IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 16, Issue 11 Ver. IV (Nov. 2017), PP 36-38
www.iosrjournals.org

Intra- Abdominal Retroperitoneal Rhabdomyosarcoma in Adult: A Rare Case Report & Review of Literature

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Abstract: *Rhabdomyosarcoma (RMS) is one of the typical tumors of childhood and adolescence, representing over 50% of soft tissue sarcomas in those age groups. Soft tissue sarcomas make up > 1% of all adult malignancies, and RMS represents 3% of all soft tissue sarcomas. The most well-known primary tumor sites incorporate the head and neck district (35%), trailed by the genitourinary and extremity primaries. Intra-abdominal rhabdomyosarcoma is still rarer in adult population.*
A 61 years old male patient presented with complain of lump in right iliac fossa associated with burning pain but not associated with vomiting, nausea or loose motion or intestinal obstruction. CECT abdomen suggested retroperitonum tumor on right side below the level of kidney extending to midline and inferiorly below level of common iliac bifurcation & encasing the right common iliac & origin of external iliac artery and The infrarenal IVC & right mid ureter are compressed & completely encased by mass lesion. CT guided biopsy suggested Rhabdomyosarcoma, spindal cell type.
Rarity of this disease and location prompted us to report this case & review of literature. Our own is most likely the third instance of intra abdominal retroperitoneal rhabdomyosarcoma reported in literature.
Keyword*: Rhabdomyosarcoma (RMS), Adult, Intra-Abdominal, Soft Tissue Sarcoma*

Date of Submission: 01 -11-2017 Date of acceptance: 09-11-2017

I. Introduction

Rhabdomyosarcoma (RMS) is one of the typical tumors of childhood and adolescence, representing over 50% of soft tissue sarcomas in those age groups .Rhabdomyosarcoma are uncommon in adult, and the literature with respect to their management is restricted. Soft tissue sarcomas make up > 1% of all adult malignancies, and RMS represents 3% of all soft tissue sarcomas. RMS can emerge in an assortment of anatomic locales all through the body. The most well-known primary tumor sites incorporate the head and neck district (35%), trailed by the genitourinary and extremity primaries. Intra-abdominal rhabdomyosarcoma is still rarer in adult population. Because of the uncommonness of adult RMS, data with respect to its clinical and biologic qualities is exceptionally limited; large, multi-institutional trials have not been performed, and only case reports have been published. Our own is most likely the third instance of intra abdominal rhabdomyosarcoma reported in literature..

II. Case Report

A 61 years old male patient came to the gastrointestinal surgery division with complain of hard mass felt on right lower flank with both lower limb edema. He give on & off symptoms of fullness in abdomen after having meal associated with burning pain but not associated with vomiting, nausea or loose motion or intestinal obstruction since 6month. On examination his vitals were stable, abdominal lump felt on right iliac fossa with diffuse tenderness but no guarding and rigidity (Fig. 1).

CECT abdomen showed a large retroperitoneal mass lesion on right side below the level of kidney extending to midline and inferiorly below level of common iliac bifurcation size 16.5(SI)×16.3(T)×9.3(AP) cm. it is near completely encasing the right common iliac & origin of external iliac artery (Fig. 2). The infrarenal IVC & right mid ureter are compressed & completely encased by mass lesion. The ascending colon, duodenum are anteriorly displaced (Fig. 3). CT guided biopsy taken & impression was Rhabdomyosarcoma (RMS), spindal cell type. All other parameters were normal. As mass was inoperable & patient had no symptoms of intestinal obstruction, chemotherapy (inj. adriomycin, inj. ifosphamide & inj. mesna) was started.


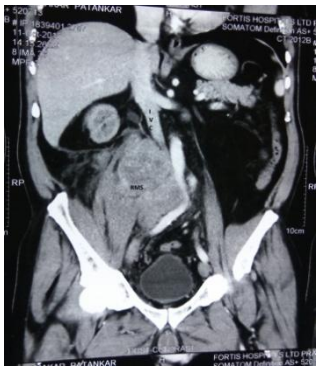
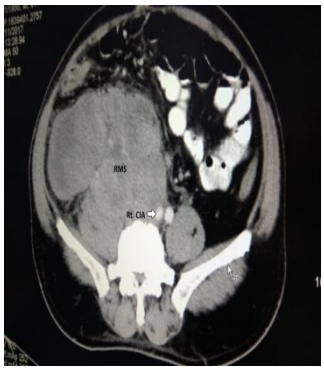

Fig.1 **Fig.2** **Fig.3**

III. Discussion & Literature Reviews

Rhabdomyosarcoma is thought to emerge from immature mesenchymal cells that are focused on skeletal muscle heredity, however these tumors are likewise known to emerge in tissues in which striated muscle is not normally found. Rhabdomyosarcoma (RMS) is one of the typical tumors of childhood and adolescence, representing over 50% of soft tissue sarcomas. Soft tissue sarcomas make up > 1% of all adult malignancies, and RMS represents 3% of all soft tissue sarcomas[1].The standard classification proposed by Horn and Enterline et.al. in 1958,into four subgroups: embryonal, alveolar, botryoid and pleomorphic and noticed that botryoid was really a subtype of embryonal [2] . Embryonal types occur in infants and toddlers, pleomorphic types arise in adults, and alveolar tumors seem to affect all age groups. Histologically, this age distribution corresponds to the histologic maturity of the RMS subtype, because embryonal types resemble embryonic tissues and pleomorphic types appear as aggressive adult cancers with malignant fibrous histiocytoma like features. However, exceptions do occur in occasional adults, and tumors with the histology of embryonal RMS or its spindle cell variant are occasionally reported [3, 4]. RMS can emerge in an assortment of anatomic locales all through the body. The most well-known primary tumor sites incorporate the head and neck district (35%), trailed by the genitourinary and extremity primaries [5]. Intra-abdominal rhabdomyosarcoma is still rarer in adult population. Patients with RMS can present with an asymptomatic mass or with signs and symptoms that are associated with the primary tumor site and are related to mass effect or complications that are secondary to the tumor. Imaging studies should include computed tomography (CT) scan or magnetic resonance imaging of the primary tumor to determine the size and possible involvement of vital organ structures; these are parameters to be considered when surgical resection is planned. Core needle biopsy may be performed for small lesions in areas that will be treated with chemotherapy/radiation or for metastatic disease [6].

There are three modalities of treating patients with RMS. These are surgery, systemic combination chemotherapy for primary cytoreduction and eradication of gross and micrometastases & radiation therapy for control of residual bulk or microscopic tumor and. Surgery incorporates complete resection of the primary tumor with encompassing edges of uninvolved tissue. In the event that microscopic residual disease is found after primary resection, re-excision of the area is indicated, before anyother nonsurgical modalities. Debulking systems have no value if CT scan suggested of inoperatibility or patient had no mass effect & intestinal obstruction symptoms and as initial biopsy and neo-adjuvant therapy results in shrinkage of the tumor allowing complete resection at second look operation [7].

The use of chemotherapy and its combination had been major point of discussion in adult RMS. Hawkins *et al*[8] concluded that there was no survival benefit with the use of chemotherapy in adult RMS. Few studies suggested that chemotherapy has same activity in adult and pediatric RMS, and when chemotherapy regimens are administered similarly to those used for treatment of pediatric patients, the outcomes for adults and children with RMS are comparable[9, 10, 11]. In a study by Ferrari *et al.*[10] the overall rate of response to chemotherapy was 85% and the authors emphasized that adults be treated on same lines as that of childhood RMS. The recommended combination of chemotherapy approach consisting of vincristine, actinomycin-d, and cyclophosphamide (VAC); mesna, Adriamycin, ifosphamide (MAID); other doxorubicin based regimen; other ifosphamide based regimen.

The dose, duration and timing of radiotherapy depend on the clinical group and the site of disease. Induction chemotherapy, followed by concurrent chemoradiation, is the current standard of care for patients with unresected disease, those with micro- or macroscopic residual disease after surgery for patients with lymph node involvement, and for patients with alveolar histology [12, 13]. RT is usually initiated between 6–12 weeks after the start of chemotherapy, omitting the concomitant administration of a radiosensitizing agent, such as an anthracycline or actinomycin D. The dose of RT is dependent upon several variables usually related to the extent of the disease, such as regional lymph node involvement.

Iyad sultan et al. in there study reported that adults with rhabdomyosarcoma had significantly worse outcome than children (5-year overall survival rates, 27 %±1.4 % and 61 %±1.4 %, respectively; P<.0001). Tumors in adults were more likely to be at an unfavorable site (65 % v 55 %; P<.0001) and to have histologies that are unusual during childhood, particularly the pleomorphic subtype (19 %) and not otherwise specified (43 %) [14]. Favorable prognostic factors in RMS are: undetectable distant metastasis; favorable anatomic destinations (orbit, nonparameningeal head/neck, and genitourinary nonbladder/prostate regions); grossly complete surgical removal of the localized tumor at the time of diagnosis. Age, location, nodal status, and histologic subtype do not appear be associated with survival in adults with rhabdomyosarcoma treated with multimodal therapy. Metastatic disease at presentation and poor response to chemotherapy are emphatically associated with poor prognosis. Future systemic therapies should be targeted to patients with localized/locoregional disease and partial responders to conventional chemotherapy and radiotherapy [15].

Our case is rare presentation of intra-abdominal rhabdomyosarcoma in adult.Andrew M. Kaplan et al. [16] reported probably the first &Sanjay Kumar Yadav et al [17] reported second well documented case report of non-hepatobiliary, adult, intraabdominal rhabdomyosarcoma in the English language literature. Our own is most likely the third instance of intra abdominal retropertonial rhabdomyosarcoma reported in literature.

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