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A case of central nervous system anomaly: syntelencephaly

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Objective

Syntelencephaly is a very rare central nervous system anomaly also known as middle interhemisferic variat holoprosencephaly (HPE) which described in 1993. Syntelencephaly is characterized by abnormal midline union of the posterior frontal and parietal lobes with variable fusion of thalami. Interhemisferic fissures always present anteriorly and posteriorly and fetal face is usually normal also a spesific type of disgenesis of corpus callossum (CC) (genu and splenium parts of CC are presentes but corpus is absent) accompanied with this anomaly and aziygos anterior cerebral artery is always presents this two signs can be accepted patognomic for syntelencephaly also neuronal migration disorders can be associated with syntelencephaly.

Methods

We present a case of syntelencephaly in a 27 weeks fetus, who refered our center because of polihydramniosis. It was the second pregnancy of the patient and there were no significant history due to this pregnancy and patient had been refused all anuploidi screening tests and fetal anomaly scaning. The 2D ultrasonography was used to carry out ultrasonic examination and there was no polhidromniosis but a dismorfic csp was present. İn the mid- sagittal plane genu and splenium parts of CC were present but corpus of CC was absent and radial sulcus were seen. Posterior and anterior interhemisferic fissures were seen but lateral vetricules were fused in the middle. Talamus and fetal face and fetal development were normal.

Results

This case illustrates the acurate diagnosis of syntelencephaly and discuss the features differentiating it from other more common forms of HPE.

Conclusion

This case illustrates the acurate diagnosis of syntelencephaly and discuss the features differentiating it from other more common forms of HPE.