Local recurrence and multi-organ metastasis of primary retroperitoneal leiomyosarcoma in unusual locations after surgical resection

Hira Lal[1], Zafar Neyaz[1*], Vinay Kumar Kapoor[2], Biju Pottakkat[2], Pallav Gupta[3]

1. Department of Radiodiagnosis, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, India

2. Department of Surgical Gastroenterology, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, India

3. Department of Pathology, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, India

* **Correspondence:** Dr. Zafar Neyaz, MD, Assistant Professor, Department of Radiodiagnosis, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Rae Bareilly Road, Lucknow, Uttar Pradesh, India. 226014
(*zafarneyaz@hotmail.com*)

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ABSTRACT

We report a case of retroperitoneal leiomyosarcoma in a 52-year-old woman, who developed local recurrence, pulmonary, hepatic, peritoneal, pancreatic, subcutaneous and intramuscular metastases two years after surgical resection of the primary tumor. Metastasis to the pancreas, subcutaneous or muscular tissue is very rare; however, presence of a lesion in these locations in a known case of leiomyosarcoma might indicate a metastatic tumor deposit.

CASE REPORT

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A 52-year-old female underwent surgical resection for a retroperitoneal soft tissue mass 2 years prior to presentation. At laprotomy, a large lobulated soft tissue mass was found arising from the left retroperitoneal area. The tumor was involving the left perirenal space and was infiltrating into the sigmoid mesocolon. An extracapsular excision was done. The postoperative course was uneventful. Gross pathological examination revealed a circumscribed mass measuring 10x14x18 cm. Outer surface of mass was bosselated and congested at places. The cut surface was firm having variegated appearance with solid grayish white areas, foci of hemorrhage, necrosis, mucoid and cystic degeneration. The cysts ranged from 0.3 to 0.5 cm. Microscopic examination demonstrated structure of a tumor consisting of sheets and fascicles of spindle cells having oval to oblong pleomorphic nuclei and moderate amount of eosinophilic cytoplasm (Fig. 1). Foci of necrosis, dilated and congested blood vessels, and frequent mitosis were seen. On immunohistochemistry, the tumor cells showed positivity for desmin and smooth muscle actin (Fig. 2, 3), and were negative for c-kit and S-100. Based on above findings a diagnosis of retroperitoneal leiomyosarcoma was made. Patient was not offered any chemotherapy or radiotherapy in view of the poor response to these therapeutic options.

The patient was lost follow up for two years. Then she reported to the outpatient clinic with a subcutaneous lump in the left paraumbilical location which was gradually increasing in size. There was no symptom. She was readmitted and a chest x-ray showed multiple nodular lesions in both lungs (Fig. 4). Ultrasound of abdomen revealed multiple hypoechoic liver lesions, a lesion in the head of pancreas, a subcutaneous mass lesion in left paraumbilical region and a large mass in retroperitoneum displacing the left kidney (Fig. 5-7). Color Doppler showed no significant vascularity inside the retroperitoneal lesion (Fig. 8). A contrast enhanced CT scan of chest and abdomen was done which confirmed the above findings. Additionally it demonstrated a peritoneal deposit near the splenic flexure, multiple subcutaneous deposits, and an intramuscular lesion in the left gluteus maximus muscle (Fig. 9-12). A core biopsy was performed from the subcutaneous nodule in the left paraumbilical location and histopathology revealed a metastatic deposit from leiomyosarcoma. Revision surgery was not considered because of the disseminated disease.

DISCUSSION

Leiomyosarcoma is the second most common primary retroperitoneal tumor in adults after the liposarcoma [1]. It is an uncommon malignant neoplasm of smooth muscle origin that tends to arise in the retroperitoneum, peripheral soft tissues, genitourinary tract, gastrointestinal tract, large vessels and rarely in bones [2]. Recent literature suggests that majority of the previously labeled alimentary tract leiomyosarcomas were actually gastrointestinal stromal tumors, with different immunohistochemistry. The etiology of leiomyosarcoma is not clear; however, in some cases prior irradiation and Epstein-Barr virus have been implicated [3, 4]. About 20-67% of cases of leiomyosarcoma arise in the retroperitoneum [2]. Retroperitoneal leiomyosarcomas are more common in females. Although no age group is exempted, it usually affects middle-aged to older adults [2, 5].

CT is the primary imaging modality for the assessment of retroperitoneal leiomyosarcomas as well as for the evaluation of metastatic disease [2]. The imaging appearance of leiomyosarcoma is non-specific and histopathology with immunohistochemistry is essential for the correct diagnosis. Most retroperitoneal leiomyosarcomas grow silently and by the time of diagnosis the primary tumor masses are usually quite large in size with extensive necrotic or cystic change [2, 5]. Calcification is rare in these tumors. Sometimes, retroperitoneal leiomyosarcomas may extend into the retroperitoneal veins and inferior vena cava (IVC). Rarely, leiomyosarcomas may arise from the IVC itself, with predominantly intraluminal growth [1]. Moderate contrast enhancement is seen at the periphery of the large primary and metastatic lesions [2]. As the metastases increase in size, they often demonstrate cystic necrosis.

MRI is also utilized in the evaluation of primary retroperitoneal leiomyosarcoma due to its multiplanar capabilities and superior soft tissue contrast resolution [1, 2]. MRI can better show the tumor margins, the site of origin and involvement of adjacent structures. Additionally intravenous Gadolinium may be used to asses the tumor vascularity and vessel involvement. Like CT scan, leiomyosarcomas have a non-specific appearance on MRI. On MRI the tumor is usually well defined, isointense to muscle on T1 weighted images, intermediate to hypointense to fat on spin-echo T2 weighted images and predominantly hyperintense on spin-echo T2 images with fat saturation [2]. The signal intensity of the tumor also varies depending on the amount of cystic spaces and hemorrhagic areas.

Complete surgical resection with wide margins reduces the rate of local recurrence; however, in retroperitoneum it is difficult to procure wide margins all the way around tumor due to the major vessels and other important structures [2]. Even when complete excision is believed to have been accomplished, local recurrence rates are as high as 40-77% [1]. Leiomyosarcomas have propensity for hematogenous spread and infrequently metastasize to lymph nodes. Distant metastases are present at the time of diagnosis in approximately 40% of cases and most patients who survive the primary tumor will eventually develop metastases [1]. Liver and lungs are the most common sites of metastasis in patients with leiomyosarcoma [2, 5]. Other manifestations of tumor spread include mesenteric or omental metastases, retroperitoneal lymphadenopathy, soft tissue metastases, bone metastases, splenic metastases, and ascites.

Rarely leiomyosarcomas have been reported to metastasize to pancreas, stomach, small bowel, cardiac chambers, submandibular salivary gland, skin, scalp, subcutaneous tissue, and skeletal muscles [3, 6-16]. Fewer than 20 cases of pancreatic metastases from the leiomyosarcoma have been reported in the literature [6-8]. Metastasis to the subcutaneous tissue and muscle is also very uncommon. In the series by McLeod et al. the incidence of soft tissue metastasis was about 7% [5]. One of their patients had metastasis in the adductor muscles from the leiomyosarcoma of the uterus. Subcutaneous tissue metastasis in the left flank has been described in a case of uterine leiomyosarcoma ten years following total abdominal hysterectomy [13]. Aslan et al. have reported temporalis muscle metastasis from a uterine leiomyosarcoma [14]. Two other cases of metastasis to the skeletal muscle have been reported from the uterine leiomyosarcomas [15-16]. In our case, in addition to the commonly reported metastatic sites, metastatic deposits were seen in rare sites like pancreas, subcutaneous tissue and the left gluteus muscle.

Among leiomyosarcomas of all sites, the retroperitoneal leiomyosarcomas have the worst prognosis and about 80%-87% of these patients die within 5 years [1]. Cure of the primary tumor is difficult because of late presentation, origin within deep tissue, inability to achieve wide surgical margins, and relative insensitivity to chemotherapy and radiotherapy [1, 2]. Single or few metastatic lesions have been surgically treated in various reports [8, 13]. Distant metastases including pancreas, subcutaneous tissue, and muscle lesions, as reported in our patient, are usually associated with widespread disease and carry an extremely poor prognosis. One other important feature of the metastatic deposits in leiomyosarcoma is the late occurrence as long as 10 years after resection of the primary lesion [13]. A long term follow up with CT scan at least once a year is important in these patients.

To conclude, the leiomyosarcomas are known for giving rise to metastatic deposits in unusual locations. In a known case of leiomyosarcoma a newly developed subcutaneous, pancreatic, or muscular lesion should be investigated and possibility of a metastatic deposit should be considered.

TEACHING POINT

In a known case of leiomyosarcomas long term follow up with imaging is essential to detect local recurrence as well as distant metastasis. As these tumors are known to produce metastatic deposits in rare locations, any newly developed lesion even in odd location should be investigated for the presence of metastasis.

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FIGURES

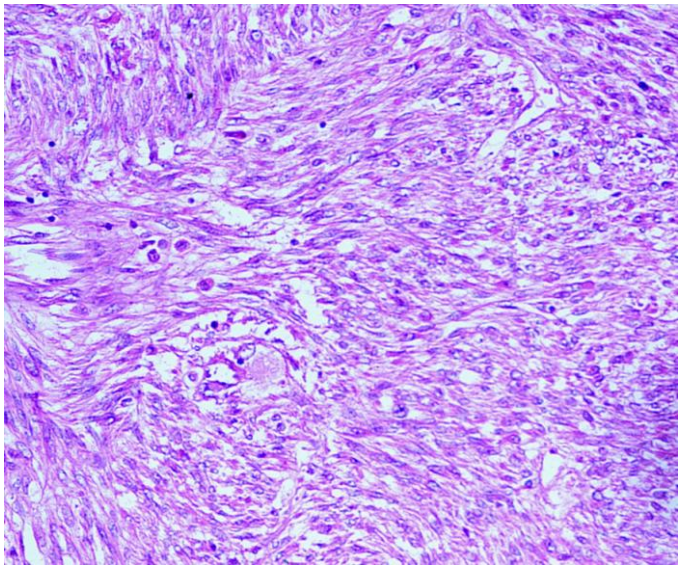

Figure 1: 52-year-old woman presenting with recurrent retroperitoneal leiomyosarcoma and multi-organ metastasis in unusual locations. Microscopic appearance of the primary retroperitoneal mass, showing tumor cells consisting of sheets and fascicles of spindle cells having oval to elongated hyperchromatic nuclei, abundant eosinophilic cytoplasm & frequent mitosis (H&E x200).

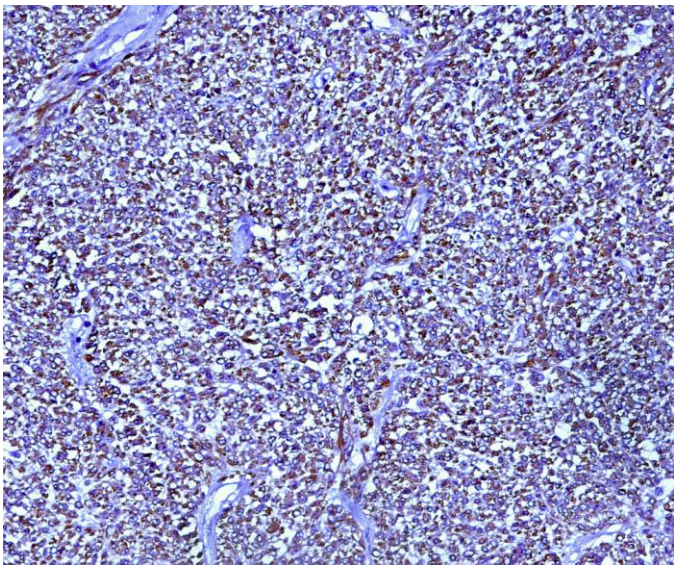

Figure 2: 52-year-old woman presenting with recurrent retroperitoneal leiomyosarcoma and multi-organ metastasis in unusual locations. Immunohistochemistry of the primary retroperitoneal mass. Tumor cells are showing cytoplasmic positivity for desmin (IHC x200).

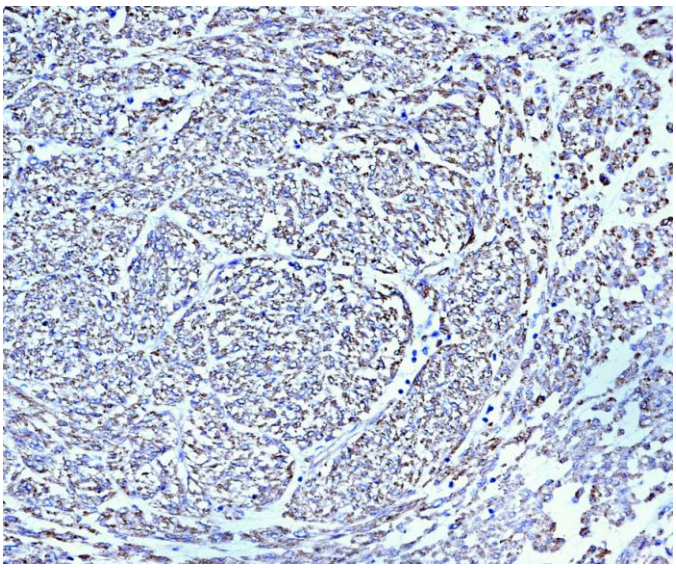

Figure 3: 52-year-old woman presenting with recurrent retroperitoneal leiomyosarcoma and multi-organ metastasis in unusual locations. Immunohistochemistry of the primary retroperitoneal mass. The tumor cells showing cytoplasmic positivity for smooth muscle actin (IHC x200).

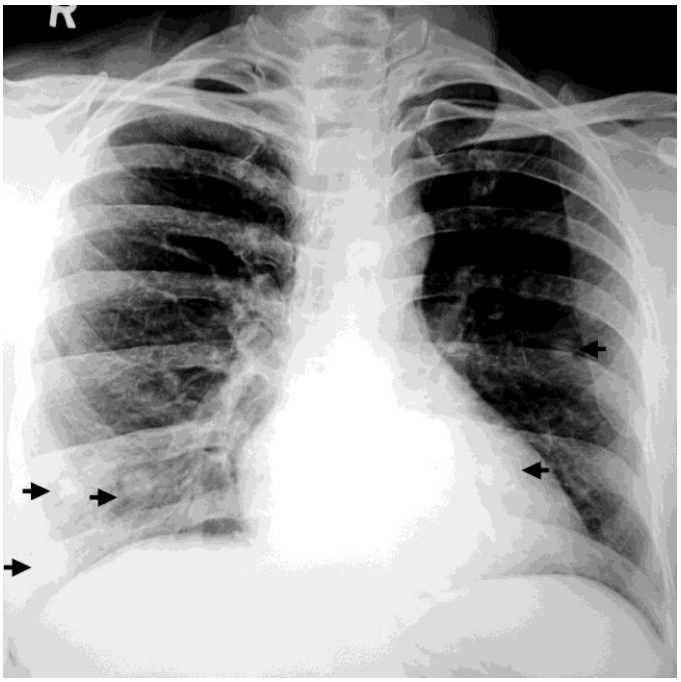

Figure 4: 52-year-old woman presenting with recurrent retroperitoneal leiomyosarcoma and multi-organ metastasis in unusual locations. Chest x-ray shows multiple well defined nodular lesions in both lungs (arrows). (Chest X-ray PA view, erect posture)

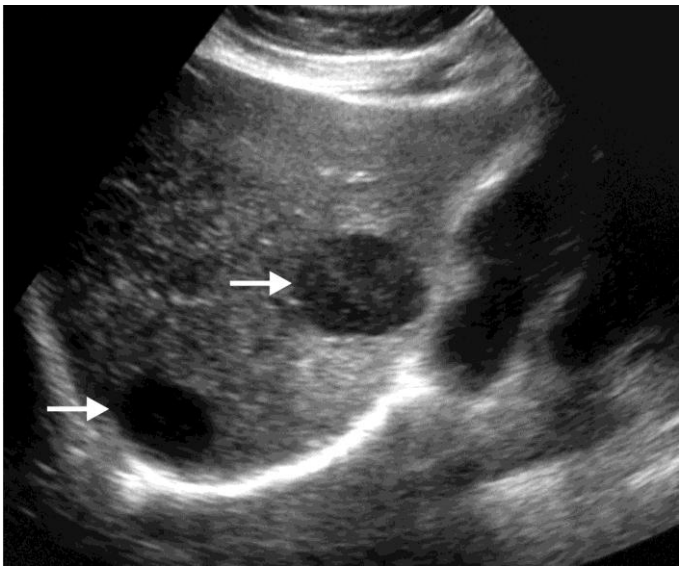

Figure 5: 52-year-old woman presenting with recurrent retroperitoneal leiomyosarcoma and multi-organ metastasis in unusual locations. Ultrasound image shows two hypoechoic liver lesions of 2.5 cm and 3.0 cm diameter (arrows). (Transabdominal ultrasound, HD 11 XE Phillips, C5-2 probe)

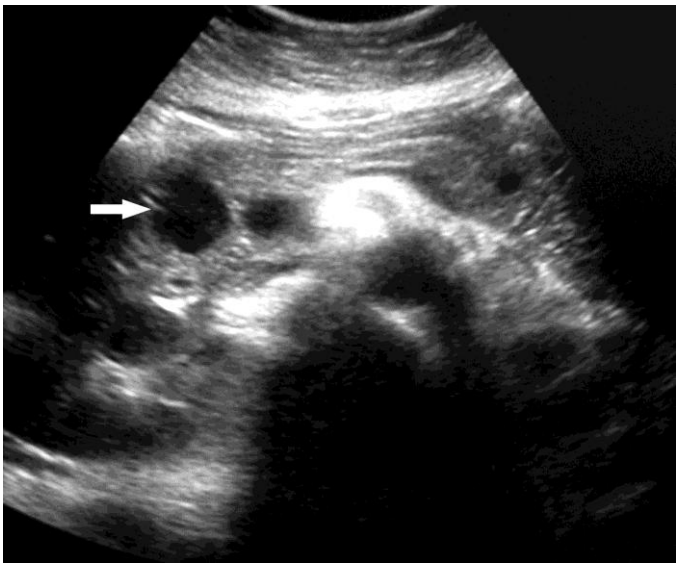

Figure 6: 52-year-old woman presenting with recurrent retroperitoneal leiomyosarcoma and multi-organ metastasis in unusual locations. Ultrasound image shows a rounded 1.5 cm diameter hypoechoic lesion in the head of pancreas (arrow). (Transabdominal ultrasound, HD 11 XE Phillips, C5-2 probe)

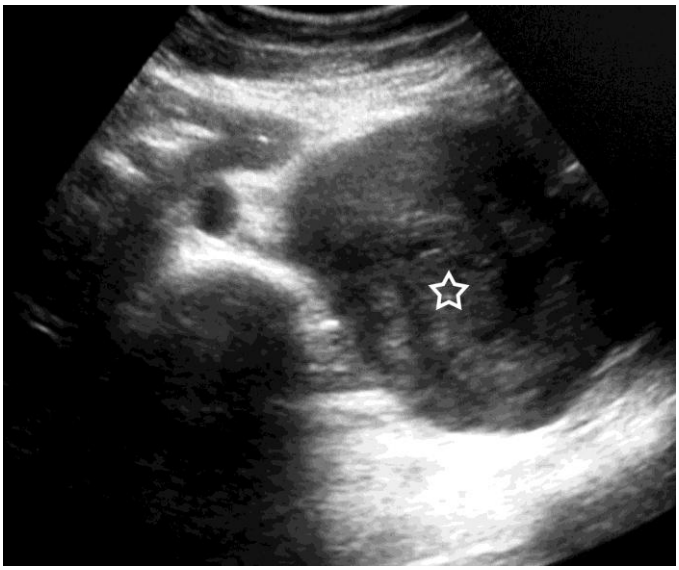

Figure 7: 52-year-old woman presenting with recurrent retroperitoneal leiomyosarcoma and multi-organ metastasis in unusual locations. Ultrasound image shows a large 6.8 x 7.8 x 7.9 cm recurrent retroperitoneal mass (star). (Transabdominal ultrasound, HD 11 XE Phillips, C5-2 probe)

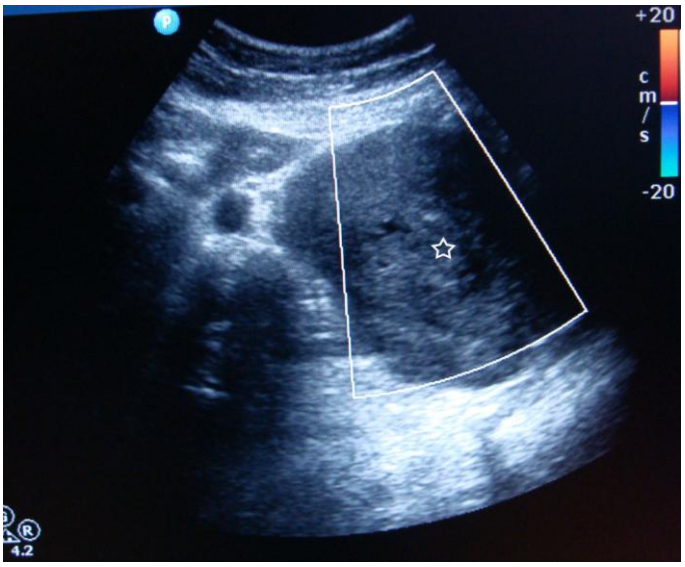

Figure 8: 52-year-old woman presenting with recurrent retroperitoneal leiomyosarcoma and multi-organ metastasis in unusual locations. Color Doppler image shows no significant vascularity in the retroperitoneal mass (star). (Transabdominal ultrasound, HD 11 XE Phillips, C5-2 probe)

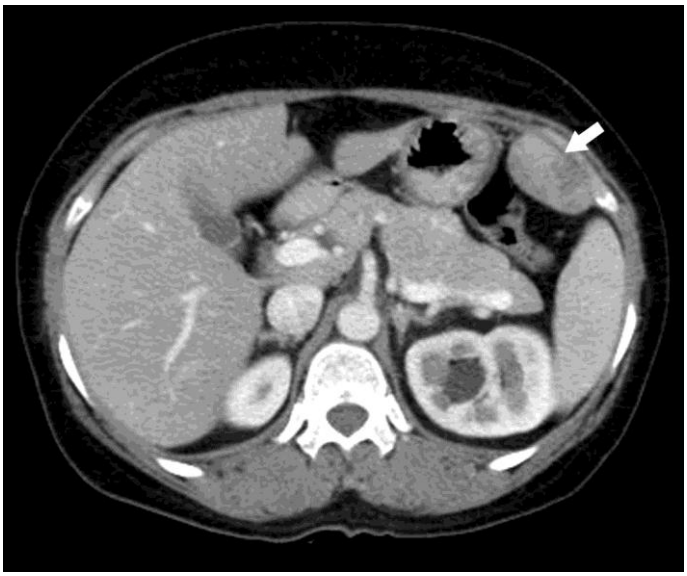

Figure 9: 52-year-old woman presenting with recurrent retroperitoneal leiomyosarcoma and multi-organ metastasis in unusual locations. Axial contrast enhanced CT images soft tissue window (64-Channel multidetector Brilliance CT, Philips Medical systems, 120 kV, 250 mAs, 3 mm reformation and window width/level of 360 / 60 HU, 100 c.c. of 300 mg/ml iodine concentration non-ionic contrast given by hand injection) at the level of pancreatic body shows a 2.0 x 3.4 x 4.0 cm peritoneal deposit near splenic flexure (arrow).

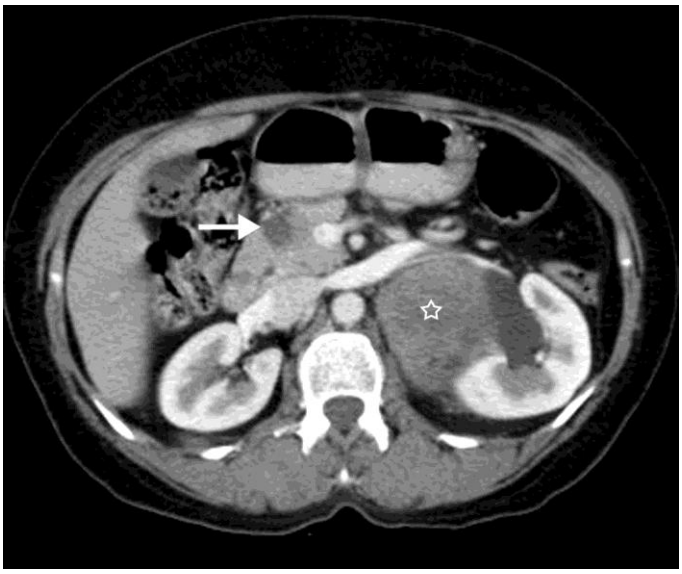

Figure 10: 52-year-old woman presenting with recurrent retroperitoneal leiomyosarcoma and multi-organ metastasis in unusual locations. Axial contrast enhanced CT images soft tissue window (64-Channel multidetector Brilliance CT, Philips Medical systems, 120 kV, 250 mAs, 3 mm reformation and window width/level of 360 / 60 HU, 100 c.c. of 300 mg/ml iodine concentration non-ionic contrast given by hand injection) at the level of renal hilum shows a large 6.8 x 7.8 x 7.9 cm recurrent mass in left perirenal location (star) and a 1.5 cm diameter pancreatic head metastasis (thin arrow).
Figure 11.

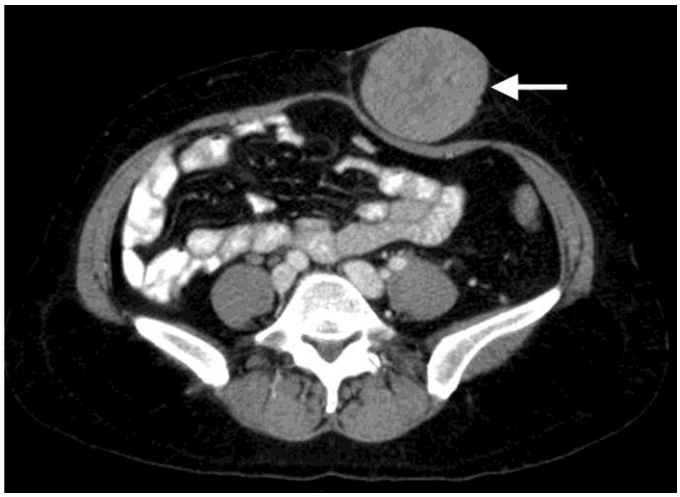

Figure 11: 52-year-old woman presenting with recurrent retroperitoneal leiomyosarcoma and multi-organ metastasis in unusual locations. Axial contrast enhanced CT images soft tissue window (64-Channel multidetector Brilliance CT, Philips Medical systems, 120 kV, 250 mAs, 3 mm reformation and window width/level of 360 / 60 HU, 100 c.c. of 300 mg/ml iodine concentration non-ionic contrast given by hand injection) at the level of umbilicus shows a large 5.0 cm diameter subcutaneous deposit in left paraumbilical region (arrow).

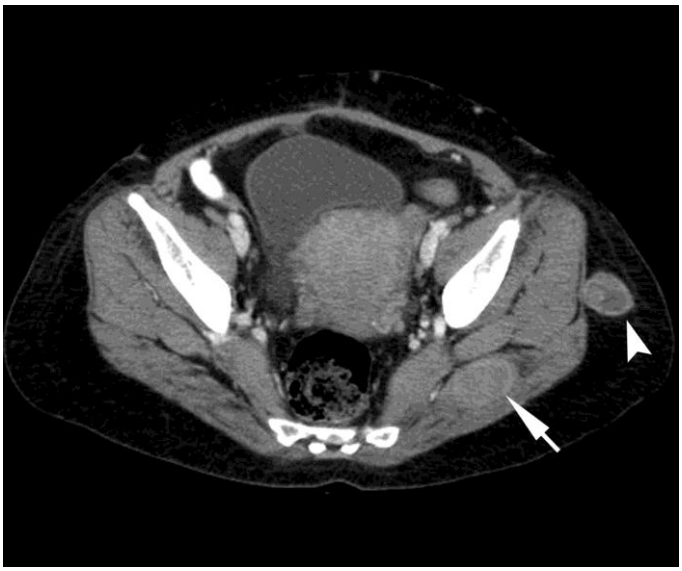

Figure 12: 52-year-old woman presenting with recurrent retroperitoneal leiomyosarcoma and multi-organ metastasis in unusual locations. Axial contrast enhanced CT images soft tissue window (64-Channel multidetector Brilliance CT, Philips Medical systems, 120 kV, 250 mAs, 3 mm reformation and window width/level of 360 / 60 HU, 100 c.c. of 300 mg/ml iodine concentration non-ionic contrast given by hand injection) in the pelvic region shows a 2.0 x 2.5 x 4.3 cm subcutaneous deposit in left gluteal region (arrowhead) and a 2.5 x 3.1 x 4.4 cm metastasis in left gluteus maximus muscle (arrow).

Differential diagnosis	USG	CT	MRI
Retroperitoneal leiomyosarcoma	• May appears solid but often contains cystic spaces with irregular walls. • The solid components are usually isoechoic relative to liver but can be hyperechoic. The cystic spaces usually contain low level echoes, but anechoic cysts are also seen.	• Often large and heterogeneous, central low density due to haemorrhage, necrosis or cystic change may be seen. • Moderate contrast enhancement at the periphery of the large primary & metastatic lesions, whereas smaller lesions may be homogeneous. Calcification is uncommon.	• Seen as a non-fatty mass, large lesions tend to be heterogeneous with areas of necrosis. Small lesions may be homogeneous. It is isointense to muscle on T1w and hyperintense on T2w images.
Gastrointestinal stromal tumor (GIST)	• Often large (more than 10 cm) at presentation, with heterogeneous enhancement and central necrosis. Hematogenous spread to the liver may produce cystic metastases. Usually seen in submucosal or intraluminal position in the stomach, jejunum and ileum; however, can occur in the mesentery, omentum and retroperitoneum.	• May show air or bowel contrast inside the tumor due to communication with bowel. • Calcification and lymph node metastasis is rare. • Small GISTs may show intense enhancement after contrast administration; however, variable enhancement seen in larger tumors. • No tendency for vascular invasion or venous thrombosis.	• The signal intensity varies depending on the degree of necrosis and hemorrhage. • The solid portions of tumor tend to be isointense relative to skeletal muscle on T1w images and hyperintense on T2w images, and enhance after administration of gadolinium. • Areas of hemorrhage within the tumor will show signal intensity, depending on the age of the hemorrhage.
Lymphoma	• May present as a hypoechoic mass lesion in retroperitoneum which is relatively hypovascular on color Doppler. Encasement of adjacent organs may be noted with associated lymphadenopathy. • Discrete Hypoechoic lesion may be noted on other organs like liver, spleen depending on stage.	• Size small to bulky and vary in shape from round or oval to irregular mass, may encase kidneys, pancreas and great vessels. • Usually seen as mildly enhancing, homogenous masses; however necrosis may be seen. The vessels remain patent despite tumor encasement.	• Lymphoma masses are low to iso-signal intensity on T1w images and moderately high signal on T2w imaging, less than the high signal associated with cysts. After the administration of gadolinium, usually mild enhancement is seen.

Table 1. Differential diagnosis of retroperitoneal leiomyosarcoma.

Etiology	Largely unknown, post radiation and Epstein-Barr virus implicated in some cases.
Incidence	Rare tumor, About 20–67% of leiomyosarcoma arise in Retroperitoneum.
Gender ratio	More common in women
Age predilection	Middle-aged to older adults
Risk factors	Unclear, post radiation and Epstein-Barr virus have been implicated.
Treatment	Surgical resection
Prognosis	Poor prognosis, about 80%-87% of patients die within 5 years.
Imaging findings	Imaging appearance is non-specific. Often large and heterogeneous, central low density due to haemorrhage, necrosis or cystic change. Moderate contrast enhancement at the periphery of the large primary & metastatic lesions, whereas smaller lesions may be homogeneous. Calcification is uncommon. Sometimes may extend into the retroperitoneal veins and inferior vena cava.

Table 2. Summary table of retroperitoneal leiomyosarcoma

ABBREVIATIONS

CT = Computed tomography
IVC = Inferior vena cava
IHC = Immunohistochemistry

KEYWORDS

Computed tomography; Leiomyosarcoma; Metastasis; Pancreas; Retroperitoneum; Skeletal muscle

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