The perinephric space and associated extra-renal pathology. Multi-modality imaging findings of normal anatomy and varying causes of perinephric masses.

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**Learning objectives**

To describe and illustrate the anatomy of the perirenal space

To discuss various pathological processes that can occur in the perirenal space such as infection, metastases, lymphoma and liposarcoma, as well as the rare conditions of capsular leiomyoma and perirenal malakoplakia

To describe and demonstrate the imaging findings of these various conditions on ultrasound, CT and MRI
Background

The perinephric (perirenal) space is a retroperitoneal area that contains the kidney, proximal ureter, adrenal gland, perirenal fat, lymphatics and blood vessels. Disease affecting this space may arise within or outside it. The space is involved in multiple neoplastic as well as non-neoplastic conditions. Many perinephric lesions have arisen from the kidney and adrenal gland. Others spread there via the blood or lymphatics. The imaging characteristics of multiple perinephric masses are described on multiple modalities (ultrasound, CT and MRI). We describe and illustrate the imaging characteristics of perinephric pathological processes including lymphoma, infection, metastases and liposarcoma on multiple modalities. We also show a rare case of perirenal malakoplakia.
Findings and procedure details

Anatomy

The perinephric space is a cone-shaped retroperitoneal compartment located between the anterior and posterior pararenal spaces (1, 2)(Fig. 1). It contains the kidney, proximal ureter, adrenal gland, fat and blood vessels. Gerota's fascia marks the anterior boundary and Zuckerkandl's fascia marks the posterior boundary.

A. Lymphoma

Perirenal lymphoma is usually clinically silent but may present with flank pain, haematuria, a palpable mass or even acute renal failure (3). It is most often due to the extension of retroperitoneal or renal lymphoma, but in 10%, it is found in the perirenal space alone (3). 3-8% of all patients with lymphoma will have renal/perirenal involvement (4). NHL (Non-Hodgkin's lymphoma) is the most common type.

**Imaging findings**

Perirenal lymphoma appears as a large contiguous retroperitoneal mass that forms a rind around the kidneys and is homogeneous attenuation on CT with mild enhancement (Fig. 2). Less extensive cases appear as thickening of Gerota's fascia or nodules in the perirenal space. On MRI, the mass is low T1/low-to-isointense T2, with again a degree of enhancement post gadolinium. There may be invasion of the kidney or a degree of renal compression (5). On ultrasound, hypoechoic tissue of variable thickness is seen to surround the kidney (6).

B. Infection

Perinephric infection is common (1). Abscesses may result from a urinary tract infection or from infection developing in a perinephric haematoma or urinoma (7). Risk factors include diabetes and immunosuppression. Infections may be bacterial or fungal and present with flank pain, pyrexia and elevated inflammatory markers.

**Imaging findings**

On CT, findings include thickening of Gerota's fascia, perinephric fat stranding and loss of the normal perinephric fat planes (8). Appearances can be focal or diffuse. Following administration of IV contrast, there is a heterogeneously enhancing mass with collections appearing as a cystic part containing enhancing septa (Figs. 3 & 4). Gas may be present.
The adjacent kidney can be enlarged. Ultrasound has been shown to be insensitive in showing extension of inflammation into the perinephric space, but is good at showing a focal abscess. They appear as multilocular fluid collections with internal echoes (1).

C. Metastases

Due to a rich blood and lymphatic supply, the perirenal space is a potential site for metastatic disease (4). However, isolated perirenal metastases are rare (8). Those neoplasms that do metastasize to the perirenal space include lung, malignant melanoma, breast and prostate carcinomas, and lung being the most common.

*Imaging findings*

They appear as single or multiple soft-tissue masses around the kidney. Breast metastases can be more infiltrative (1). Metastases to the kidneys may also infiltrate into the perinephric space by directly extending through lymphatic’s or via capsular vascular flow (4). Arterial enhancement occurs with malignant melanoma and breast metastases, but not with lung or prostate (Figs. 5, 6 & 7).

D. Liposarcoma

Liposarcomas are malignancies of adipose tissue that correspond to 19-35% of all soft tissue sarcomas (10, 11). It is the most common malignant retroperitoneal tumour (1, 9) with 56% the well-differentiated type and 37% the de-differentiated type. They peak in the 6\(^{th}\) and 7\(^{th}\) decade and tend to be large at the time of presentation.

*Imaging findings*

The well-differentiated type has a large macroscopic fat component, appearing as fat attenuation on CT and are usually round or lobulated, displacing normal structures (Figs. 8 & 9). On MRI they are predominately high signal on T1 and isointense on T2. They become dark on fat saturation sequences. They may contain non-fatty areas that are low signal on T1 and iso-to-hyperintense on T2. These non-fatty areas enhance post contrast (12). The more high-grade types have similar signal and attenuation to muscle on T1 MRI and CT respectively. On T2 sequences, they show intermediate-to-high signal. Some types have a cystic component with reticular enhancement. Non-fatty areas are hypointense on T1 and iso-to-hyperintense on T2. Solid areas show enhancement.

E. Capsular Leiomyoma
Renal leiomyomas are rare and benign (9). They are most often asymptomatic and may present with an abdominal mass, pain and haematuria (13). Those that occur in the perirenal space are exophytic renal tumours arising from the renal capsule.

**Imaging findings**

On imaging they are well-defined soft tissue enhancing masses (Figs. 10, 11 & 12). On ultrasound they appear as well-defined, low echogenic masses with low doppler signal (14). CT and MRI show a soft tissue density/signal mass (14). Capsular leiomyomas show an arterial supply from the capsular artery on angiography. There is an absence of adjacent renal parenchymal distortion or lymphadenopathy. Those that are larger in size are heterogeneous in appearance secondary to haemorrhage and cystic degeneration.

**F. Extra-adrenal Paraganglioma**

An extra-adrenal paraganglioma is a rare neuroendocrine tumour that arises from neuroectodermally-derived paraganglionic cells throughout the body (15). They can occur at any age, but most often occur in the 4th or 5th decades. If hormonally active (functioning paragangliomas), they present with symptoms related to catecholamine secretion (headache, palpitations, sweating, hypertension). Non-functioning paragangliomas present as an enlarging palpable mass or pain.

**Imaging findings**

Extra-adrenal paragangliomas occur mainly in the retroperitoneum, most frequently in the infrarenal area near the origin of the IMA (15). They appear as para-aortic soft tissue masses with homogeneous enhancement or central areas of low attenuation (Fig. 13). Punctate calcification or foci of haemorrhage can be seen. MIBG (metaiodobenzylguanidine) scan often shows increased uptake by the mass. MRI shows the mass to be iso/hypointense on T1 and hyperintense on T2.

**G. Malakoplakia**

Malakoplakia is an uncommon granulomatous inflammatory condition (16). It is thought to arise as an abnormal response to infection. It can affect any system in the body (gastrointestinal, bone, lungs, lymph nodes, skin) but is most common in the genitourinary system, most often the urinary bladder. Malakoplakia is often associated with urinary tract infections (Escherichia coli) or immunosuppression (organ transplantation, chemotherapy, prolonged steroid use). Patients develop mass-like inflammatory change that can look like malignancy. The final diagnosis is histological. In our presented case (Fig. 14), there are bilateral perinephric masses, which are close to but separate to the kidneys. A CT-guided biopsy confirmed the diagnosis as
malakoplakia. No cases of this perinephric malakoplakia without renal involvement can be found in the literature, with most describing a renal mass with perinephric extension (17).

**Imaging findings**

Renal malakoplakia is seen as multiple poorly-enhancing lesions in the kidneys that causes renal enlargement. It can occur bilaterally and may extend into the perirenal region (17). Lesions appear as ill-defined low attenuation on CT and have variable echogenicity on ultrasound. In our case of perinephric malakoplakia, the masses are slightly heterogenously enhancing with surrounding fat stranding. The kidneys were unremarkable.
**Fig. 1:** Normal perinephric anatomy. The anterior border of the perinephric space is bounded by Gerota’s fascia (white arrows). The posterior border is bounded by Zuckerkanell's fascia (red arrows). The perinephric space is marked with an asterisk.

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Fig. 2: Bilateral homogeneous attenuation perinephric masses in an adult patient later confirmed to have lymphoma

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Fig. 3: Sagittal reformatted view of an adult with a peripherally enhancing collection in the perinephric region in keeping with an abscess. The underlying cause can be seen posteriorly as a calcified density, a small renal stone that has eroded through the kidney.

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**Fig. 4:** Axial CT of the same patient in Figure 2 showing a peripherally enhancing collection in the perinephric region in keeping with an abscess. The underlying cause can be seen posteriorly as a calcified density, a small renal stone that has eroded through the kidney.

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Fig. 5: Arterial phase CT in an adult with metastatic lung carcinoma. The mass was confirmed as a metastatic deposit. Lack of arterial enhancement is noted.

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Fig. 6: Sagittal reformatted view of an arterial phase CT in the same adult with metastatic lung carcinoma as in Figure 5. The mass was confirmed as a metastatic deposit. Lack of arterial enhancement is noted.

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Fig. 7: Bilateral perinephric masses in an adult patient confirmed following biopsy as metastatic deposits.

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**Fig. 8:** Coronal CT Urogram in a 69-year-old patient. An ill-defined low attenuation mass is seen surrounding the lower pole of the right kidney. This mass contains areas of fat-density. The underlying right kidney is unaffected.

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Fig. 9: Axial CT Urogram in the same patient as Figure 9. The low attenuation mass is again seen posteromedial to the right kidney. Areas of fat density within the mass are again noted.

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**Fig. 10:** Axial T1 MRI image in a 60-year-old patient. A well-defined isointense mass is noted adjacent to the lateral border of the right kidney

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Fig. 11: Coronal T2 MRI image in the same patient as Figure 10. The mass is again well-defined but low signal on T2.

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Fig. 12: Renal ultrasound image in the same patient in Figures 10 & 11, performed at the time of biopsy. The mass is seen as a well-defined hypoechoic mass surrounding the upper pole of the right kidney. The mass was confirmed as a capsular leiomyoma.

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Fig. 13: CT Abdomen in a 56 year old female patient who presented with left-sided abdominal pain. The image shows a heterogenously enhancing mass in the perinephric region on the left.

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**Fig. 14:** CT Abdomen in a 62-year-old female who presented with a history of abdominal bloating and weight loss. The coronal reformatted image shows bilateral soft tissue-attenuation perinephric masses, larger on the left. Biopsy confirmed malakoplakia.

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Conclusion

Perirenal masses may arise from within or outside the perinephric space. We describe normal perinephric anatomy important in the understanding of disease pathology, as well describe and illustrate multiple examples of perinephric masses on multiple imaging modalities.
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