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RETROPERITONEAL LEIOMYOMA: A CASE REPORT

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ABSTRACT

Tumors of the retroperitoneum often present as clinical challenge because of poorly accessible location and late presentation with vague symptomatology. Primary tumors of this site include those of kidney, pancreas, adrenal gland, lymph node, germ cells and soft tissue; the benign neoplasms outnumbered by the malignant counterparts. Soft tissue tumors, predominantly smooth muscle, neural and adipocytic, are seen less commonly, accounting for almost 15% of the tumors of this region. Primary leiomyoma of the retroperitoneum is an extremely rare occurrence and should be diagnosed with utmost prudence. We hereby present a case of a lady with a rapidly growing large retroperitoneal mass suspected to be a malignant tumor of unknown nature but post surgical resection turned out to be a case of retroperitoneal leiomyoma.

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INTRODUCTION

Leiomyoma represents the most common gynaecologic neoplasm. Most of them are detected in women of middle aged group. These histologically benign tumors, which originate from smooth muscle cells, usually arise in the genitourinary tract but may arise in nearly any anatomic site.[(1)] Although there

have been reports on various atypical localizations for leiomyomata, their occurrence in the retroperitoneum is extremely rare.[2] Most of the cases in literature diagnosed clinically as retroperitoneal growths, were looked at with high suspicion of malignancy without suspecting their leiomyomatous nature.[3] The retroperitoneal origin of such leiomyomata is still an issue of dispute.

CASE REPORT:

A 38-year-old multiparous woman was referred to our Cancer surgery department with the 6-month history of vague abdominal discomfort, occasional nausea, vomiting and urinary incontinence. The previous medical history including gynecological was noncontributory. Her general condition and vital signs were normal. On examination, there was an extremely large firm painless palpable abdominal mass with smooth surface, extending from the suprapubic region to the epigastrium and completely filling the abdominal cavity. Except for mild neutrophilia, all other parameters including tumor marker levels (CEA, CA19-9, CA125 and AFP) were normal. Systemic examination, including gynecological, did not reveal any abnormalities.

Ultrasonography (USG) revealed a large 25 ×22 cm in diameter solid tumor, filling most of the abdominal cavity. Abdominal computed tomography (CT) confirmed USG finding revealing a large well encapsulated intraabdominal tumor 30×27×23 cm in size, extending from the pelvis to epigastrium and almost completely filling the pelvic and abdominal cavity and compressing the right kidney and colon. Since a definitive diagnosis was not possible and the mass was large and rapidly growing, the patient was taken up for surgical exploration laparotomy. At laparotomy, a huge encapsulated firm tumor was seen arising from the right retroperitoneal region and extending to the whole abdominal cavity. The mass was ligated at the base and resected completely, including hysterectomy and resection of colonic segment. Informed consent was obtained from the patient for use of the clinical data. Histopathological examination revealed intermediate sized spindle shaped cells with spindled to ovoid nuclei, arranged in bundles and cords. The capsule was intact, thin and fibrous (Figure 1,2).

A tentative diagnosis of benign spindle cell lesion was made and as leiomyoma is the first differential diagnosis, immunohistochemistry for smooth muscle actin (SMA) was performed which turned out to be positive, thus making a final diagnosis of retroperitoneal leiomyoma (Figure 3,4).

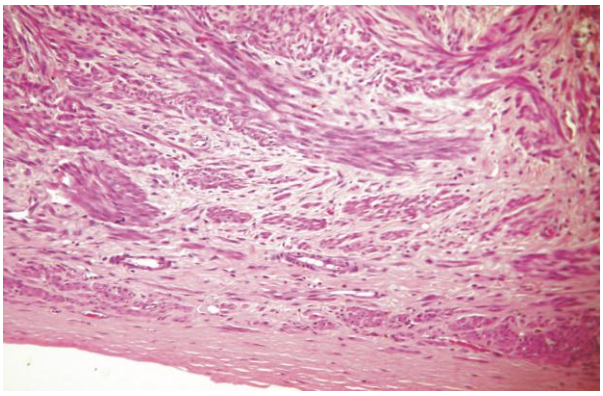

Figure 1: Photomicrograph showing capsulated tumor composed of spindle shaped cells (Hematoxylin & Eosin x 100).

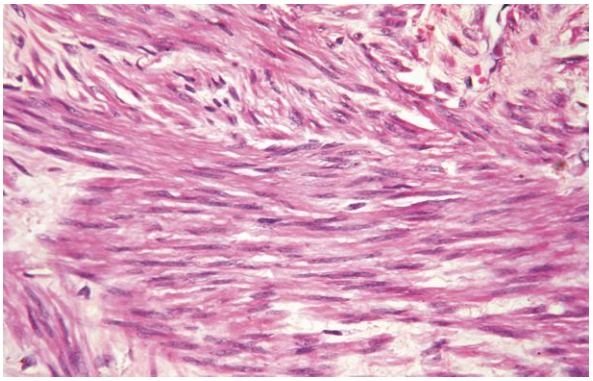

Figure 2: Spindle cells with cigar shaped nuclei without atypia (Hematoxylin & Eosin x 200).

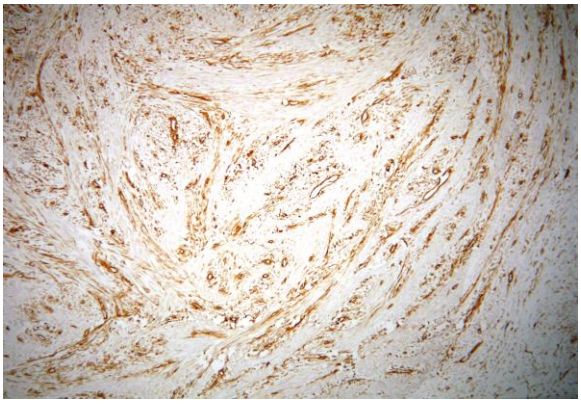

Figure 3:Tumor cells positive for Smooth muscle actin (x 40).

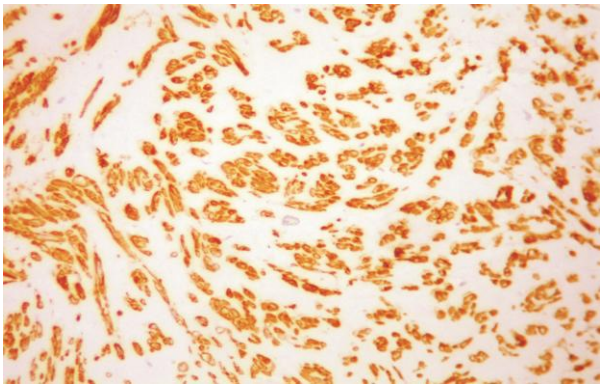

Figure 4: Cytoplasmic positivity for Smooth muscle actin (x 200).

Post-operative period was uneventful with no symptoms or recurrence reported at six months follow up.

DISCUSSION:

The retroperitoneal space is limited anteriorly by the peritoneal covering, posteriorly by posterior abdominal wall, superiorly by the 12th rib and vertebra, inferiorly by the base of the sacrum and iliac crest, and laterally by the site borders of the quadratus lumbora muscles. It contains: connective tissue, the adrenals, kidneys and ureters, aorta and its branches, inferior vena cava and its tributaries and lymph nodes. The relative paucity of vital structures, and the abundance of loose connective tissue in this area, results in a generally late clinical presentation of space occupying lesions.[4] Symptoms tend to be related to gastrointestinal, urinary or vascular

compromise, when large lesion size and compression/invasion of adjacent structures severely limits the curative treatment options.

In adults, retroperitoneal neoplasms primarily include lymphoproliferative (Hodgkin's and non-Hodgkin's lymphoma), parenchymatous epithelial tumors (renal, adrenal, pancreas) and metastases. Soft tissue (mesenchymal) tumors at this site are less common, sarcomas accounting to 15%, representing the second most common site for the origin of malignant mesenchymal tumors, after the deep tissues of the lower extremity. It has been observed that at all sites, benign soft tissue lesions outnumber their malignant counterparts by a ratio of at least 100:1. On the other hand, in the retroperitoneum sarcomas are more prevalent than their benign counterparts. [4,5]

. Since retroperitoneal smooth muscle tumors are more often malignant than benign, prompt and accurate preoperative radiological assessment is of paramount importance.

Ultrasonography examination provides good localization for retroperitoneal masses, though CT and especially magnetic resonance imaging (MRI) are most useful screening tools in evaluating and distinguishing the exact nature of the tumor and its relationship with adjacent organs and vascular structures. However, no radiological diagnostic modality appears highly sensitive or specific in ruling out malignancy and differential diagnosis on the basis of radiological finding alone is difficult.[2] Therefore, a definitive diagnosis requires histopathological examination of the tumor. Sampling of the retroperitoneal mass under USG or CT guidance preoperatively may allow microscopic examination, although the results may lack certainty due to small amount of specimen. Hence, the final determination of the nature of tumor is to be accomplished with a complete examination of resected specimen.

Our patient presented with a rapidly growing retroperitoneal mass which was clinically suspected to be a malignant tumor of unknown nature. Biopsy was not possible because of the poorly accessible location. In toto resection of the tumor was done followed by histopathological examination and immunohistochemical examination which rendered the diagnosis of retroperitoneal leiomyoma.

Retroperitoneal location of leiomyomas is of extremely rare occurrence. Most of the published case reports diagnosed the cases clinically as retroperitoneal growths with high suspicion of malignancy without suspecting their leiomyomatous nature.[3] Pathologic origin of these lesions are not certain. Poliquin and coworkers observed a 40% association of retroperitoneal leiomyomas with uterine

counterparts or a history of hysterectomy due to uterine leiomyomata.[6] Zaitoon suggested the parasitic theory for such tumor growth while Stutterecker et al claimed that Müllerian cell rests or smooth muscle cells in the retroperitoneal vessels wall are the putative origin.[3] Kho and Nezhat proposed an 'iatrogenic' origin for such growths while analyzing a case series of extra-uterine leiomyomata, mostly of retroperitoneal or intraperitoneal location with no visible connection to the uterus.[7]

Histologically, the distinction of benign leiomyoma and malignant leiomyosarcoma (especially low grade) may also be difficult. The histopathological parameters used for differential diagnosis include gross tumor size, the presence of nuclear atypia, pleomorphism and necrosis and the mitotic activity as the most useful guide to prognosis. On light microscopy, leiomyoma consists of monomorphic spindle cells arranged in interweaving fascicles which are separated by variable amounts of hyalinized collagen. Smooth muscle cells are elongated with eosinophilic cytoplasm and uniform, cigar-shaped nuclei. Usually, there is no cytologic atypia or necrosis and mitotic index is less than 5 per 10 high-power fields. It is very important to distinguish coagulative from hyalinizing necrosis as the presence of coagulative necrosis, even in absence of significant atypia would lead to a diagnosis of sarcoma.[8] In addition, immunohistochemical staining with estrogen, progesterone receptors, desmin, calponin, h-caldesmon, Ki-67 and p53 may be helpful in differential diagnosis of leiomyoma from leiomyosarcoma.

A complete surgical excision is the only curative treatment for retroperitoneal smooth muscle tumors, regardless their benign or malignant nature. A minimum of 3cm margin is essential but is rarely feasible due to invasion of adjacent structures by the tumor. The feasibility of complete resection is one of the important factors influencing the survival of patients. Curative surgery is difficult with large retroperitoneal sarcomas and those in close proximity to vital structures and involving adjacent organs. Aggressive surgery remains mandatory in retroperitoneal leiomyosarcoma. Abdominal hysterectomy along with the resection depends on the age of the patient, her symptomatology, and associated uterine myoma. The overall prognosis of patients with retroperitoneal leiomyoma is good with a small potential for local recurrence.[9]

In case of leiomyosarcoma, the addition of adjuvant radiotherapy after surgical resection is associated with a reduced risk of local recurrence, and a longer recurrence-free

interval, but there has generally been no suggestion of an impact on survival. With the exception of childhood soft tissue sarcomas such as rhabdomyosarcoma and extraskeletal Ewing's sarcoma, the benefits of adjuvant chemotherapy for soft tissue sarcoma remain controversial. The use of adjuvant chemotherapy following surgical resection for retroperitoneal leiomyosarcoma is not a standard approach.[(10)]

Conclusion

Leiomyomata have been described in several unusual locations, their occurrence in retroperitoneum is however extremely rare. Retroperitoneal sarcomas are more prevalent than their benign counterparts and till date, no radiological diagnostic modality appears highly sensitive or specific in ruling out malignancy. Poorly accessible location and rarity often makes retroperitoneal tumors a clinical puzzle. Leiomyoma should always be considered in the differential diagnosis of tumors at this site.

Conflict of Interest Statement-

There is no conflict of interest.

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