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PRIMARY MEDIASTINAL NONSEMINOMATOUS GERM CELL TUMOUR IN A YOUNG FEMALE

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Abstract : Primary mediastinal nonseminomatous germ cell tumours are uncommon tumours. Here we report a young female with primary mediastinal nonseminomatous germ cell tumour. This case is reported for the rarity of presentation of this curable tumour.

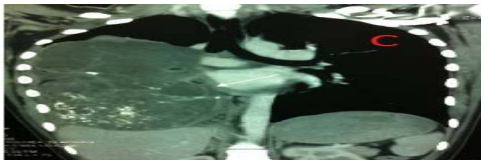
Keyword : Nonseminomatous, Germ cell, Mediastinum

INTRODUCTION :

Primary Mediastinal Nonseminomatous Germ Cell Tumour (PMNSGCT) is an uncommon neoplasm that arises nearly exclusively in males (1). They are more frequent in Klinefelter's syndrome and have the risk of development of haematological neoplasia that is not therapy related (2). Treatment comprises of cisplatin based chemotherapy followed by surgical resection of residual mass.

CASE REPORT :

Fifteen year old female presented with breathlessness of one month duration. On examination she had decreased breath sounds in the right hemi-thorax. Chest roentgenogram showed a homogenous opacity in the right middle and lower zones. Positron emission tomography (PET) showed a 16 * 11 * 10 cm multicystic mass with enhancing septa and calcifications in right paracardiac region and middle and lower zones of right hemi thorax with standardized uptake value (SUV) of 2.8. Uterus & bilateral adnexal structures were normal. Her serum tumour markers were alpha foetal protein (AFP) – 848 ng/ml; Beta human chorionic gonadotropin (HCG) – 2.3 mIU/ml; Lactate dehydrogenase (LDH) – 733 . Computerized tomography (CT) guided biopsy of the mass showed cystic structures lined by keratinized squamous epithelium, mucinous epithelium, ganglion cells and smooth muscle fibres suggestive of Teratoma. Bone marrow study was normal. She was diagnosed as Primary Mediastinal NSGCT. After 2 cycles of Bleomycin/Etoposide/Cisplatin (BEP) her AFP decreased to 17 ng/ml and
HCG and LDH were normal. However her CT showed a static response. She planned for reassessment for surgery after BEP # 4.



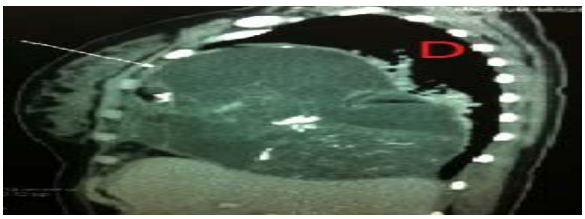
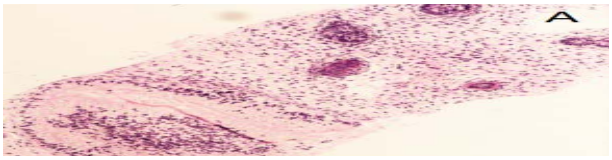
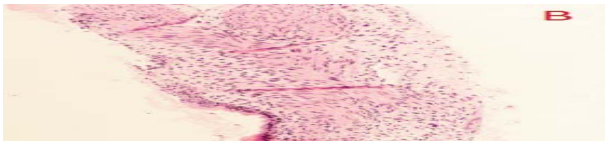
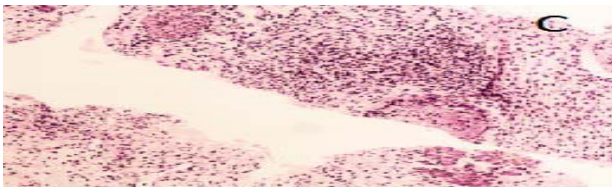


Figure 1. A.Upfront chest roentgenogram showing homogenous opacity in right mid and lower zones. B, C & D. Upfront positron emission tomography showing showing 16 * 11 * 10 cm multicystic mass with enhancing septa and calcifications in right paracardiac region and middle and lower zones of right hemi thorax.







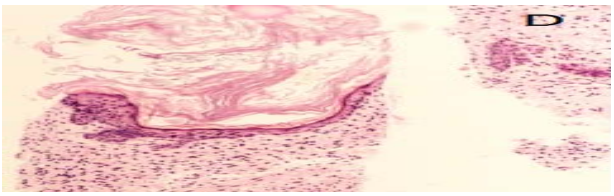


Figure 2. A. Eosin and Hematoxylin stain of mediastinal mass biopsy, 100x power showing ganglion cells. B. Smooth muscle fibres. C. Glandular structures. D. Keratinised squamous epithelium.

DISCUSSION :

Extragonadal germinal cell syndromes are rare tumours that comprise 5 – 10 % of all germ cell tumours. More than 90 % of tumours predominantly affect young males

3) and the predominant site is anterior mediastinum

(4)They are located in the midline regions including pineal gland, mediastinum, retroperitoneum or sacro-coccyx with no evidence of a primary tumour in the gondads by physical and radiologic examination. The proposed theories of pathogenesis include local transformation of primordial germ cells misplaced during embryogenesis and reverse migration of occult carcinoma in situ lesions in the gonad

(5). Mediastinal germ cell tumours comprise 50 – 70 % of all germ cell tumours in which mature teratoma constitutes 30 - 40 % and malignant germ cell tumours 60 - 70 %. Among the malignant mediastinal germ cell tumours seminoma constitute 40 % and non-seminomatous germ cell tumours constitute the remaining 60 %. The usual presentation includes dyspnoea, chest pain, cough and superior venacaval obstruction syndrome. The differential diagnosis includes lymphoma, leukemia, thymic tumour, thyroid tumour and neurogenic tumour. The workup includes tumour markers including AFP, HCG, LDH, contrast CT of the chest, bone marrow study with cytogenetics. Histological confirmation is mandatory if tumour markers are not elevated.

Stage I mediastinal germ tumour include tumours confined to mediastinum without microscopic invasion of adjacent structures. Stage II includes tumour confined to mediastinum with microscopic or macroscopic infiltration of adjacent structures. Stage III A includes tumour with intrathoracic metastasis and Stage III B includes tumour with extrathoracic metastasis. The most common associated haematological malignancy in PMNSGCT includes acute myeloid leukemia and myelodysplastic syndrome. The treatment of PMNSGCT is multimodality treated with four cycles of BEP chemotherapy (Bleomycin + Etoposide + Cisplatin) every 3 weeks followed by resection of residual mass

(6). Patients demonstrating complete necrosis have excellent long term survival (7). The presence of elevated serum tumour markers before resection does not affect outcome if the resection is complete (8). Recurrent or refractory tumours can be treated with high dose chemotherapy with etoposide plus carboplatin followed by autologous stem cell transplantation (9). The immediate toxicities includes nausea, vomiting, cytopenias and nephrotoxicity and the delayed toxicities include infertility, cisplatin induced neurotoxicity, bleomycin induced pulmonary toxicity and etoposide induced secondary leukemias.

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