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Incidentally Discovered Giant Leiomyosarcoma of Renal Vein: A Case Report

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Abstract **Case Report**

***Background*:** Leiomyosarcoma originating from the renal vein (RVLMS) is extremely rare. Only few cases are reported in the literature. ***Case Report*:** We report here an exceptional case of a huge LMS of the right renal vein incidentally discovered, occurring in a 73-year-old female patient. ***Conclusions*:** This tumor lacks specific clinical manifestations and specific imaging features. Only pathological features make their diagnosis. Standard treatment consists of radical nephrectomy followed by chemotherapy and/or radiotherapy. The prognosis factors are not well identified. Overall prognosis of renal vein LMS is poor.

Keywords: Leiomyosarcoma, Retroperitoneal Tumor, Pathology, Incidentally Discovered.

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BACKGROUND

Angiogenic leiomyosarcoma is a rare soft tissue sarcoma, which mostly occurs in the inferior vena cava. A leiomyosarcoma arising from the renal vein is even rarer. To date, only 67 cases are reported in literature [1].

Similar to leiomyosarcoma arising from the inferior vena cava, renal vein leiomyosarcoma (RVLMS) lacks specific clinical manifestations. Most patients are asymptomatic, and the tumor is discovered incidentally [2].

CASE REPORT

We present the case of a 73 years old female patient with a medical history of a sigmoid adenocarcinoma operated in 2009. During a follow up consultation, the clinical examination found a large fixed mass in the right flank. Non abnormalities were observed in routine laboratory examination, except a ferriprive anemia: hemoglobin 11 mg/dL (12-14 mg/dL). A computed tomographic scan imaging demonstrated a 17 cm a left lateroaortic mass attached to the jejunal loop; a biopsy of the mass was performed and sent to the laboratory of pathology.

RESULTS

Histological examination of the biopsy was in favor of a smooth muscle tumor. There was insufficient material to distinguish benign from malignant. The mass excision was performed and appeared to have an attachment to the renal vein.

Gross examination (IMAGE 1) of the specimen found a fasciculate greyish-white mass measuring 17 x 15 cm, attached to a vascular structure and microscopic examination showed a spindle cell proliferation with moderate eosinophilic cytoplasm with elongated blunt ending vesicular nuclei showing nuclear pleomorphism in areas. Some cells show multinucleation. Mitotic figures were 9/10 hpf Immunohistochemistry was performed and showed a positif staning of Desmin, SMA and H- Caldesmon and the diagnosis of leiomyosarcoma was retained.

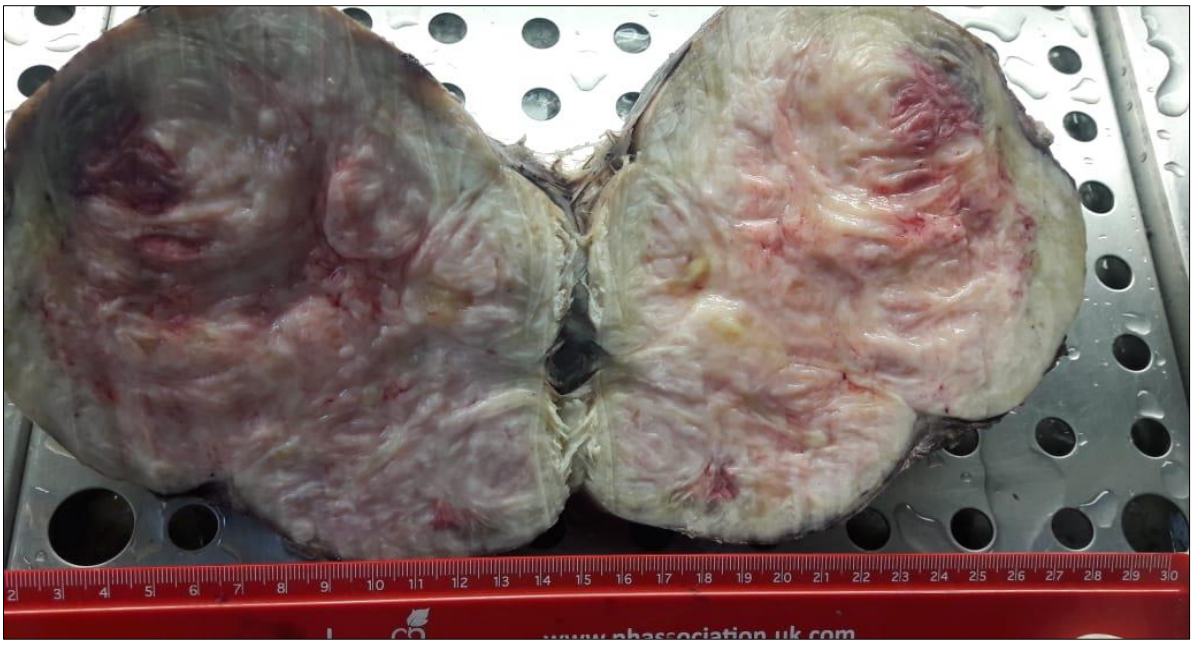

Image 1: Gross image of the tumor

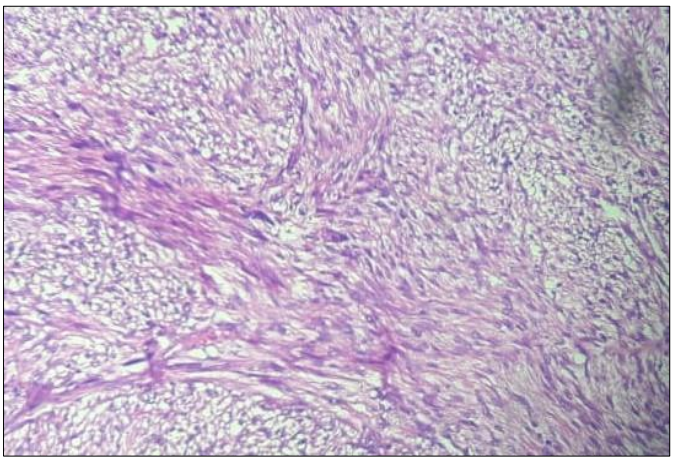

Image 2 :HE staining

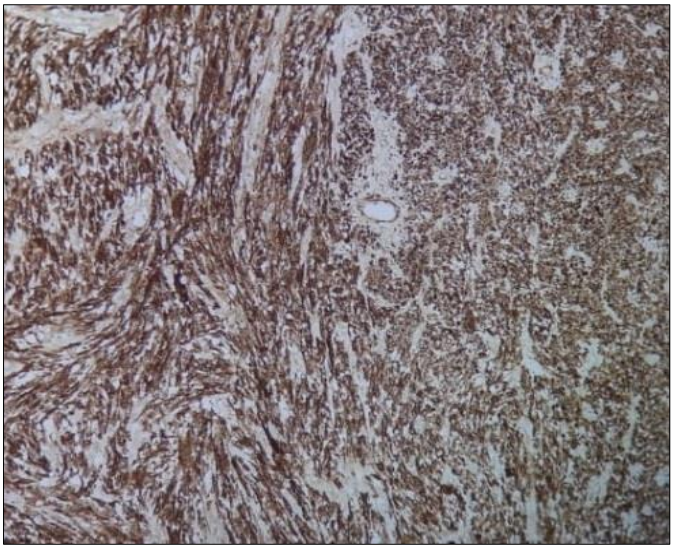

Image 3: SMA staining

Image 4: Desmin staining

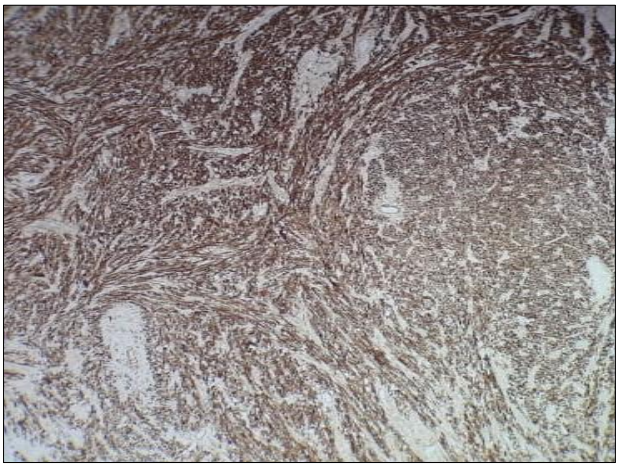

Image 5: H Caldesmone staining

DISCUSSION

LMS of vascular origin are an uncommon group of tumors [3]. The most common site is IVC (70% of all vascular LMS) [4]. Primary renal vein LMS are exceptional. Their clinical and imaging features can significantly overlap with those of advanced primary renal neoplasms, particularly renal cell carcinoma with venous extension [5]. In the 4th edition of the WHO Classification of Tumors of the Urinary System and Male Genital Organs, LMS of the renal vein is considered an entity of renal LMS [6].

A few cases of LMS of the renal vein have been reported so far [4-8]. It predominantly occurs in women (76.1%), on the left side (68.7%) and affects older population, with the peak occur-ring at age 60–69 years. Various theories have been suggested regarding this clinical presentation. Female preponderance is supported by the theory that estrogenic stimulation leads to growth and proliferation of smooth muscle tumors [9]. The more frequent involvement of the left renal vein is suggested by its longer length compared to the right renal vein [9-11]. In our case, the tumor involved rather the right renal vein.

From the clinical point of view, RVLMS presents difficulties in making diagnosis, because it is uncommon, has no specific symptoms and no pathognomonic radiological features. Presenting symptoms are unspecific, abdominal pain was reported in 49.3%. Imaging studies of LMS (magnetic resonance imaging (MRI) and contrast-enhanced CT) are nonspecific but helpful in delineating the relationship to adjacent structures, particularly in the retro peritoneum [12]. The location of LRV is more important than the size of the tumor. It can overlap with much more common RCC with venous extension. LRV is usually located in the hilum of the kidney. The bulk of the tumor lies predominantly or entirely outside the hilar parenchyma or the tumor is limited to the renal vessels [13].

With regard to biopsy, retroperitoneal mass is usually detected on abdominal CT scans. When imaging is not diagnostic of a retroperitoneal liposarcoma, image-guided CNB of to obtain the sample for diagnosis. However, sometimes, pre-surgical differentiation

between leiomyoma and leiomyosarcoma is very difficult and the final description of tumor characteristics usually requires excision and histological examination.

Grossly, LMS are large, solid, gray-white, soft to firm, and variably necrotic [5]. They usually generate the displacement of structures rather than invasion [12]. Microscopically, the morphological features of renal vein LMS are identical to those of LMS arising at other sites Typical LMS shows spindle-shaped cells with plump, blunt-ended nuclei and moderate to abundant, pale to brightly eosinophilic fibrillary cytoplasm. The cells are set in long intersecting fascicles parallel and perpendicular to the plane of section. Moderate nuclear pleomorphism is usually noted, although pleomorphism may be focal, mild, or occasionally absent. Mitotic figures, including atypical ones, are typically easy to find. Tumor cell necrosis is often present in larger tumors [12]. Immunohistochemically, at least one myogenic marker (SMA, desmin, or h-caldesmon) is positive in 100% of cases, and >70% of cases are positive for more than one of these markers. None of these is absolutely specific for smooth muscle, and positivity for two myogenic markers is more supportive [12].

With regard to treatment, the only potentially curative treatment for RPS is surgery with macroscopically complete resection [9-11]. The role of ChT and RT in RPS is not proven and still under investigation. It is generally recommended, that in case of RT administration, it should be delivered in the preoperative setting and possibly within a clinical trial [9]. Postoperative RT should not be ad-ministered routinely in R0 and R1 resections [9].

According to the current literature, the risk of local recurrence and distant metastasis is significantly increased when the tumor is greater than 3 cm in diameter, and when the margins of the resected specimen are positive for tumor cells [14, 15]. Grignon *et al*., reported that the probability of local recurrence after operation is 40%, and that distant metastases can reach the lung, liver, skin and soft tissues [16]. Aguilar *et al*., analyzed 30 cases reported in the literature and found that 30% of patients (average follow-up 78 months) had no local recurrence or distant metastases, 23% had local recurrence and distant metastases but were still alive (average follow-up 48 months), and 37% died following local recurrence [14]. Brandes *et al*., [17], reported that RVLMS exhibits a poor 5-year survival rate. In our case, the patient is alive and without local recurrence or distant metastases 27 months after surgery. Adjuvant therapy and regular follow-up both play important roles in the treatment of RVLMS.

CONCLUSION

Renal vein leiomyosarcoma is a very rare tumor, that lacks specific clinical manifestations and specific imaging features. Most patients are diagnosed intra-operatively or following postoperative pathology. Early and complete resection is considered the first choice of treatment, and whether or not to preserve the kidney depends on the patient's condition.

List of Abbreviations

LMS: Leiomyosarcoma.
IVC: Inferior Vein Cava.
RCC: Renal cell carcinoma.
RPS: retroperitoneal sarcoma.
RVLMS: Renal vein Leiomyosarcoma.
CNB: Core needle biopsy.

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