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RESEARCH ARTICLE

A RARE CLINICAL CASE SERIES OF LEIOMYOSARCOMA OF SOFT TISSUE ORIGIN AT OUR INSTITUTE

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Abstract

Introduction:- LMS is a type of rare cancer that grows in the smooth muscles of the body. LMS is a aggressive cancer. LMS incidence is between 5-10% of soft tissues sarcoma cases and is more common in adults. It often present as an enlarging painless mass. Imaging with proper Biopsy and IHC aids in the diagnosis and management incudes a multidisciplinary approach.

Objectives:- Due to rarity of this disease case series of LMS of soft tissue origin is documented here.

Material and Methods:- 3 cases of primary LMS of soft tissue origin is documented in this case series along with history, imaging, diagnosis and treatment. 1. LMS of Lung – A 51 year old female patient presented with complaints of cough and difficulty in breathing since 3-4 months. Patient was planned with chemotherapy. 2. LMS of Pelvis – A 44 year old female patient presented in OPD with complaints of pain in leg due to nerve compression. Patient underwent surgery followed by which chemotherapy was given. 3. LMS of Pararenal Space – A 33 year old female presented with dull aching pain in left lumbar region since 2-3 months. Patient managed with surgery, chemotherapy and radiotherapy.

Conclusion:- Due to rarity of the sites of LMS mentioned in the case series little is known about its clinical behaviour and treatment outcome. A multidisciplinary approach is needed for the optimal management of the disease. Surgery with a curative intent is the corner stone of treatment for localized disease along with the combination of neoadjuvant or adjuvant chemotherapy and radiotherapy. Due to its aggressive behaviour and rarity, further researches are required for the definitive treatment.

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Introduction:-

Case1:-

Lms Of Lung:-

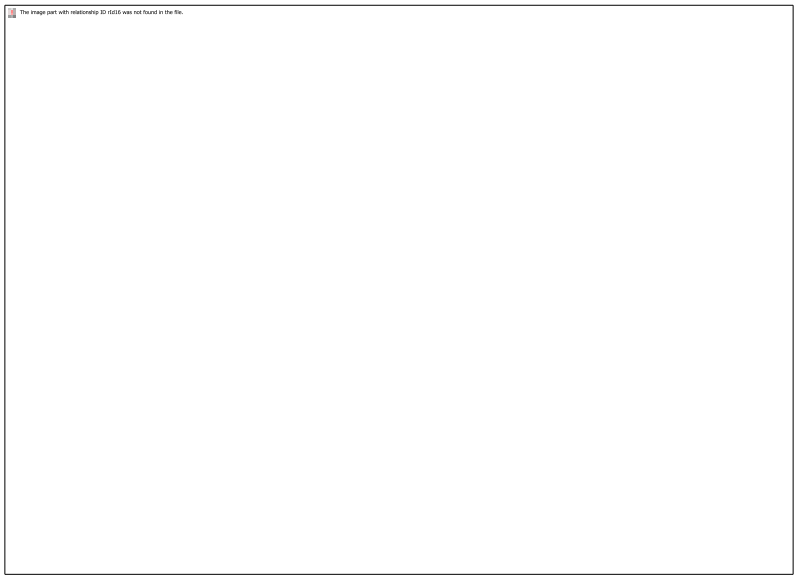
Primary pulmonary LMS (PPL) is an extremely rare malignant tumor originating from smooth muscles of pulmonary parenchyma , pulmonary arteries and bronchi. It accounts for < 0.5 % of all malignant pulmonary tumors (1). Patients with PPL may be asymptomatic or may present with symptoms those similar to primary lung cancer. Diagnosis based on radiological finding of CT scan. Biopsy and IHC markers are mandatory for the diagnosis of PPL. If surgically operable surgery is definitive and primary mode of treatment like lobectomy , pneumonectomy or bronchial sleeve resection procedures can be performed. Radiochemotherapy plays an important role in case of incomplete resection of tumor or if recurrence is there. Early detection and complete surgical resection of PPL contributes to an increased survival time of patients (3) .

Case Report:-

A 51 year old female patient with complaints of cough with expectoration and difficulty in breathing since 3-4 months presented in our OPD. There was no history of addiction and no significant family history. The patient had no history of fever (2), anorexia, hemoptysis, chest pain or weight loss. The vitals were normal. Physical examination revealed dullness to percussion , decreased tactile tremitus and asymptomatic chest expansion lung auscultation revealed decreased breath sounds on the right lung. Other physical examination were normal. Laboratory examination were normal including bloodchemistry , blood tumor marker , liver and renal function test , sputum examination was negative for acid fast bacilli.

HRCT Thorax done revealed gross pleural effusion on right side with 7.9X 11.5 X 12.3 cm relatively well defined soft tissue density mass lesion in RUL. Along with multiple variable sized nodular soft tissue density lesion in left lung parenchyma largest 1.2 x 1.2 cm f/s/o – Malignant neoplastic etiology with lung metastasis. Trucut biopsy from right lung mass demonstrated that the tumor was composed of spindle cells, with marked nuclear pleomorphism and numerous mitotic figures. There was no epithelial differentiation in the tumor. Immunohistochemical staining demonstrated the strong expression of vimentin , smooth muscle antigen with calretinin positive in mesothelial cells. PCK, CD34 , TTF1 and EGFR were negative. The cell proliferation index of the tumor using antigen Ki 67 was 15-20 %. The feature were in favour of high grade sarcoma with evidence of smooth muscle differentiation indicating leiomyosarcoma.

CT Thorax:- suggestive of 9.8 X9.4 cm well defined soft tissue density mass in right upper lobe.



H & E report showing spindle cells with marked nuclear pleomorphism and numerous mitotic figures.

Patient underwent CT scan of thorax and abdomen suggestive of large bronchocentric hilar, parahilar soft tissue density lesion 9.8 X 9.4 cm involving upper and middle lobe region on right side with peribronchial and endobronchial extension involving the lobar bronchus. Medially lesion appears to involve the carina , mediastinal pleura. Superiorly having broad base indentation over the RPA. Laterally abducts the right lateral chest wall and endothoracic region with multiple pulmonary nodules 10 - 15 mm diameter involving both lung field. There were no other metastatic lesion observed elsewhere. As the disease was not surgically resectable , patient was planned for 3 cycle of Chemotherapy AI regimen. (inj Adriamycin 60mg/m2 on day 1 , inj ifosphamide 1.5gm/m2 day 1-4, inj mesna 225mg/m2 day1-4).

Follow up study :- Post 2 cycles of chemotherapy patient experienced sudden breathlessness at home and succumbed.

Discussion:-

PPL is a rare mesenchymal tumor originating from the smooth muscle cell of the bronchial or blood vessel wall. PPL can be classified into intraluminal, intrapulmonary and pulmonary vascular (4) , out of which intrapulmonary type of PPL is most common. Most of the patient are symptomatic. In ordery to differentiate PPL from bronchogenic carcinoma biopsy and IHC are must. Vimentin is a strong mesenchymal tumor marker. Proper radiological imaging with CT , MRI or PET Scan for the detailed delineation of tumor and to rule out metastasis is required. Treatment of PPL ends to achieve local and systemic control of the tumor, while preserving function and quality of life. If surgically operable surgery should be definitive approach. While surgery , chemotherapy and radiotherapy all contributes as multidisciplinary approach for the proper management of such tumors. Prognostic indicator of PPL consist of tumor size, extent of bronchial invasion and degree of malignancy.

Conclusion:-

PPL is a rare tumor that grows rapidly and its diagnosis is challenging to differentiate PPL from other pulmonary tumors. Early diagnosis and treatment is mandatory. The goal of the treatment is to obtain local and systemic control of the sarcoma along with preserving functioning and quality of life. An increase awareness of PPL leading to an early diagnosis and the performance of a complete surgical resection with adjuvant radio and chemotherapy in the selected patient may improve the prognosis of the people with PPL. Otherwise PPL is a very aggressive tumor with increased morbidity and mortality. Due to its aggressiveness and rarity further research are required for the definitive treatment so that morbidity and mortality could be reduced.

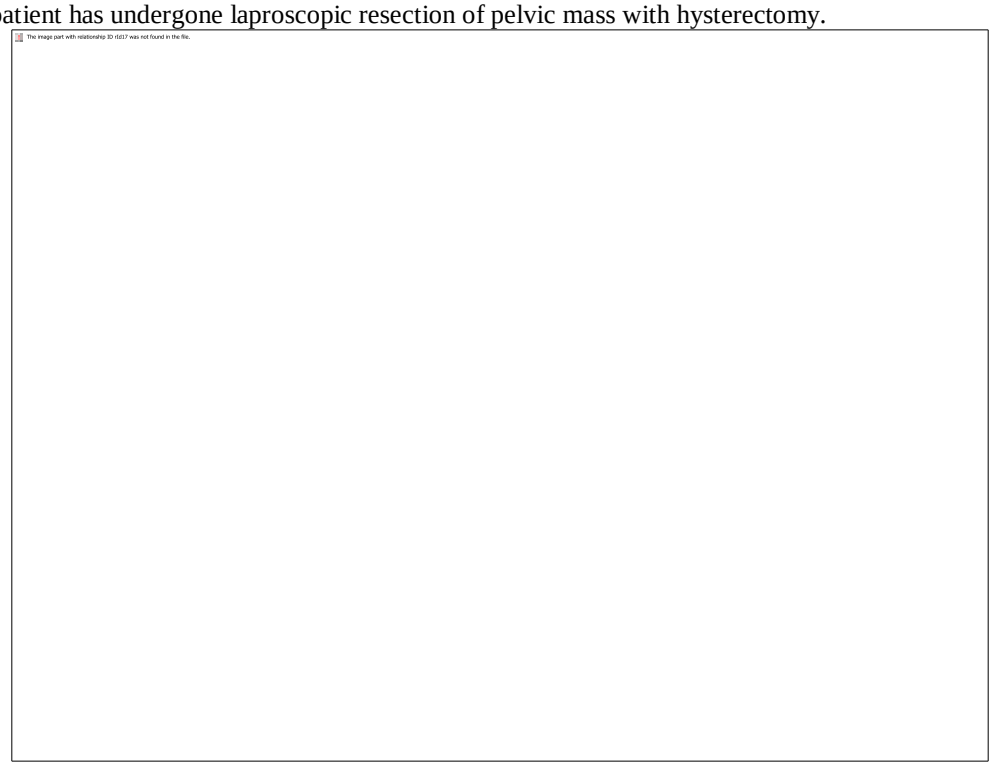
Case 2:-

Lms Of Pelvis:-

LMS is a soft tissue sarcoma derived from smooth muscle. The most frequently involved organ are the retroperitoneal space, extremities and uterus. LMS can occur at any site however there is a lack of convincing data on pelvic non uterine LMS. The biological behaviour including the associated metastasis and prognosis required further exploration. Biopsy and IHC are must for diagnosis. Usually the patients of extrauterine LMS present with abdominal pain, distension and lump in the pelvis. CT Scan, MRI, PET and USG are used in the examination and follow up of such patients. The treatment methods and prognostic factors require extensive research because of the disease high malignant potential. Surgery, chemotherapy and radiotherapy as a multimodality approach are required for the treatment. (5)

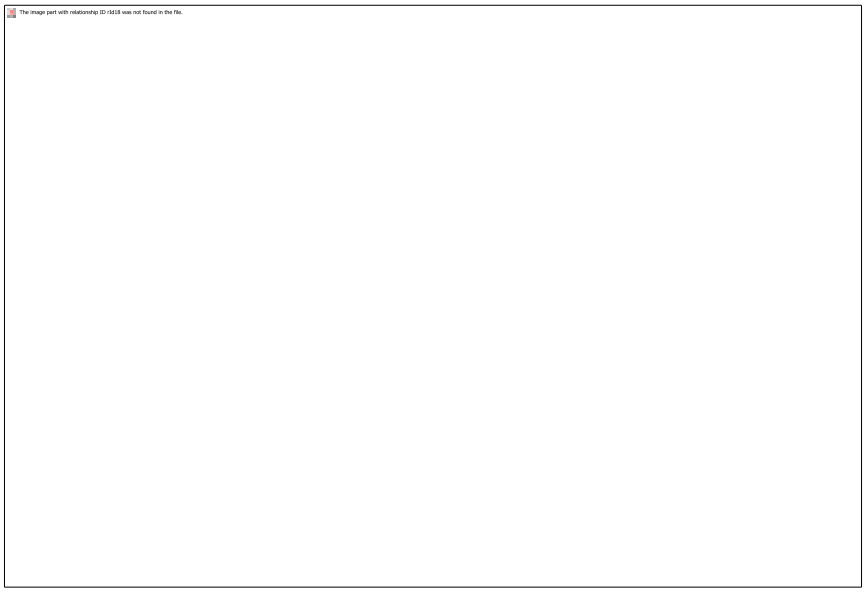
Case History:-

A 44 year old female patient had complaints of pain in leg due to nerve compression in year 2019. On USG there was an incidental finding of pelvic mass. Biopsy of the lesion showed spindle cell neoplasm. IHC done on imaging report showed Desmin, SMA positive ,MyoD , S100 Negative and Ki 67:- 15- 20 % suggestive of leiomyosarcoma. For which patient has undergone laproscopic resection of pelvic mass with hysterectomy.



IHC report positive for Desmin marker.

No preop imaging and post op histopathological report of the patient was available. Patient had history of receiving inj Doxorubicin from 12/5/20 to 31/8/20 outside in private hospital. There was disease free interval of 2 years. Patient presented with pedal edema in our OPD in January 2022. Clinical examination revealed a hard pelvic lump of size 9 X10 cm with illdefined margins and a laparotomy scar. CT Scan done on 18/01/2022 was suggestive of lobulated heterogeneously enhancing solid cystic lesion in pelvis extending into abdominal cavity more on right side into subhepatic region size 5.8X9.8X8cm with moderate ascities suggestive of recurrence.



CT Scan suggestive of lobulated solid cystic lesion in pelvis right side subhepatic region 5.9 X 9.8X8cm.
Serum CA 125 was 214.86 U/ml. Second line chemotherapy with 3 cycles ofinj Gemcitabine and inj Docetaxel was given. Last cycle on 29/09/2022. CT scan done on 14/11/22 showed solid lesion of size 14.5 X 20 X 18.5 cm against previously mentioned 5.8X9.8X9 cm in pelvis adherent to pelvic wall abutting aorta and partially encasing bilateral common and internal iliac arteries, adherent to sigmoid colon at few places. 7.1 X12.2 X 10.5cm lesion noted anterior to right kidney displacing it with moderate ascities and pleural effusion. Patient was started on inj Eribulin based chemo and received 2 cycles of chemotherapy. Post 2 cycles due to disease progression with gross ascities and pleural effusion patient was admitted in ward and succumbed during treatment.

Discussion And Conclusion:-

Tumor stage, size, relapse status and mitotic index are associated with overall survival and progression free survival in all leiomyosarcoma. The disease stage at diagnosis is an important predictor of survival. (6) There is lack of standardized treatment guidelines and specialized investigation focusing on nonuterine LMS of the pelvic cavity. Thus there is an urgent need to identify clinical characteristics of nonuterine LMS to provide early diagnosis and proper treatment.

Case 3:-
Lms Of Pararenal Space
Introduction:-

The retroperitoneum is an anatomical space located behind the abdominal or peritoneal cavity. These space are the anterior pararenal space, posterior pararenal space and the perirenal space. LMS are the second most common sarcomas to affect the retroperitoneum accounting for 2.8% of cases (9) and may grow to large sizes before detection and may be an incidental finding. LMS mostly arises from the large blood vessels in the retroperitoneum including the IVC and renal veins affecting women more commonly (7) . Mostly retroperitoneal LMS present as a large soft tissue masses and when symptomatic may cause compressive symptoms including pain. Biopsy is mandatory for diagnosis with the typical microscopic pattern of intersecting sharply marginated fascicles of spindle cells. The most useful immunohistochemical stains are SMA , Desmin and H- Caldesmon which are positive in more than 70% of cases. Additionally, focal positivity for Keratin, EMA , CD34 and S100 proteins may be seen. Important genetic predisposition to LMS are seen with Li-Fraumeni syndrome and the hereditary form of Retinoblastoma.

The imaging is the important part of evaluation which includes CT Scan , MRI and if possible PET CT scan. The AJJC on cancer staging TNM staging (8) system is the most common method of staging. The staging is based on pathologic findings and includes tumor size , nodal status, metastasis and tumor grade. If the patients performance status allows, first line treatment consist of surgical resection with the goal of negative margin. The use of

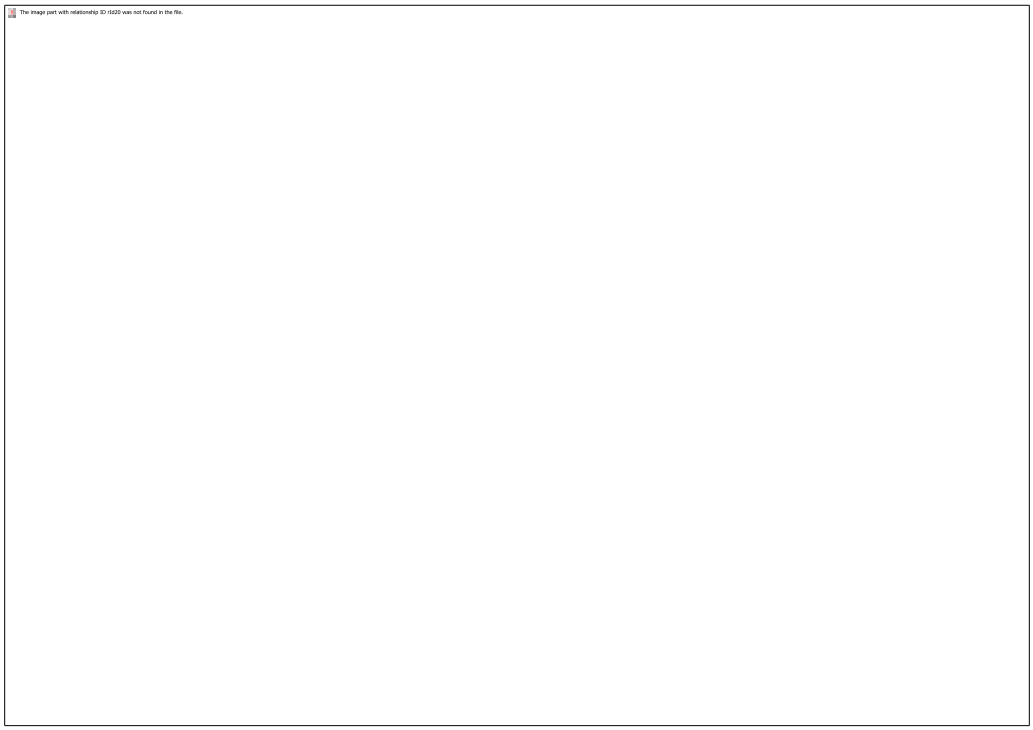
chemotherapy and radiotherapy has been described in both the adjuvant and neoadjuvant settings. Retroperitoneal LMS are often fatal owing to local recurrence and distant metastases.

Case History:-

A 33 year old female presented with dull aching pain in left lumbar region since 2-3 months. Patient has history of hypothyroidism since 2 years. Rest personal and family history was not significant. On clinical examination Per abdomen finding was a bulky mass of approximately 8 x 6 cm in left lumbar region with ill defined margins extending towards posterior aspect. Lump was mobile and non tender. Blood investigation were within normal limit and no serum tumor markers were done. USG A+P was suggestive of well defined lobulated hypoechoic lesion of size 6.4 X 6.5X8.3 cm in upper pole of left kidney and splenic hilum abutting the pancreas with increased vascularity around the lesion. CECT A+P was suggestive of a solid mass lesion measuring 6.3X7.1X7.8 cm with necrotic area in anterior pararenal space abutting body and neck region of the pancreas without infiltration and posteriorly related to perirenal space compressing left adrenal gland.

CT scan suggestive of solid mass measuring 6.3 X 7.1 X 7.8 cm in anterior pararenal space abutting body and neck of pancreas.

Patient undergone left adrenalectomy on 28/04/23. Intra operative finding was a mass in left suprarenal space 8X7 cm in size, soft, encapsulated. Adrenal gland was adhered to the mass with dense vascularity all around the tumor. Post operative HPR was suggestive of gross encapsulated nodular, globular form tissue 9X8X7 cm with spindle shaped cells and elongated nuclei. IHC done was immunoreactive for Desmin , SMA, CD34 and H- Caldesmon , Ki67 was 15-20% feature suggestive of leiomyosarcoma.



IHC positive for SMA marker.

Patient was then planned for AI regimen, inj Adriamycin 60mg/m2 on day 1 ,injifosphamide 1.5gm/m2 day1-4 , injmesna 225mg/m2 day 1-4. Post 4 cycles of AI regimen PET CT SCAN was done which showed no uptake at post op site or distant metastasis. Patient is now planned for post op radiotherapy to a dose of 60Gy/30#.

Discussion:-

Retroperitoneal LMS are often typically large at diagnosis and are difficult or impossible to resect with clear margins. Radiological imaging play a key role in determining the possibility of surgical resection. 4 cycles of post op chemotherapy in the form of AI regimen was given to the patient and now patient is on radiotherapy to the post op site. Patient tolerated chemotherapy well. ‘

Conclusion:-

Retroperitoneal LMS is an important malignant neoplasm with absent or non specific clinical symptoms. Pathologic features with biopsy and IHC confirms the diagnosis. Radiologic imaging plays a vital role for staging and for surgical approach , as well to differentiate it form other differential diagnosis. Multimodality approach as required for the treatment including surgery, chemotherapy and radiotherapy. Long term disease free survival and overall survival is under assessment.

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