboidal to columnar to polygonal in shape with mild to moderate eosinophilic cytoplasm, vesicular nuclei with prominent 1 to 4 nucleoli at places with areas showing cells having hyperchromatic nuclei. (Fig b). Perivascular formation lined by layer of cuboidal/low columnar hyperchromatic epithelial like cells (Schiller Duvel Bodies). (Fig. c)

Patient was followed up in oncology ward, based on histopathology report further blood workup was carried out showing normal Beta HCG levels, raised Alpha Feto Protein (AFP) level >1000 ng/mL (reference levels 0-8) and LDH levels were 150 IU/L (refrence range 100-190). Usg testis was within normal limits. Based on characterstic histopathology, raised serum alphafoetoprotein levels and normal USG Testis diagnosis of primary Mediastinal tumor (Germ cell tumor) of mediastinum -Yolk sac type was confirmed.Patient was started on chemotherapy

Discussion

Lesions of the mediastinum are rare. The prevalence of mediastinal tumours foundin less than1% of population. The mediastinal masses span a broad histopathological spectrum with 50% occurring in anterior mediastinum.The most frequent etiologies of anterior mediastinal masses are thymic malignancies and lymphoma ifollowed by thyroid and other endocrine tumors, benign teratoma, malignant GCTs, and benign thymic lesions[5].

Among the germ cell tumours, benign teratomas are the most common .Seminomas and non seminomatous GCTs (NSGCTS) are rare malignant GCTs. Seminomas represent 25 to 50% of malignant GCTS exhibit normal alphafoetoproteinlevela (AFP). NSGCTs, comprising of yolk sac tumours, embryonal carcinoma, choriocarcinomas and mixed GCTs. commonly associated with raised alphafoetoprotein levels[6].

Germ cell tumors (GCTs) are classified as extragonadal if there is no evidence of a primary tumor in the testes. Primary mediastinal germ cell tumor in majority cases manifests as anterior mediastinal masses and in differential diagnosis, thymic diseases, thyroid goiter and lymphomas should be considered[7].

The treatment regimens of extragonadal and gonadal YSTs are similar since they share histological patterns. According to the International Germ Cell Cancer Collaborative Group (IGCCCG), PMYST is seen to be associated with a poor prognosis with a 40% to 50% overall Survival.[8] Surgical resection as the primary treatment modality is not recommended in mediastinal GCTs because of the likelihood of early metastasis. Chemotherapy combined with surgical resection, carried pre or post chemotherapy play a vital role in treatment. Complete resolution of serum AFP marker occurs in less than 5% of patients that affects the overall survival, asin patients who have decrease in serum of AFP after chemotherapy as well as residual tumor surgical excision survival rates increase.[4]

Conclusion

Mediastinal tumours are rare. Primary extragonadal mediastinal germ cell tumours are neoplasm wherein a high index of suspicion is needed to diagnose them as their prognosis is poorer and treatment protocol is different from that of the primary in testis/ovary.

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