**Introduction**

Autosomal dominant DICER1 mutations are among the causes of early-onset familial cancer. DICER-1 mutation has been shown in pleuropulmonary blastomas as well as ovarian tumors, thyroid, parathyroid, pituitary, adrenocortical and testicular tumors. It is important to be aware of the risk for the development of other cancers in the follow-up of these cases. DICER-1 mutation was detected in two cases who presented with different rare endocrine tumors.

**Case 1**

8.5 year old girl

- Complaint: Deepening voice and hirsutism
- Family History: Parents are first degree cousins
  - Aunt had a history of thyroid cancer
- Physical Examination:
  - Height was significantly above the familial target height percentile
  - Thelarche: Tanner stage 1, pubic hair development: stage 5
  - Size of the clitoris: 3x1cm
  - Increased muscle mass
- Laboratory evaluation:
  - Total testosterone: 231.6 ng/dl (N<10)
  - USG: A mass of 43 mm in the left adrenal area
- Pathologic diagnosis: Adrenocortical cancer
- Treatment: Chemotherapy + Mitotane

During the follow-up visits, a simple ovarian cyst with the diameter of 3cm was detected and disappeared spontaneously.

**Laboratory Results**

<table>
<thead>
<tr>
<th>Testosterone (ng/dl)</th>
<th>TSH (mIU/L)</th>
<th>FT4 (ng/dl)</th>
<th>Calcitonin (pg/ml)</th>
<th>Thyroid USG</th>
<th>LDH (U/L)</th>
<th>AFP (IU/ml)</th>
<th>B-hCG (Ng/ml)</th>
<th>CA-125 (U/ml)</th>
</tr>
</thead>
<tbody>
<tr>
<td>CASE-1</td>
<td>231.6</td>
<td>2.26</td>
<td>1.48</td>
<td>-</td>
<td>Normal</td>
<td>273/179</td>
<td>0.945</td>
<td>&lt;2</td>
</tr>
<tr>
<td>CASE-2</td>
<td>&lt;2.5</td>
<td>2.65</td>
<td>1.59</td>
<td>&lt;2</td>
<td>8mm nodule</td>
<td>681</td>
<td>2.21</td>
<td>&lt;2</td>
</tr>
</tbody>
</table>

**Case 2**

6.5 year old girl

- Complaint: Abdominal pain
- Family History: Parents are second degree cousins
- Physical Examination:
  - A mass was palpated in the abdomen
  - Pubertal development was Tanner stage 1
- Laboratory evaluation:
  - Elevated CA-125 and LDH levels
- USG: A mass of 15 cm in diameter with right adnexial origin
- Pathologic diagnosis: Sertoli-Leydig cell tumor
- Treatment: Chemotherapy

During follow-up, a solid nodule (8 mm in long axis) in the left lobe of the thyroid gland was detected. Fine needle aspiration biopsy revealed a benign lesion.

**Conclusion**

As seen in our cases, DICER-1 mutation should be considered in the presence of multiple organ involvement in endocrine cancers and other endocrine organ pathologies should be kept in mind during the follow-up period.