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A RARE CASE OF PRIMARY MEDIASTINAL YOLK SAC TUMOUR

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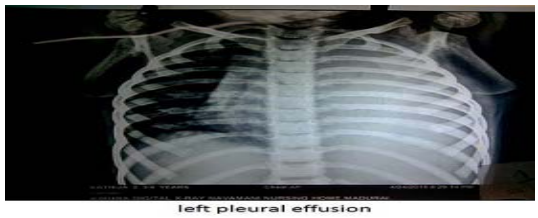
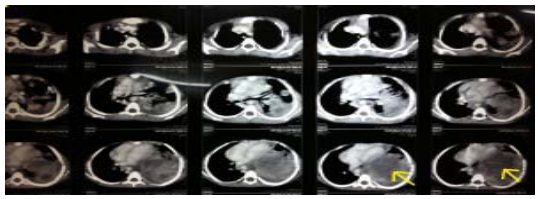
Abstract : Yolk sac tumour is a subtype of germ cell tumour and highly malignant neoplasm. It has been detected in several extragonadal sites in addition to usual presentation at ovaries and testes. Primary mediastinal yolk sac tumour is rare and carries grave prognosis. Here we present a case of 3 year old Female child with hemorrhagic pleural effusion followed by rapid evolution of intrathoracic mass and grossly elevated alpha fetoprotein (AFP) which later turned out to be mediastinal yolk sac tumour.

Keyword :primary yolk sac tumour, mediastinum, extra gonadal

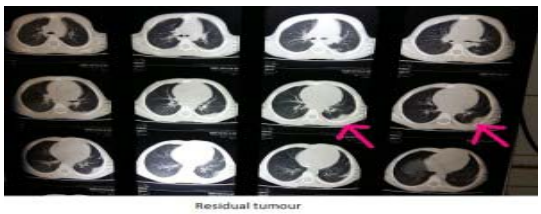
CASE REPORT:

3 year old girl, first born to non consanguineous parents, apparently normal before, developed high grade fever, cough, breathlessness for 2 days. On examination: child was dyspnoeic, pale, No lymphadenopathy, BCG scar present. Dull percussion note and diminished breath sounds elicited in left hemithorax. Mantoux test and IgM Elisa for HIV were negative. chest xray showed left sided pleural effusion. Pleural tap was hemorrhagic with no malignant cells; CT thorax revealed massive pleural effusion on left side with no evidence of mass. She improved after intercostal drainage, antibiotics and later discharged. Child returned with similar symptoms after 2 months. Chest xray showed mediastinal widening ; Diagnostic thoracocentesis revealed hemorrhagic effusion which aroused suspicion of malignancy. Repeat CT showed irregular hypodense mass (13 x 10 cm) with mediastinal shift suggestive of pulmonary tumour. Surgical excision of tumour followed by histopathological study showed malignant tumour composed of polygonal cells with ill defined borders, raised Nuclear: Cytoplasm ratio; and focal necrotic areas. Immunohistochemistry studies were positive for Alpha Fetoprotein, Neuron Specific Enolase, Pan Cytokeratin, CD 117 or C- KIT suggestive of yolk sac tumour. Beta HCG - negative. Serum alpha fetoprotein level was elevated (5800 IU/L). Child was started on chemotherapy – bleomycin (15 IU/m2); cisplatin (20 mg/m2); etoposide (100 mg/m2) once in every 3 weeks. Follow up CT thorax after 6 chemotherapy cycles showed significant reduction in size of tumour. Serum AFP – 32 ng/ml





CT Thorax - Pulmonary tumour



Residual Tumour

DISCUSSION:

Germ cell tumours are classified as extra gonadal if there is no evidence of primary tumour in testis or ovary. These constitute 2-5% of all germ cell tumours. From 4th to 6 th week gestation, the germ cells migrate through midline dorsal mesentery. A remnant of tissue anywhere along the migration course can be the site for germ cell tumour, in future like mediastinum (most common), retro peritoneum, sacrococcygeal region, pineal gland and neurohypophysis. This tumour arises from malignant transformation of misplaced primordial germ cells. They are of 2 types ,

1)Benign (80%) – teratoma, teratodermoids.

2)Malignant (20%) - Non Seminomatous germ cell tumours (NSGCT) – teratocarcinoma, choriocarcinoma, embryonal carcinoma, yolk sac tumour and Seminoma or Dysgerminoma. Yolk sac tumour is defined as tumour characterised by numerous patterns that recapitulate the embryonic yolk sac, allantois, extra embryonic mesenchyme and show bimodal age distribution (16 to 18 months & 25 to 35 years). Most common complaints are cough, fever, chest pain, dyspnoea and weight loss. Incidence of haematological malignancies is 200 to 300 fold higher - most commonly acute myelogenous leukaemia and myelodysplasia with megakaryocyte abnormalities. Histologically, both extragonadal & gonadal tumours are same. Most common pattern is micro cystic or reticular pattern. Schiller duval bodies are pathognomic. Immunohistochemistry for Yolk Sac tumour is positive for AFP, Glypican-3, placental alkaline phosphatase. Markedly elevated AFP is virtually diagnostic and useful for prognosis. CT is the imaging modality of choice to determine the exact location of mediastinal tumour and its relationship to adjacent structures. These tumours are chemo sensitive and radio resistant. Treatment and prognosis depends on the type of cancer, tumour location and its size. Regimen - BEP (bleomycin, etoposide, cisplatin) 4 cycles at 3 weeks interval with residual tumour surgical excision after AFP levels are reduced to normal limits ,followed by 2 cycles of chemotherapy. If serum markers are elevated, give salvage chemotherapy (etoposide, ifosfamide, cisplatin). Thymectomy is performed routinely because thymus is often replaced totally by tumour. Long term follow up is needed to detect late recurrences and development of testicular tumour after initial diagnosis of Extra Gonadal Germ Cell Tumour.

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