Learning objectives

The purpose of this exhibition is to describe CT and MR image findings of idiopathic retroperitoneal disease and its mimickers presented as renal, para-renal or peri-ureteral soft tissues and to search the differential image patterns

Background

The idiopathic retroperitoneal fibrosis, IgG4-related disease, Erdheim-Chester disease and Urinary lymphoma involving urinary system show similar image features to each other. Some cases are open to be misdiagnosed because of overlapping image findings

Imaging findings OR Procedure details

1. **Idiopathic Retroperitoneal Fibrosis**

A range of diseases characterized by the presence of a fibro-inflammatory tissue, which usually surrounds the abdominal aorta and the iliac arteries and extends into the retroperitoneum to envelop neighboring structures.

Retroperitoneal fibrosis is generally idiopathic, but can also be secondary to the use of certain drugs, malignant diseases, infections, and surgery.

Potential pathogenesis: a local inflammatory reaction to antigens in the atherosclerotic plaques of the abdominal aorta

==> **chronic periaortitis**

The center of fibrosis is usually located at the level of the aortic bifurcation; this abnormal tissue bifurcates to follow the common iliac arteries.

CT and MRI are the most reliable imaging modalities.

CT: A homogeneous plaque, iso-dense with muscle, surrounding the lower abdominal aorta and the iliac arteries, and often enveloping the ureter and the inferior vena cava.
• CE: in the early, inflammatory stages of idiopathic retroperitoneal fibrosis, but not in later stage

MRI: provides a better definition against the surrounding tissue

• T1WI: Hypo SI
• T2WI: Hyper SI in the early or active stage due to tissue edema with hypercellularity (good for therapeutic response evaluation) --> Hypo SI in the late stage
• Inhomogeneity on T2WI suggests malignant retroperitoneal fibrosis

*Cases of IPF: See Fig 1. - Fig 5. images*

2. *IgG4 related sclerosing disease*

Elevation of serum IgG4 levels and infiltration of abundant IgG4 positive plasma cells into various organs are rather specific to AIP patients.

Systemic Ds entity: extensive IgG4-positive plasma cells and T-lymphocyte infiltration of various organs.

Autoimmune pancreatitis is a indicator of IgG4-related sclerosing Disease *(Figure 6).*

Histology: zonal tubulointerstitial nephritis with IgG4 plasma cell-rich inflammatory infiltrate, fibrosis, and tubular atrophy

Image findings> CT and MRI are the most reliable imaging modalities.

Involvement of kidney:

1. *(Mostly) bilateral / multiple & Predominantly involve the cortex*

   • Small peripheral cortical nodule
   • Well-defined or ill-defined round lesions
   • Well-defined wedge-shaped lesion
   • Diffuse patchy involvement

2. Diffuse rim of soft tissues around the Kidney
3. Irregular nodules in bilateral renal sinuses

4. Diffuse wall thickening of the pelvis

Involvement of ureter:

1. irregular periureteral soft tissue lesion with hydronephrosis
2. LN enlargement is relatively rare

Involvement of urinary bladder:

1. Focal soft tissue mass: resembles idiopathic retroperitoneal fibrosis
2. LN enlargement is relatively rare

**Cases of IgG4 related sclerosing disease:** See Fig. 7 - Fig. 13, images

3. **Lymphoma**

Primary lymphoma is rare because kidney does not normally contain lymphoid tissue. but, the kidney is one of the common sites of secondary lymphoma (hematogeneous spread or direct extension from retroperitoneal LN).

Non Hodgkin Lymphoma is more common than Hodgkin Disease.

Image findings> CT and MRI are reliable imaging modalities.

Involvement of kidney:

1. *(Mostly) bilateral / multiple round masses with retroperitoneal LN enlargement*
   - Poor enhancement, compared with non-involved renal parenchyma
2. Contiguous retroperitoneal extension: resembles idiopathic retroperitoneal fibrosis
3. Solitary mass
4. Diffuse infiltration of whole renal parenchyma
5. Perirenal soft tissue lesion surrounds renal parenchyma
Involvement of ureter:

1. irregular periureteral soft tissue lesion without hydropnephrosis
2. LN enlargement is common

_Cases of lymphoma involvement: See Fig 14. - Fig 18. images_

**4. Erdheim-Chester disease**

Erdheim-Chester disease (ECD) is a rare xanthogranulomatous non-Langerhans cell histiocytosis first described by Jakob Erdheim and William Chester in 1930.

Pathognomonic lesions: Bone lesion

- All patients reported in literature
- Bone scan: Bilateral symmetric uptake of bone within the metadiaphyses of the appendicular skeletons
- Plain x ray: Osteosclerosis of the diaphyses and the metaphyses of long bones with sparing of the epiphyses (Ddx. from LCH: Osteolysis with rarely involvement of the long bones)

Renal and perirenal involvement of (ECD) is frequent (29% of ECD)

- May be seen as isolated location of the disease
- Asymptomatic

Image findings> CT and MRI are reliable imaging modalities for diagnosis of renal-perirenal involvement of ECD.

**CT findings** of Renal and/or perirenal involvement:

1. (Mostly)Homogeneous, hypoattenuated soft tissue infiltration with weak contrast enhancement in the renal fossae
   - Extension of soft tissue into renal sinuses, pedicles and proximal ureters
2. Perirenal soft tissue lesion surrounds renal parenchyma
   - "Hairy kidney" appearance

**MR findings** of renal and/or perirenal involvement:
1. Iso- signal intensified soft tissue infiltration to muscle on T1- and T2-weighted images

2. Slight homogeneous enhancement after Gd injection

Cases of ECD: See Fig 19. - Fig 24. images

Images for this section:

Fig. 1: The first case of IPF
Fig. 2: The first case of IPF
Fig. 3: The first case of IPF

Diffuse infiltrative and patchy soft tissue mass around aorta and iliac vessels with encasement of aorta/iliac vessels (periaortic soft tissue +)
- Poorer enhancing features than adjacent normal parenchymae
- Hot uptake on PET-CT

Hydronephrosis (+) (d/t periureteral involvement)
Excretory delay (+) (d/t periureteral involvement)
LN enlargement (+)

Parapelvic soft tissue (-)
Renal parenchymal soft tissue (-)
Perirenal soft tissue(-)
Fig. 4: The first case of IPF - Follow up images After Pd therapy

Idiopathic retroperitoneal fibrosis (CASE1) – After Pd medication

<table>
<thead>
<tr>
<th>Markedly improved state</th>
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<tbody>
<tr>
<td>- Diffuse infiltrative and patchy soft tissue mass around aorta and iliac vessels with encasement of aorta/iliac vessels (periaortic soft tissue +)</td>
</tr>
<tr>
<td>- Hydronephrosis</td>
</tr>
<tr>
<td>- Excretory delay</td>
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<tr>
<td>- LN enlargement</td>
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</table>
Fig. 5: The second case of IPF
Fig. 18: The fourth case of renal lymphoma

Infiltrative soft tissue mass involving bilateral renal sinuses
- Encasement of renal cortex
  (Perirenal soft tissue mass)
- Parapelvic soft tissue (+)
- Perirenal space extension (+)
  - Encasement of renal hilar vessels without obstruction

Poorer enhancing features than adjacent normal parenchyma

Multiple LN enlargement around aorta and IVC

Hydronephrosis (-)
Periaortic soft tissue (-)

→ Low grade B cell lymphoma
Fig. 17: The fourth case of renal lymphoma
Fig. 16: The third case of renal lymphoma
**Fig. 19:** The first case of renal involvement of Erdheim-Chester disease
Fig. 20: The first case of renal involvement of Erdheim-Chester disease
**Erdheim-Chester disease (CASE1) – Important finding for D/dx**

<table>
<thead>
<tr>
<th>Bone scan</th>
<th>Orbital CT</th>
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<td>Hot uptake of long bones (extremities)</td>
<td>Bilateral Retro-ocular masses sheathing the optic nerves</td>
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<tr>
<td>CT coronal reconstructed image: Heterogeneous patchy sclerotic change of covered long bones (both femur heads and shafts)</td>
<td>Bilateral Diffuse infiltration of the retro-ocular fat</td>
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**Fig. 21:** The first case of renal involvement of Erdheim-Chester disease
Fig. 22: The second case of renal involvement of Erdheim-Chester disease

Erdheim-Chester disease (CASE2)

A 41-year old man

- Parapelvic soft tissue (+)
- Encasement of renal hilar vessels (-)
- Excretory delay (-)
- Renal parenchymal lesion(-)
- LN enlargement (-)

- Homogeneous, poorer enhancing features than adjacent normal parenchymae
- Hydronephrosis (+)
- Periaortic soft tissue (-)
**Fig. 6**: Schematic illustration showing the relationship between IgG4 related sclerosing disease and auto-immune pancreatitis.
Fig. 23: The second case of renal involvement of Erdheim-Chester disease
Fig. 15: The second case of renal lymphoma

Lymphoma (CASE2)

A 67-year old man

Infiltrative soft tissue mass involving Rt. renal hilum and Rt. Renal parenchyma
- Perirenal soft tissue mass (+)
- Parapelvic soft tissue (+)
- Encasement of renal hilar vessels without obstruction

Poorer enhancing features than adjacent normal parenchymae

Multiple LN enlargement around aorta and IVC

Hydronephrosis (+)
Periaortic soft tissue (+)
→ Diffuse large B cell lymphoma
Lymphoma (CASE1)

A 5-year old man

Multiple well-defined round soft tissue masses at both renal parenchymae
- Poorer enhancing features than adjacent normal parenchymae

Perirenal soft tissue mass (-)
Parapelvic soft tissue (-)
Encasement of renal hilar vessel (-)

LN enlargement around aorta and IVC (-)
Hydronephrosis/Excretion delay (-)
Periaortic soft tissue (-)
  → Precursor B lymphoblastic lymphoma

Fig. 14: The first case of renal lymphoma
**Fig. 7:** The first case of IgG4 related sclerosing disease.
Fig. 8: The first case of IgG4 related sclerosing disease
Fig. 9: The second case of IgG4 related sclerosing disease
Fig. 10: The second case of IgG4 related sclerosing disease
**Fig. 11:** The third case of IgG4 related sclerosing disease
**Fig. 12:** The third case of IgG4 related sclerosing disease

- Homogeneous T1WI and T2WI low SI feature with poorer enhancing features than adjacent normal parenchymae
- Encasement of renal pelvis and proximal ureter with smooth marginated tapering
- Hydronephrosis (+)  Renal parenchymal lesion (-)
- LN enlargement (-)  Periaortic soft tissue (-)
Fig. 13: The fourth case of IgG4 related sclerosing disease - the pattern of systemic disease (extra-renal involvement)
Fig. 24: The second case of renal involvement of Erdheim-Chester disease
**Conclusion**

To search the differential image patterns is important for appropriate therapeutic responses.

Most of IPF cases involving the urinary system including kidneys show soft tissue infiltration and fibrosis around abdominal aorta and iliac arteries. The renal involved IPF cases tend to show soft tissue mass in the renal sinus and perirenal rim of soft tissue.

The most common findings of IgG4-related sclerosing disease is bilateral, multiple renal parenchymal soft tissue lesion. In spite of wall thickenings of renal pelvis or upper ureter, the cases of total occlusion of urinary tract or severe hydronephrosis are rare.

The renal lymphoma is hematogenous spread or extension from retroperitoneal site. The most common findings of renal lymphoma are multiple soft tissues with poor enhancement. Most cases of lymphoma show retroperitoneal lymphadenopathy. The ureteral obstruction or vascular occlusion is relatively rare.

The pathognomonic lesion of Erdheim-Chester disease (ECD) is the osteosclerosis of long bones. The renal involvement of the ECD tends to show homogeneous, poorly enhanced tissue infiltration in the renal sinus with perirenal rim.

**References**


**Personal Information**