25th European Congress of Pathology – Lisbon, Portugal

Plenary Residents Seminar

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Clinical History:

- 46 years old Brazilian male with a right renal mass.
- Past medical history:
  - High blood pressure.
  - Perianal abscesses.
- No relevant family history.
Abdominal CT-scan:

“(…) a 4.5cm well defined, solid mass located in the middle third of the right kidney (…) with heterogeneous enhancement(…)“

Surgery: radical right nephrectomy.
Gross findings:

- 10x6x4 cm right kidney with a 4x3,5x3,5 cm well circumscribed and encapsulated round mass in the middle third.

- The cut surface was heterogeneous and white-tan with a solid consistency.

- No gross invasion of the renal sinus, renal pelvis or ureter.
Microscopic findings (I):

- Well-demarcated encapsulated tumor;

- Alternation of paucicellular and cellular areas;

  **Paucicellular areas:**
  
  - Hyalinized and edematous stroma with reticular vascular channels.

  **Cellular areas:**
  
  1. Rich vascular network with thin walled vessels lined by flattened to plump endothelial cells, sometimes with a hemangiopericytomatous appearance.
Microscopic findings(II):

- Cellular areas:
  - II. Stromal cells:
    - Polygonal cells with large and eosinophilic cytoplasm, sometimes with multiple and fine microvacuoles (lipid–laden cytoplasm);
    - Central or peripherally located round nuclei with occasional hyaline globules.

- Occasional moderate cellular atypia and exceedingly rare mitotic figures.

- Focal stromal hemorrhage.

- Inflammatory infiltrate of mast cells, lymphocytes and plasma cells.
Differential diagnoses:

I. Hemangioblastoma

II. Epithelioid Angiomyolipoma

III. Hemangiopericytoma

IV. Clear cell renal cell carcinoma

V. Extraintestinal GIST

VI. Adrenal cortical carcinoma (metastasis)

VII. Paraganglioma
### Immunohistochemistry

<table>
<thead>
<tr>
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<th>Positive</th>
<th>Negative</th>
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<tbody>
<tr>
<td>Stromal Cells</td>
<td>Vimentin</td>
<td>MNF -116</td>
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<tr>
<td></td>
<td>S100 protein</td>
<td>EMA</td>
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<td></td>
<td>NSE</td>
<td>Synaptophysin</td>
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<td></td>
<td>α-inhibin</td>
<td>Chromogranin</td>
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<tr>
<td></td>
<td>CD10 (focal)</td>
<td>CD57</td>
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<td>Desmin</td>
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<tr>
<td></td>
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<td>CD117</td>
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<td>MiTF</td>
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<td></td>
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<td>HMB45</td>
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<tr>
<td>Vascular network</td>
<td>CD34</td>
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</tbody>
</table>

Ki-67: < 1%
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<thead>
<tr>
<th>Tumor Type</th>
<th>Immunohistochemistry</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epithelioid Angiomyolipoma</td>
<td>HMB45, MiTF, muscle markers + (\alpha)-inhibin -</td>
</tr>
<tr>
<td>Hemangiopericytoma</td>
<td>CD34 and EMA +</td>
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<tr>
<td>Clear cell renal cell carcinoma</td>
<td>Pan-cytokeratin and EMA +</td>
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<tr>
<td>Extra-intestinal GIST</td>
<td>CD117 and CD34 +</td>
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<tr>
<td>Adrenal cortical carcinoma / paraganglioma</td>
<td>Synaptophysin + / Synaptophysin, chromogranin and CD57 +</td>
</tr>
</tbody>
</table>
Renal Hemangioblastoma
Follow up:

- No adjuvant treatment.
- Genetic study for von Hippel-Lindau disease in progress.
- NED/8 months.
References:


- Zhao M et al, PAX8 expression in sporadic hemangioblastoma of the kidney supports a primary renal cell lineage: implications for differential diagnosis. HUM PATH.

