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RESEARCH ARTICLE

LEIOMYOSARCOMA MIMICKING AN ADRENAL GLAND TUMOR

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Abstract

Leiomyosarcoma is a malignant smooth muscle tumor. It is the second most common primary retroperitoneal tumor behind liposarcoma. The retroperitoneal léiomyosarcoma represents 11% of all retroperitoneal malignant tumors. The age of discovery is between 40 and 60 years.

Introduction:-

Leiomyosarcoma is a malignant smooth muscle tumor. It is the second most common primary retroperitoneal tumor behind liposarcoma. The retroperitoneal léiomyosarcoma represents 11% of all retroperitoneal malignant tumors. The age of discovery is between 40 and 60 years. The sex ratio M/F is 1/6, clearly for the female sex.1,2.

Observation:-

We report the case of a patient with retroperitoneal Leiomyosarcoma treated in our institution.

She is a 60-year-old patient with no pathological history, consulted for diffuse abdominal pain with a feeling of heaviness. On physical examination, the abdomen was painful as a whole without visceromegaly..

The abdominal CT showed a large tumor mass with intraperitoneal development, forward of the pancreas, the left kidney and the adrenal gland, retro gastric, and contracting intimate relations with the wall of the colon, of lobule contours and heterogeneous density and presents some microcalcification.

The patient underwent a complete surgical excision removing the left adrenal gland; the surgical specimen weighing 200g and measuring 11x8x6 cm. The neighboring structures were unharmed.

Surgical pathology of the operative specimen confirmed the diagnosis of Leiomyosarcoma.

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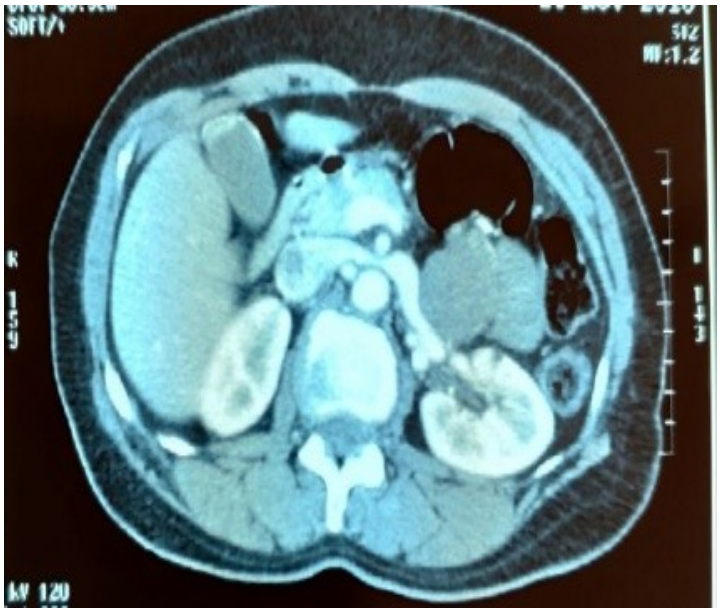
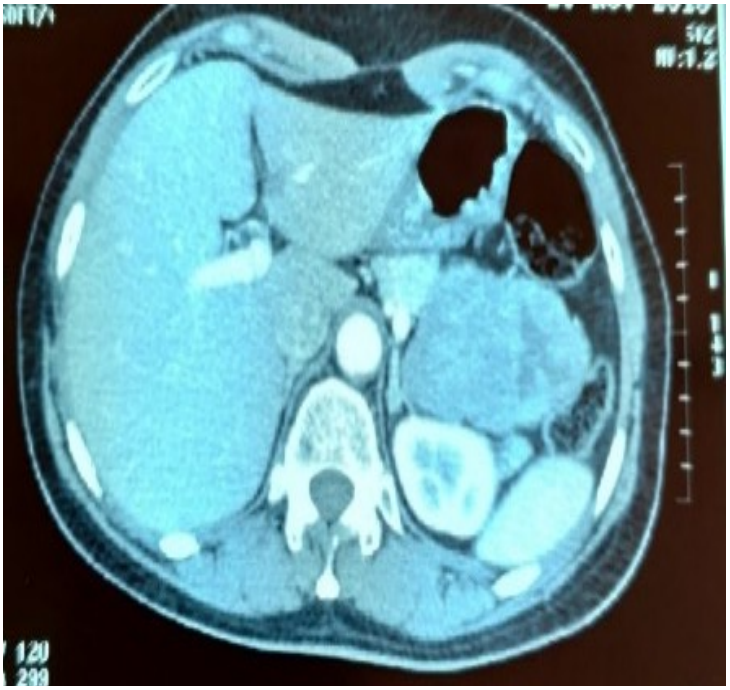
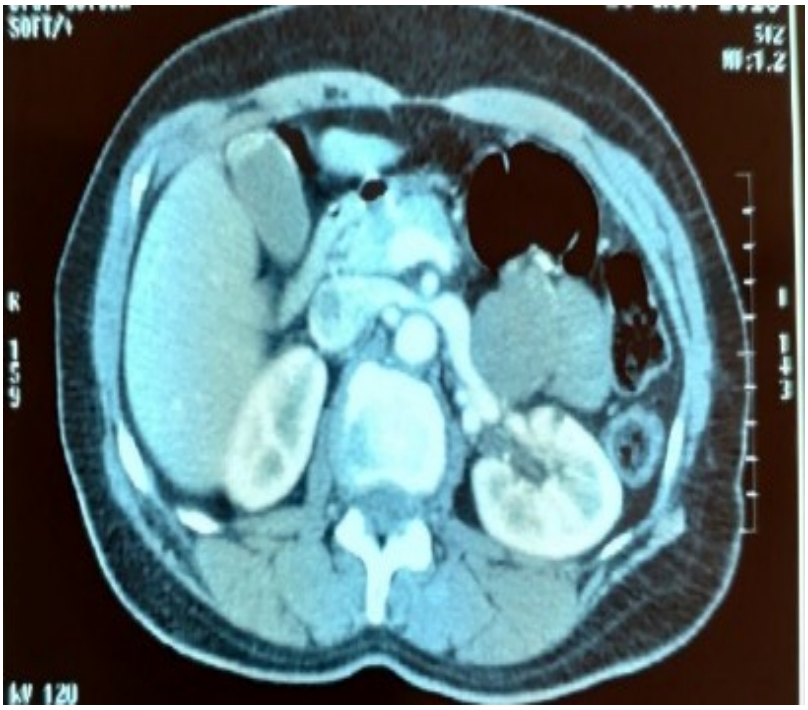
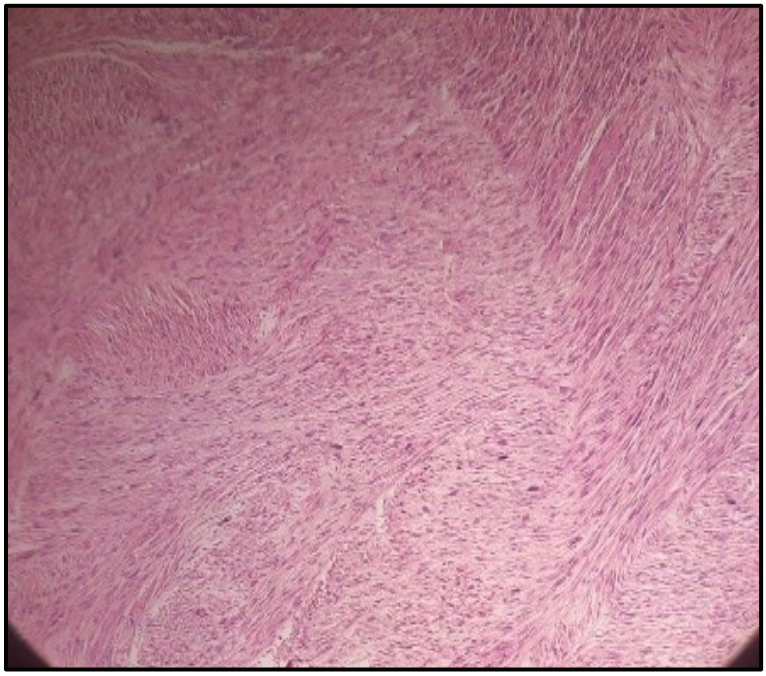






A transverse contrast-enhanced CT scan of the abdomen showed a large retroperitoneal mass



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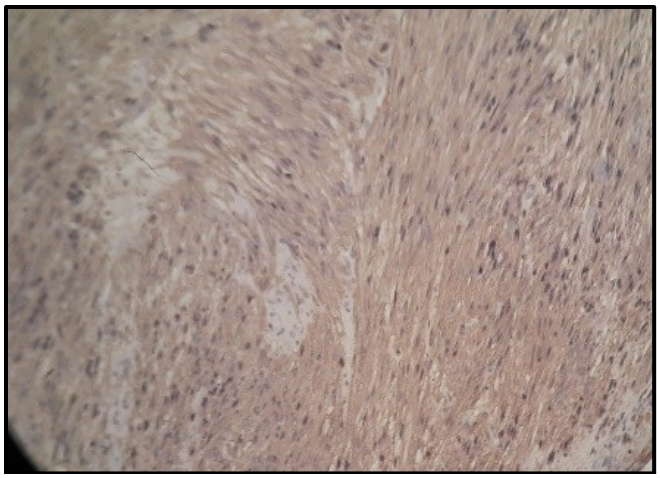
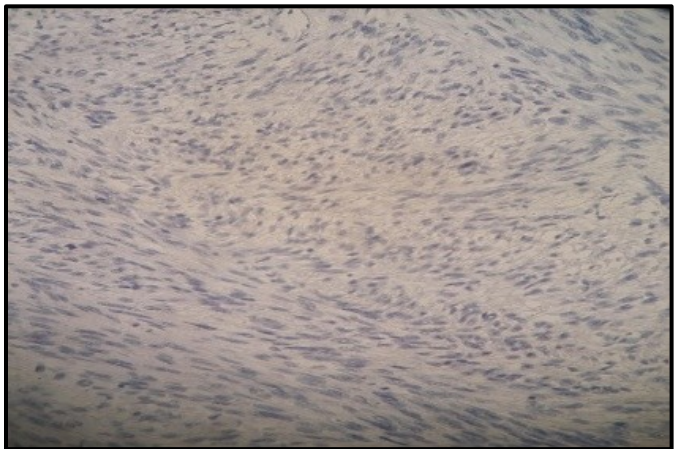


Overexpression of anti-caldesmon antibody x400.



Lack of antibody marking CD117 x400.

Discussion:-

The retroperitoneum is an expansible cavity; that is why this tumor can remain asymptomatic for a long time and be discovered in an advanced stage. Most often revealed between the fifth and seventh decade.[3]

Clinical symptoms are late and not specific. It results from the development of the tumor in the retroperitoneum and the compression of the adjacent organs: pain, digestive symptoms (nausea, vomiting, constipation), venous or lymphatic obstruction.

The discovery of a retroperitoneal tumor (TRP) less than 5 cm is rare and often lucky. Obstruction of the urinary tract is uncommon and is caused by the progressive development of the tumor. When the diagnosis is very late, the physical examination detects a large abdominal mass;[3,4]

CT and MRI are the two radiological exams of choice for the retroperitoneum. Indeed, a CT scan is recommended to detect, characterize, preoperative assessment, and monitor TRP.

CT scan confirms the tumor's retroperitoneal origin, carries out a complete extension assessment at both supra and infra-diaphragmatic level (search for lung, bone, hepatic and peritoneal metastases) to ensure post-therapeutic follow-up, the tracking of a locoregional recurrence. Finally, it allows the realization of a scanned biopsy.

MRI helps check the intraspinal extension of tumors close to the spine, studying relationships with vascular axes, and muscles invasion.[1,4]

The diagnosis of Leiomyosarcoma is based on histology and immunohistochemistry, highlighting fuso-cellular elements with a hyper-chromatic nucleus. The number of mitoses, pleomorphism, and necrosis determines malignancy.

Immunohistochemistry shows a positive reaction to antibodies anti-Vimentin and anti-Actin, and a negative reaction to antibodies anti-PS100, anti Desmine, and anti-Cytokeratin.

The standard treatment is based on complete excision of the tumor and its locoregional extensions in a single piece. Getting healthy margins requires surgeons to perform extensive resection of the organs next to the tumor, even if they are apparently not invaded, particularly the kidney and the digestive segments attached to the tumor.[5]

Indeed, the quality of the surgical excision is constantly the determining prognostic factor in all multifactorial studies.

The quality of the tumor resection must be defined according to the criteria of the UICC (International Union against Cancer):
RX = Presence of residual tumor cannot be assessed
R0 = No residual tumor
R1 = Microscopic residual tumor
R2 = macroscopic residual tumor
the survival rates after five years being: R0: 70%, R1: 45%, and R2: 15%.[6]

The TRPs, by their volume, repress the various abdominal viscera and are only separated by pseudo-capsule rich in tumor cells.[4]

Preoperative radiotherapy impacts local control and is well tolerated. In particular, the contribution of innovative techniques, Intensity Modulated Radiotherapy (IMRT), is prominent and allows the reduction of toxicities.

Chemotherapy should be systematically discussed in order to improve local and systemic control. Based on protocols comprising anthracyclines and alkylating agents and prescribed as induction for high-grade tumors, it facilitates the surgical procedure and limits the functional sequelae.[4,7]

Intraperitoneal chemotherapy has given disappointing results. In a metastatic situation, chemotherapy gives a response rate that does not exceed 20% to 30%. The histological grade established according to the criteria of the National Federation of Cancer Control Centers "FNCLCC" constitutes a determining prognostic factor for recurrence and overall survival. Grade III tumors recur five times more than low-grade tumors.

The quality of the first tumor resection is also a main prognostic factor. Survival reaches 91% at three years in R0 surgery, whereas it is only 48% after surgery in positive margins.

Therefore, it is not surprising that specific tumor locations, such as aortic, superior mesenteric, or vena cava involvement, may be correlated with a poor prognosis.

The follow-up makes it possible to detect local or peritoneal recurrences seen in 44% to 85%. The management of recurrence is multidisciplinary, involving surgeons, oncologists, and radiotherapists because of their poor prognosis and a frequently higher grade of malignancy than the primary tumor.

The surgery of metastatic recurrences is possible if their number is limited, if the time of occurrence compared to the initial treatment is sufficient, and especially if they are extirpable in full.

The other lesions require chemotherapy or supportive care. The prognosis for metastatic recurrence is reserved, with a median survival of around 12 months.[2,5,7]

Conclusion:-

Retroperitoneal leiomyosarcomas are rare tumors whose symptoms are insidious, linked to the mass effect. The scanner and the MRI make up the essential complementary examinations. Surgery remains the treatment of choice.

The quality of the surgical excision determines survival without recurrence and overall survival. High-grade tumors or positive margins are part of a multimodal approach combining chemotherapy and radiotherapy.

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