

Primary mediastinal germ cell tumor - successful curative resection following chemotherapy - a case report

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Abstract

Primary mediastinal germ cell tumours are relatively rare and account for only a small portion of all the mediastinal tumours. We present a case of a primary mediastinal germ cell tumour in a 14 years old Afghani boy.

Introduction

Germ cell tumors account for approximately 1% of

all the malignancies occurring in men, with testis being the most common site of origin. Extra gonadal germ cell tumors can occur in the midline of the body from the pineal gland through to the mediastinum, retro peritoneum and sacrum. The different subtypes of this entity are made up of seminomas and nonseminomatous germ cell tumors (i.e., embryonal carcinoma, choriocarcinoma, yolk sac tumors, mixed germ cell tumors, mature and immature teratomas). Germ cell tumors are primarily treated with Cisplatin based

chemotherapy and the surgical resection is reserved for residual masses.

Case Report

A 14 years old Afghani patient presented with a 6-8 months history of dyspnea and left breast tenderness. He also gave one year history of weight loss, anorexia, malaise and non-productive cough. There was no history of bone pain.

On physical examination, bilateral gynaecomastia with left breast tenderness and multiple nodules were present. The breathing was harsh with decreased breath sounds on the left side and trachea was shifted to the right. Genital examination showed normal external meatus and skin, right testicular swelling which was mobile and non-tender and left testicle was almost atrophied. Heart sounds were normal.

Initial laboratory investigations revealed hemoglobin of 11 G/dl, hematocrit of 34, a white blood cell count of 8700/dL with 60.5% neutrophils and 26% lymphocytes, ESR of 40mm first hour creatinine of 0.7 mg/dl, LDH of 1796 IU, calcium of 8.5 mg/dl, β -HCG of 9.3 mIU/ml and alpha-fetoprotein of 9600 IU/ml.

LFTs showed bilirubin of 0.40 mg/dl, albumin of 3.33, SGPT of 14 and ALP of 333 (Table).

Table. Temporal assessment of tumour markers.

| Tumor Marker | Pre Chemotherapy | Post Chemotherapy | Normal values |
|---------------------|------------------|-------------------|---------------|
| AFP (IU/ml) | >30000 | 9.65 | 0.5-5.5 IU/ml |
| β hCG(mIU/ml) | 9.3 | <2 | 0-5mIU/ml |
| LDH (IU) | 1796 | 346 | 253-548IU |

Chest X-ray (Figure 1) showed a mass in the left chest abutting the heart and raised left hemidiaphragm secondary to phrenic nerve involvement.

CT scan of the chest (Figure 2) showed a mediastinal mass measuring 10.4x7.9x16 mm and occupying left hemithorax. The tumour was in the proximity to the heart and great vessels. The heart was pushed to the right side across midline with accentuated pericardial fluid trapped between the heart and described mass. It had well defined margins and left paratracheal lymphadenopathy was present.

After a month, Video Assisted Thoracoscopy (VATS) was done which revealed a gelatinous mass reaching upto the chest wall between 2nd and 5th Inter-costal spaces which was haemorrhagic. The biopsy of the mass showed a mixed germ cell tumor (Yolk sac tumour) with ter-

atomatous component. The patient was started on Chemotherapy consisting of Cisplatin, Etoposide and



Figure 1. Chest X-ray is being shown. Notice the mass in the left chest abutting the heart and raised left hemidiaphragm secondary to phrenic nerve involvement.



Figure 2. CAT scan of the tumor is being shown. Notice the proximity of the tumor to the heart and great vessels.

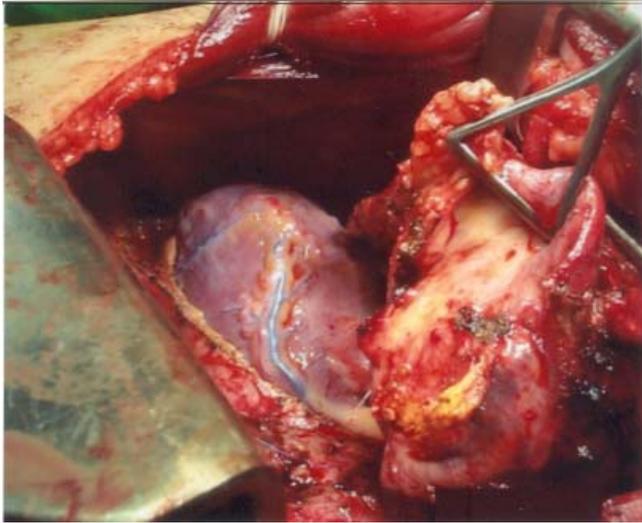


Figure 3. The resection of the actual tumor is being shown. Notice the anterior basilar segment of left lower lobe attached to the tumor which is also resected with the mass. The underlying heart is also seen.

Dexamethasone and received 8 cycles in total. After the shrinkage of tumor size, the residual mass in the mediastinum which was involving the phrenic nerve, pericardium, lung and portion of left ventricle, posterolateral thoracotomy was done and the entire tumor along with pericardium was removed (Figure 3). Post operative course was uneventful and the patient was discharged on 7th Post operative day.

Discussion

Mediastinal tumours are rarely observed in the clinical setting. Mean age of presentation is 35 years and no specific sex predilection has been observed. Clinical picture is vague and hence clinical diagnosis remains a dilemma. However, almost all tumours can be picked up on chest radiograph.¹ Mediastinum is documented to be the most common extra gonadal site of germ cell tumours while 95% of all the germ cell tumours are located in the gonads. They have been shown to originate from intratubular testicular germ cells and can either be seminomatous or non-seminomatous. Extragonadal germ cell tumours, which are a less common variant (5%), have been hypothesized to arise from migration of the germ cells along the urogenital ridge and most of them being the non-seminomatous variant.² However, primary mediastinal germ cell tumours remain a relatively rare clinical entity. Primary mediastinal germ cell tumors such as uchoriocarcinomas, teratomas with yolk sac component and embryonal carcinomas have been reported in the literature. Cases of co-existing primary gonadal and extra-gonadal germ cell tumours have also been cited.³ Workup of mediastinal mass consists of CT imaging of the chest, a biochemical assessment of tumour markers and fine needle aspiration or core biopsy. Testicular ultrasound

has also been practiced to rule out co existing germ cell tumours in the testis and the mediastinum.^{1,2}

Earlier literature had shown that FNA did not prove to be a good diagnostic tool for mediastinal germ cell tumours so management approach was not based on it.³ Based on the tumour markers and histologic assessment, primary germ cell tumour was suspected. Surgical resection of mediastinal germ cell tumours is recommended to follow chemotherapy after normalization of markers.² Sternotomy and posterolateral thoracotomy have been used before for anterior and anterosuperior mediastinal masses.² Mariel E Gels et al had successfully shown thoracotomy to be a successful postchemotherapy intervention in disseminated non seminomatous testicular germ cell tumours.⁶ Same approach has been used successfully for resection of intrathoracic primary germ cell tumour.³ In light of the cited literature VATS and Thoracotomy were performed after normalization of the tumour markers in the presentation. The tumor mass in the mediastinum which was involving the phrenic nerve, pericardium, lung and portion of left ventricle were completely resected via thoracotomy. The surgery was unremarkable and no major complications were observed.

Though multiple primary tumours remain a rather uncommon clinical observation, it is a potential risk in individuals with germ line mutations and epimutations in the DNA repair genes. Suter C M et al very recently studied two individuals with multiple primary tumours and elucidated an epimutation involving allele-specific and mosaic hypermethylation of the DNA mismatch repair gene MLH1.⁷ A similar genetic aberration in our patient may be responsible for the co existing primary tumours.

Hence, we support and recommend the workup comprising CT imaging of the chest, and temporal biochemical assessment of tumour markers for mediastinal germ cell tumours.

References

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