CASE REPORT

SYMPLASTIC LEIOMYOMA OF INFERIOR VENACAVA: PRESENTING AS A RETROPERITONEAL MASS

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ABSTRACT: Atypical/symplastic leiomyoma is infrequently encountered in surgical pathology. It is characterized by presence of multinucleated giant cells with pleomorphic nuclei and little or no mitotic activity we present a case of symplastic leiomyoma of inferior venacava, which is a rare variant of leiomyoma, and which presented as a retropitoneal mass in 55 year old women.
KEYWORDS: atypical leiomyoma, retroperitoneal mass, inferior venacava.

INTRODUCTION: Leiomyomas, which were first described by Virchow in 1854 are benign soft tissue tumors arising from smooth muscle and may develop in any location where smooth muscle is present. Frequently encountered in the uterine smooth muscle, where they are called "Fibroids", these tumours also arise from vascular walls more frequently seen in peripheral blood vessels compared to the central vessels and have been identified in the vena cava in only a few cases. (1) Leiomyoma which contain cells with moderate to severe atypia but without cell necrosis and mitotic index of fewer than 10 mf/10 hpf. Are called as Atypical or Symplastic Leiomyoma.[1] "Leiomyoma containing giant cells with pleomorphic nuclei and little or no mitotic activity" is called the Symplastic Leiomyoma[2] as per World Health Organization [WHO] classification. It is composed of tumor cells with variation in size and shape, hyperchromatic nuclei and multinucleated forms but no coagulative necrosis or mitotic activity counts of 2-7 mf/ 10 hpf (by the highest count). These are benign lesions even though there is high cellularity, numerous widely distributed bizarre cells and with no or low mitotic activity.

CASE PRESENTATION: Patient aged 55 years came with complaints of pain in right lumbar & iliac region of one month duration. Pain was colicky in nature. On clinical examination a 10×8 cm, immobile mass was palpated in the right hypochondrium and right lumbar region, firm in consistency. Ultrasonography revealed a retroperitoneal space occupying lesion. Computed tomogrphy revealed a well-defined mass lesion compressing the inferior venacava (Fig. 1). Retroperitoneal biopsy was done and showed a spindle cell tumor. Excision of retroperitoneal mass (Fig. 2) with inferior venacava resection & Dacron grafting was performed. (Fig. 3)

GROSS PATHOLOGY: A large irregular grey white tissue mass measuring 12.5×12×8 cm. External surface was bosselated, with a linear groove measuring 6cm in length. Cut section showed grey white whorled appearance. (Fig. 4)

MICROSCOPY: Histological findings revealed a tumor composed of thick bundles/fascicles of spindle cells, the fascicles were seen to intersect with each other at right angles (Fig. 5). The cells

are spindled and possess elongated nuclei with bipolar cytoplasmic process, several cells show enlarged nuclei with irregular contours, rare mitosis and mild lymphocytic infiltrates (Fig 6). A diagnosis of atypical/Symplastic leiomyoma was given.

IMMUNO-HISTOCHEMISTRY: Immunohistochemistry revealed SMA (Smooth Muscle Actin) positivity. (Fig. 7)

Diagnosis of atypical leiomyoma arising from inferior vena cava was thus confirmed by histomorphological features, immunohistochemistry and radiological investigations.

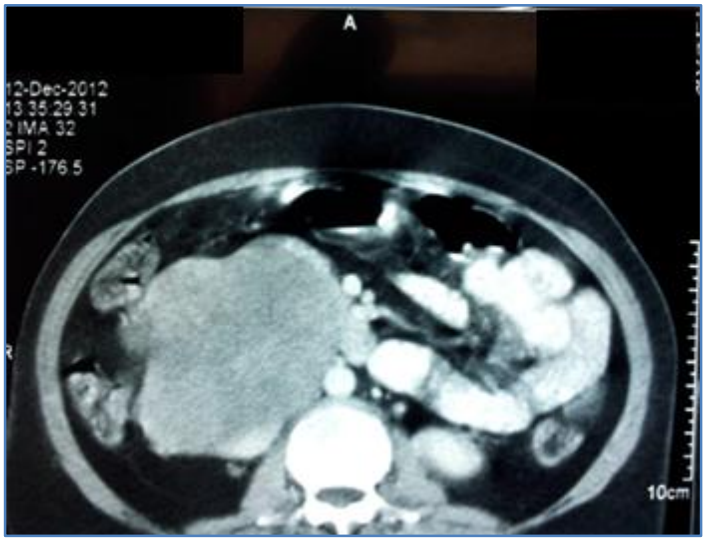

Fig. 1: Computer Tomographic picture, revealing compression on the venacava

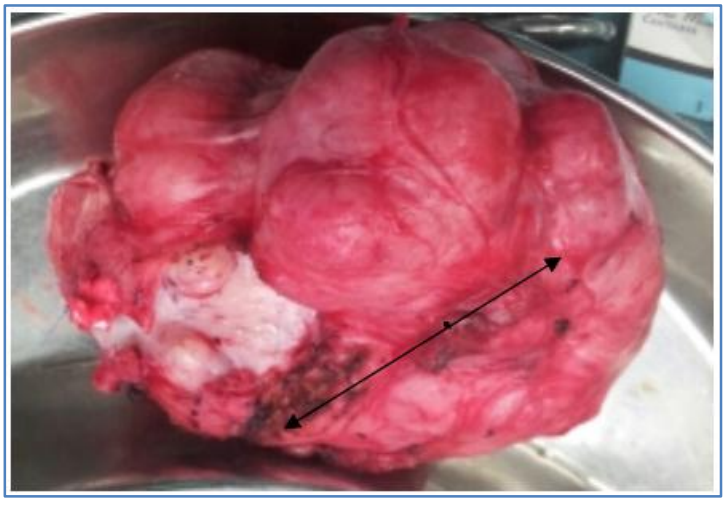

Fig. 2: Excised retroperitoneal mass nodular, with IVC groove (arrow)

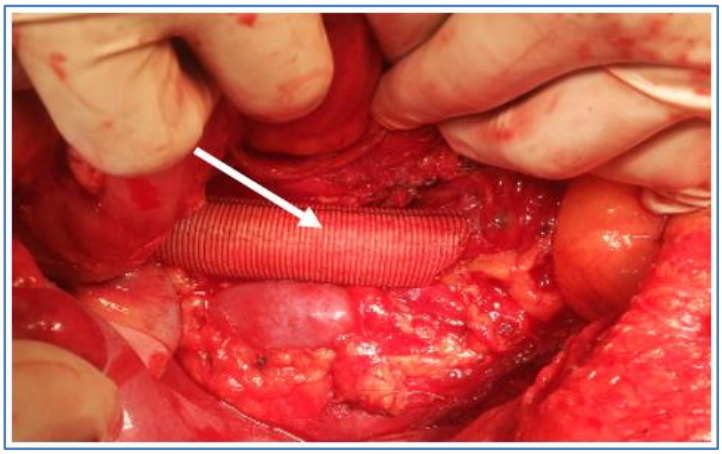

Fig. 3: Resected IVC with Dacron graft

Fig. 4: Gross picture of the retroperitoneal mass, bosselated surface, with the IVC groove (arrow) C/S whorled

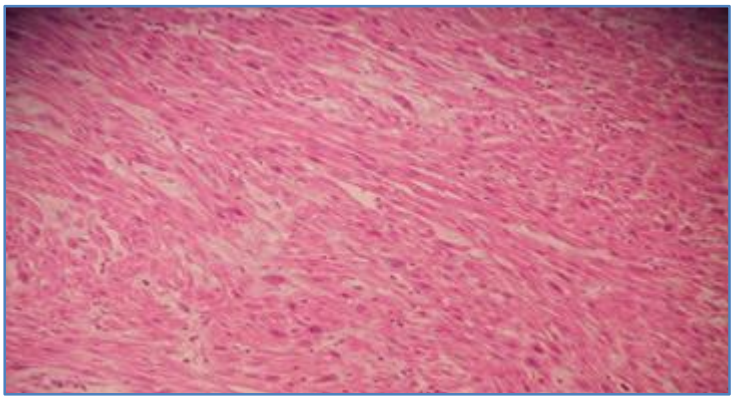

Fig. 5: Photo micro graph showing spindle cells intersecting at right angles (H & E stain, X 10x)

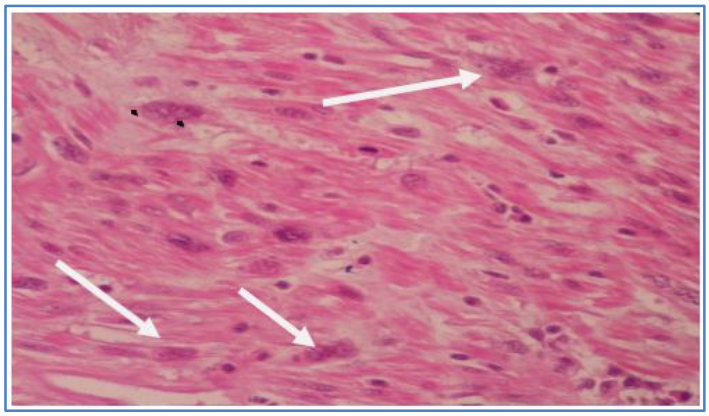

Fig. 6: Photo micro graph showing the bizarre nuclei, arrow (H & E, X 40x)

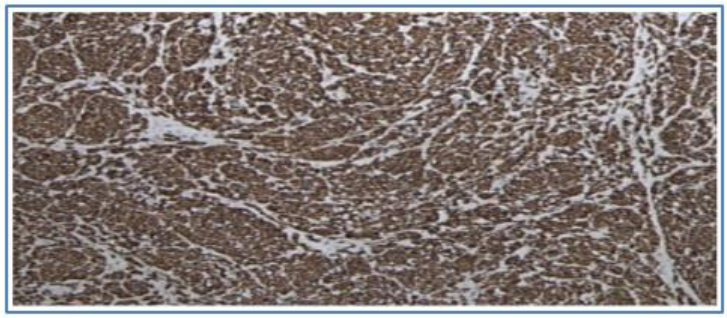

Fig. 7: Photomicrograph showing the diffuse Desmin positivity on IHC

DISCUSSION: Tumours can arise from wallof the vessels and usually present as soft tissue tumours. Most of them are venular in origin and can be endotheliomas from intima, fibrosarcoma from connective tissues, and leiomyosarcoma from smooth muscle of media.[3,4] Inferior vena cava is the commonest site of origin of venous tumors[5] followed by long saphenous vein, femoral vein, internal jugular vein, iliac vein and, very rarely, renal vein.

Most of the tumours arising from veins are malignant. Leiomyosarcomas are the most common primary malignancy of IVC that generate more than 90% of all reported IVC sarcomas, are usually thought to develop from the muscle of media. Leiomyomas also arise from the smooth muscles of media.[6,7,8]

Typical, epithelioid, cellular, hemorrhagic cellular, lipoleiomyoma and symplastic are the variants of leiomyoma.[9] Described histologically Martin and colleagues in 1960 used the word" Bizzare" for gastric Leiomyoma. Then, it was adopted as "bizarre leiomyoma" by the World Health Organization (WHO) and described as "leiomyoma containing giant cells without mitotic activity or with small and pleomorphic nuclei". The synonyms of this term have been reported as "atypical", "pleomorphic" and "symplastic".[10] The symplastic leiomyoma with a mitotic index lower than 10

mf/10 is a smooth muscle tumor which is rarely reported in the medical literature.[11,12] Only one case of IVC involvement has been reported.[13]

The various growth patterns of the lieomyoma variants mimic that of an aggressive tumou Pleomorphic tumor cells with distinct atypical nuclei and low mitotic rate are important characteristics Smplastic leiomyoma. The pleomorphic cells often have multinucleated or multilobated nucleus, can be present focally, multifocally or diffusely throughout the mass, and generate more than 25% of the tumor in most cases. These tumors can have secondary changes like, edema and hyaline changes, Coagulative tumor cell necrosis is never a feature of Symplastic Leiomyoma.[9] It may be misdiagnosed as leiomyosarcoma due to the marked nuclear atypia. The mitotic count is mainly considered in distinguishing a Symplastic leiomyoma from leiomyosarcoma. The mitotic index of leiomyosarcoma is equal to or more than 10 mf/10 hpf. It has been reported that symplastic leiomyoma with high cellularity has a benign course even if the mitotic activity of atypical cells is 2-7mf/10hpf.[9,11]

In this study, we attempted to present a rare histological variant of leiomyoma, the" Symplastic leiomyoma", with its rarer occurrence in inferior venacava.[13] Due to the presence of large atypical bizarre nuclei, one can be tempted to call the tumour as malignanat i.e. leiomyosarcoma,[13] which should be avoided by carefull search for absence of necrosis and a low/absent mitosis.

This case is presented for its rarity of type as well as origin/location of the tumour.

ABBREVIATIONS:

IVC-Inferior venacava.
MF: Mitotic figures.
HPF: High power field.

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