CASE 5
SLIDE SEMINAR GYNECOLOGICAL PATHOLOGY
ECP NICE 2019

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CLINICAL HISTORY

• 70 y.o. female with chronic renal failure candidate for kidney transplantation.
• Preoperative studies showed a 4 cm solid lesion in the left adnexal region.
• A left salpingo-oophorectomy was performed.
DIAGNOSIS

ANASTOMOSING HEMANGIOMA
• ~60 Renal / ~60 extrarenal (paraspinal soft tissues, testis, ovary, adrenal, liver, GUT, bladder)
• Benign behavior
• 2-85 y (49y renal / 65y non renal)
• Male predominance
• 60% Asymptomatic
• Difficulty in preoperative diagnosis (overtreatment) / Unfamiliarity with its histologic characteristics (misdiagnosis of angiosarcoma).
## Anastomosing Hemangioma of the Ovary

<table>
<thead>
<tr>
<th>Case No. /Age (y)</th>
<th>Tumor location</th>
<th>Clinical manifestations</th>
<th>Tumor size (cm)</th>
<th>Follow-up (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 / 70 (1)</td>
<td>Right, cortex</td>
<td>Endometrial carcinoma</td>
<td>0.2</td>
<td>NED (25)</td>
</tr>
<tr>
<td>2 / 49 (1)</td>
<td>Right, cortex</td>
<td>Bilateral serous cysts</td>
<td>0.1</td>
<td>NED (16)</td>
</tr>
<tr>
<td>3 / 77 (1)</td>
<td>Left, cortex and medulla</td>
<td>Serous cystadenoma</td>
<td>1.1</td>
<td>NED (32)</td>
</tr>
<tr>
<td>4 / 66 (2)</td>
<td>Laterality unknown</td>
<td>Metrorrhagia</td>
<td>0.5</td>
<td>NED (25)</td>
</tr>
<tr>
<td>5 / 43 (2)</td>
<td>Left</td>
<td>Leiomyomas and benign ovarian cyst</td>
<td>1.3</td>
<td>NED (4)</td>
</tr>
<tr>
<td>6 / 69 (2)</td>
<td>Right</td>
<td>Leiomyomas</td>
<td>1.5</td>
<td>NED (52)</td>
</tr>
<tr>
<td>7 / 81 (2)</td>
<td>Right</td>
<td>Metrorrhagia, adenomyosis</td>
<td>3.5</td>
<td>NA</td>
</tr>
<tr>
<td>8 / 68 (2)</td>
<td>Left</td>
<td>Ovarian tumor, ascites and increased CA125</td>
<td>3.5</td>
<td>NA</td>
</tr>
<tr>
<td>9 / 69 (2)</td>
<td>Right</td>
<td>Ovarian tumor (patient with lymphoma)</td>
<td>1.2</td>
<td>NED (13)</td>
</tr>
<tr>
<td>10 / 62 (3)</td>
<td>Right, cystic</td>
<td>Cystic ovarian mass</td>
<td>9 (4 cm solid area in an hematoma)</td>
<td>NA</td>
</tr>
<tr>
<td>11 / 50 (4)</td>
<td>Right</td>
<td>Abdominal pain and ascites</td>
<td>3</td>
<td>NA</td>
</tr>
<tr>
<td>12 / 70 (5)</td>
<td>Left, medulla</td>
<td>Asymptomatic</td>
<td>4</td>
<td>Dead of unrelated cause (21)</td>
</tr>
</tbody>
</table>
ANASTOMOSING HEMANGIOMA.
PATHOLOGY

- Well circumscribed, nonencapsulated, red-brown, spongy texture
- 1 mm-9 cm (mean 2.2 cm)
- Solitary (multifocal in end stage renal disease)
- Diffuse – vaguely lobulated
- Irregular tightly packed capillary-sized vessels with prominent anastomosing morphology
- Single layer of bland endothelial cells / hobnail morphology
- Mitoses absent or rare
- Intravascular fibrin trombi
- Extramedullary haematopoiesis
- Intracytoplasmic hyaline globules
- Intravascular extension may be seen
- Luteinized cells at the periphery
- Diffuse staining for endothelial markers (F VIII, CD31, CD34, ERG, FLI1)
- D2-40, CD8, HHV8 and epithelial markers NEGATIVE
Intravascular extension
Recurrent *GNAQ* mutations in anastomosing hemangiomas

Gregory R Bean¹, Nancy M Joseph¹, Ryan M Gill¹, Andrew L Folpe², Andrew E Horvai¹ and Sarah E Umetsu¹

Anastomosing hemangiomas are recently described benign vascular lesions that occur chiefly in the genitourinary tract and paravertebral soft tissues. Owing to their rarity and unusual cytoarchitectural features, anastomosing hemangiomas are frequently confused with low-grade angiosarcomas. The specific genetic alterations underlying these lesions are currently unknown. We performed capture-based next-generation DNA sequencing analysis on 13 anastomosing hemangiomas and identified frequent somatic mutations in the heterotrimeric G-protein alpha-subunit, *GNAQ*. Nine of 13 cases (69%) harbored a somatic mutation at *GNAQ* codon 209, a known hotspot that is commonly mutated in uveal melanoma and blue nevi, as well as various congenital vascular proliferations. No other pathogenic or likely pathogenic mutations were identified in these genetically simple lesions. The finding of a recurrent driver mutation in the G-protein signal transduction pathway provides strong evidence that anastomosing hemangiomas are indeed clonal vascular neoplasms. *Modern Pathology* (2017) 30, 722–727; doi:10.1038/modpathol.2016.234; published online 13 January 2017

2 ovarian / 1 renal cases: codon 209 GNAQ wt
DIFFERENTIAL DIAGNOSIS
Cavernous Hemangioma
Ovarian angiosarcoma

- 29 patients
- 19-77 y (mean 37 y)
- Abdominal pain and/or abdominal distension
- 64% (16/25) DOD

Ovarian angiosarcoma

- 3.5-25 cm (mean 12.5 cm)
- Unilateral. Extraovarian extension
- Association with mature teratoma / mucinous cystadenoma
- Highly atypical cytology
- Lobulated architecture
- Solid, cystic, interanastomosing vascular channels, reticular and fascicular patterns
- Brisk mitotic activity

Angiosarcoma
HGSC post-chemotherapy
Microcystic Stromal Tumor of the Ovary
Report of 16 Cases of a Hitherto Uncharacterized Distinctive Ovarian Neoplasm

Julie A. Irving, MD*† and Robert H. Young, MD‡§
Conclusions

• AE is a rare but distinct type of vascular neoplasia with benign behavior
• Ovary may be involved
• Although microscopic features may appear alarming at first sight, the absence of significant cytologic atypia, as well as the identification of the characteristic histologic pattern, should lead to an accurate diagnosis