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Research Paper

Primary Rhabdomyosarcoma of Diaphragm: Report of A Paediatric case

Medical Science

KEYWORDS : Rhabdomyosarcoma, Spindle cell, Diaphragm

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ABSTRACT

Rhabdomyosarcoma (RMS) is most common soft tissue sarcoma in children .But primary RMS arising from diaphragm is a very rare tumour, only few cases are reported in literature. Here we report a case of spindle cell RMS of diaphragm, stage III, in a 12yr old boy treating with neo-adjuvant chemotherapy.

INTRODUCTION:

RMS is highly aggressive soft tissue sarcoma, commonly occur in children.RMS arises from either undifferentiated unsegmented mesoderm or myotome derived skeletal muscle[1].Most commonly arises from head neck and genitourinary region.RMS originated from intrathoracic or retroperitoneal or paraspinal region is very uncommon with poor outcome. Primary tumour of diaphragm either benign or malignant are very rare; among them benign mesothelial and bronchogenic cyst commonest. Fibrosarcoma is common malignant lesion of diaphragm. But primary RMS arising from diaphragm is a very rare tumour, only few cases are reported in literature; spindle cell variety reported in only 3 instances. Here we present case of primary RMS of diaphragm in a 12 years old male patient.

CASE REPORT:

12 years old male patient admitted in pediatrics surgery department of our hospital with complaints of severe pain over left hemi thorax increasing during inspiration, gradual distension of abdomen along with significant loss of weight and appetite for last 3 months. Physical examination revealed distended, tense abdomen with a huge mass occupying epigastrium and left hypochondrium region. Breath sound diminished over left hemi thorax. Routine blood parameters revealed presence of decreased hemoglobin percentage and luecocytosis. Chest X-ray showed complete opacity of left hemi thorax with mediastinal shift to right. Contrast enhanced Computed Tomography scan (CECT) revealed large uncalcified heterogeneously enhancing abdomino thoracic mass 19.3x17x11.5 cm^3 sizes with internal necrosis. Left dome and crura of diaphragm are not seen separated from mass lesion. Superiorly the lesion abutting post inferior surface of heart and pericardium. No distinct fat plane between them. No obvious lymphadenopathy present. (Figure 1) CT guided trucut biopsy and histopathological examination showed malignant spindle shaped cell with eosinophilic cytoplasm positive by periodic acid Schiff staining compatible with embryonal RMS; spindle cell variety.

Immunohistochemistry showing immune-reactivity for Myo D1 & Desmin and immune-negativity for Cytokeratin, EMA, CD34 and S-100(Figure 2).He was diagnosed as Diaphragmatic spindle cell RMS, Stage III [According to Intergroup Rhabdomyosarcoma Study (IRS) pretreatment staging system] [1]

Patient was referred to us from surgical side as an unresectable and we planned for neo-adjuvant chemotherapy with VCD regimen (Inj Vincristine 1.5 mg/m2 IV Day1and Day21, Inj Cyclophosphamide 1200mg/m2 IV Day17-21, Inj Actinomycin D 0.5mg/m2 IV Day21-23, repeated 28 days interval) with proper pre and post medications. After 3 cycles of chemotherapy repeat CECT done (Figure 3) and comparison with previous one it was progressive diseases. We are planned to switch next line chemotherapy with MIAD regimen (Inj Ifosfamide 2000mg/m2 IV Day1-3 with Inj MESNA, Inj Doxorubicin 15mg/m2 IV Day1-4, Inj Dacarbazine 250mg/m2 IV Day1-4). Patient is under treatment with this chemotherapy regimen.

DISCUSSION:

RMS is a malignant tumor that can arise at any site of the body. Better supportive care and systematic application of increasingly effective multimodal treatment have dramatically improved survival over this period, with 5-year survival rates rising from approximately 10 to 20% in 1970 to about 60 to 70% currently. RMS are group of malignant primitive mesenchymal tumour; skeletal muscle origin. RMS accounts for 15% of soft-tissue sarcomas in the general population and 4% to 8% of all childhood malignancies[2]. Most of the patients are < 10 years of age at the time of diagnosis, <1 yr age at 5% cases and >10 yrs age 25% cases[3]. Histologically RMS comprises of 4 subtypes pleomorphic, alveolar, botryoid, and embryonal. Embryonal and botryoid varieties are common in infant arising from head- neck and genitourinary region. Alveolar variety more common in adolescent; mainly arises from trunk and extremity. Regional lymphatic spread occurs in 15% cases; more with paratesticular, trucal and extremity tumour and haematogenous spread occur in 15% cases[4].

Signs and symptoms depend on the anatomic site of the tumor, and prognosis varies according to tumor size, histology, localization, clinical group, and cytogenetic alterations[5]. Those arising in orbit, genitourinary tract (non bladder and prostate), head neck (non parameningeal) have the best prognosis. The unfavorable sites include extremities, retroperitoneal, intra- thoracic locations, bladder-prostate, parameningeal. Embryonal histology carries good prognosis where as alveolar variety is highly aggressive[6]. Spindle cell variety of embryonal RMS belongs to superior prognosis group (International classification of Rhabdomyosarcoma) with 5 yr survival rate 88% to 95%[7]. CT and MRI are highly effective in characterization and location of the RMS. While mediastinal involvement is well demonstrated with CT (*Marasco WJ et al*, 1991), MRI has been proved to be more helpful in defining local infiltration of chest wall and providing accurate information on the extent of diaphragmatic involvement (*Stark P et al*, 1994; *Almberger M et al*, 2001).CT or MRI of primary tumour along with PET scan is valuable to detect extend of disease[8]. Our patient belongs to IRS clinical group III; embryonal, spindle cell variety. However, the size of tumor and its location and his clinical group are indicators of poor outcome. For unresectable local or regional tumors in less favorable sites, radiation therapy is recommended early in the course of treatment. Radiation therapy was not delivered because our patient did not fit in these criteria. Initially our patient treated with VCD regimen, but as he developed progressive disease we shift to MIAD regimen because it includes the two most effective drugs against this tumor (ifosfamide and doxorubicin), aiming for a remission[9].

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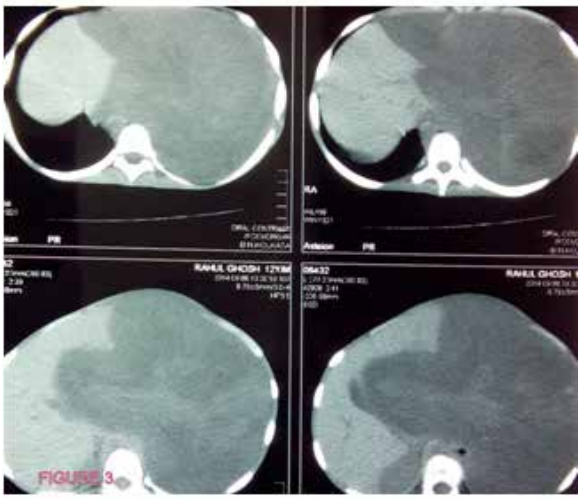
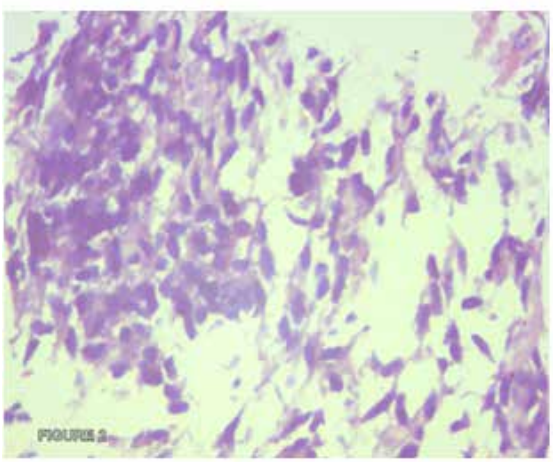
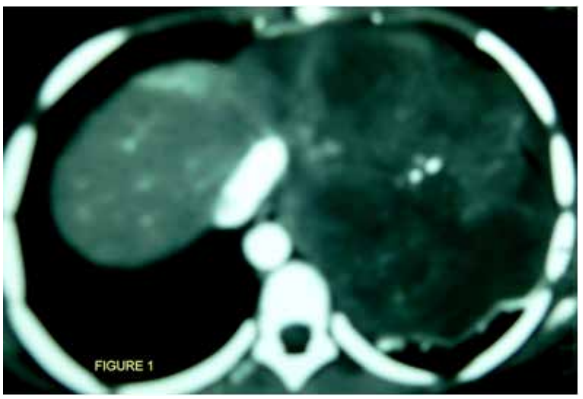
LEGENDS OF FIGURE 1: CECT Thorax reveals huge abdomino thoracic mass arising from left dome of diaphragm; involving posterior inferior of heart and pericardium.





LEGENDS OF FIGURE 2: i.e Immune-reactivity for Myo D1 & Desmin and immune-negativity for Cytokeratin, EMA, CD34 and S-100

LEGENDS OF FIGURE 3: CECT thorax and abdomen reveals increased size of mass compared to previous CTscan.



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