Right Parotid Gland Tumor

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Speaker: Dr. Kimihide Kusafuka

No declare of Interest
Patient: 44 year-old Japanese man.
Main Complain: Swelling of the right neck.

History:
*X-10 year: He noticed the swelling of the right neck.
   The right neck mass had rapidly swollen.
*X-1 year: FNAC was performed: “Suspicious of malignancy”.
He admitted to the hospital. The 4 x 4 cm unmovable mass was found in the right parotid region. MRI showed a 40x42x70mm irregular-shaped mass with high intensity in the right parotid gland. CT showed the similar findings.
*Jan. X year: Total parotidectomy and right neck dissection.
   Post-operative radiotherapy(60Gy) was performed.
*Oct. X year: FGD-PET indicated multiple bone metastases
*He died to disease, 21 months after operation.
【Operation material】
Diffuse proliferation
Less cohesive cancer cells showing eosinophilic cytoplasm and eccentric nuclei.
Immunohistochemical examination
CK7

Adenocarcinoma
Salivary duct carcinoma

GCDFP-15

AR
Apocrine-HER2 subtype!
CK5/6

Intraductal component(+)
Ki-67 high

High-grade malignancy

p53(MT+)
This tumor did not indicate the co-expression of pan-CK and vimentin. Moreover, intact expression of SMARCB1.
No expression of E-cadherin and β-catenin.
Genetic analysis
### PCR-based analysis on CDH1 gene

<table>
<thead>
<tr>
<th>Case No</th>
<th>Exon</th>
<th>Nucleotide change</th>
<th>AA change</th>
<th>Genotype</th>
<th>Mutation site</th>
<th>Mutation Taster</th>
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<tbody>
<tr>
<td>Case 1</td>
<td>4</td>
<td>c.490C&gt;T</td>
<td>p.(Pro164Ser)</td>
<td>c/t</td>
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<td></td>
<td>5</td>
<td>c.540C&gt;T</td>
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<tr>
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<td>14</td>
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<td>c.2476C&gt;T</td>
<td>p.(Pro826Ser)</td>
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<td>c.388G&gt;A</td>
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<td>Case 10</td>
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### Diagram

Case 1  
Exon 4  
C>T(G>A)
At least, this tumor is “Carcinoma ex pleomorphic adenoma, widely invasive”.

The carcinomatous component is composed of diffuse proliferation of less cohesive cancer cells with eosinophilic cytoplasm and eccentric nuclei.

It shows the salivary duct ca.-like phenotype: GCDFP-15(+), AR(+) and HER2(+).

CK(+), but vimentin(-). Intact expression of SMARCB1.

Loss of E-cadherin and β-catenin expression.

Genetically, it shows the missense mutation of CDH1 gene.
【Final diagnosis】

Salivary duct carcinoma with rhabdoid-like features (SDCRF) ex pleomorphic adenoma, widely invasive.
Originally, we reported 2 cases of SDC with rhabdoid features of the salivary glands as a new entity: Diffuse proliferation of less cohesive ovoid cancer cells with eosinophilic cytoplasm and eccentric nuclei.

After then, we collected and extracted 17 cases of SDCRF: CXPA, 9 cases (IC, 1 case); de novo, 4 cases (SDCIS, 1 case); Bx only 1 cases; Consultation cases: 3 cases.

Eight cases died of disease; 2 cases lost the follow-up but LN meta(+) at the last day.

The cancer cells are not true “rhabdoid cells”, from neither co-expression of CK and vimentin, nor round-shaped intracytoplasmic inclusion body: Therefore, we called them “rhabdoid-like cells”

Immunohistochemically, SDCRF shows no or aberrant expression of E-cadherin and β-catenin.

Genetically, approximately half cases examined shows the missense mutation or insertion of CDH1 gene, coding E-cadherin.
【another SDCRF case 1】

de novo SDCRF

E-cadherin

exon 4
G>A
【another SDCRF case 2】

SDCRF in situ

calponin

E-cadherin
**Conclusion**

SDCRF is a rare and new variant of SDC. From the morphology, immuno-phenotype of E-cadherin and CDH1 gene mutation, SDCRF is a salivary counterpart of pleomorphic lobular carcinoma of the breast (PLCB).

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Thank you for your attention♫