PNH is a malformation of cortical development, due to abnormal neuronal migration, in which a subset of neurons fails to migrate into the developing cerebral cortex and remains as nodules that line the ventricular surface. PNH is usually missed on prenatal sonographic examinations. The prenatal diagnosis is possible due to coexistence of PNH with brain malformations such as ventriculomegaly, posterior fossa anomalies, or agenesis of corpus callosum.

We present two cases of isolated PNH suspected at 24 and 20 weeks respectively, in pregnant women with unremarkable history. Our ultrasound examination revealed in both cases irregular borders of the bodies of lateral ventricles on axial and sagittal views. Maternal MRI confirmed the diagnosis in the early third trimester. PNH appeared on MRI images as periventricular nodules protruding into the ventricular lumen and demonstrating the same signal intensity as did the gray matter on T1 and T2 sequences. Antenatal findings were confirmed with postnatal MR imaging. Neurodevelopmental outcome is good in both cases without seizures at the moment (4 years old and 6 months). A genetic analysis for the FLNA mutation was performed in both cases, and the results were negative.

**Conclusion**

Isolated PNH is detectable in the fetus by both sonography and MR imaging in the mid second trimester evaluation. Irregular ventricular walls on axial view and irregular square-shaped lateral ventricles on coronal view are suggestive of PNH.