**Background**

Gain-of-function mutations in *KISS1* and *KISS1R* genes and loss-of-function mutations in the gene encoding the makorin RING-finger protein 3 (*MKRN3*) expressed only in the paternal allele are the most common genetic reasons of familial central precocious puberty (CPP).

**Aim**

We report a case of familial CPP and a pathogenetic variant in the *MKRN3* gene.

**Case**

7 years and 4 months old, girl

**Complaint:** Breast and pubic hair development of three months duration

**Medical history:** Normal

**Family history:** Unrelated parents, precocious puberty history in her father, paternal uncle and cousins (Figure 2).

**Physical examination**

- Weight: 34 kg (2.06 SDS), Height: 127 cm (0.78 SDS), BMI: 97p
- Target height 148.1 cm (-2.01 SDS)
- Predicted adult height 148.4 cm (-2.26 SDS)
- Mother's height -1.52 SDS, Father's height -3.44 SDS
- Breast Tanner stage II
- Pubic hair Tanner stage II
- Other system examinations normal

**Laboratory and imaging findings:** Thyroid function tests and routine biochemical examinations were normal. Other laboratory and imaging results are shown in Table 1.

**Table 1. Laboratory and imaging findings**

<table>
<thead>
<tr>
<th>Baseline LH (N&lt;0.1)</th>
<th>Baseline FSH (N: 0.1-4.3)</th>
<th>E₂ (N&lt;12)</th>
<th>Peak LH after LHRH test</th>
<th>Peak FSH after LHRH test</th>
<th>Bone age (Greulich-Pyle)</th>
<th>Uterus length</th>
<th>Right ovary</th>
<th>Left ovary</th>
<th>Cranial MRI</th>
</tr>
</thead>
<tbody>
<tr>
<td>0.8 IU/L</td>
<td>4.1 IU/L</td>
<td>17 pg/mL</td>
<td>18.1 IU/L</td>
<td>14.1 IU/L</td>
<td>10.5 years</td>
<td>50 mm</td>
<td>3.8 mL</td>
<td>3.3 mL</td>
<td>Normal</td>
</tr>
</tbody>
</table>

**Clinical follow-up**

- GnRH agonist treatment was started (3.75 mg leuprolide acetate every 28 days)

9 years and 3 months

- Weight 47 kg (2.25 SDS), Height 139.5 cm (0.55 SDS)
- Annual growth velocity 6.5 cm/year
- Bone age 12 years (ΔBA/ΔCA: 0.75)
- Predicted adult height 154.8 cm (-1.17 SDS)
- Breast Tanner stage III, Pubic hair Tanner stage III

**Conclusion**

In the evaluation of CPP cases, family history and genetic analysis are important in terms of early diagnosis and treatment with genetic counseling of the next generations.