Congenital Acute Lymphoblastic Leukemia with Placental Involvement: Case Report x

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Abstract

The authors describe a case of congenital acute lymphoblastic leukaemia (ALL), with extensive placental involvement, causing intratuterine fetal growth restriction and unexpected fetal demise at 33 weeks. Although unusual, other congenital tumours, including neuroblastoma and hepatoblastoma, can metastasize to the placenta. Congenital malignancy must be considered in the differential diagnosis of an abnormally large placenta. This case emphasizes the important role of careful histopathologic examination of the placenta which, combined with immunohistochemistry and clinicopathologic correlation, may establish the accurate diagnosis.
and with a worse prognosis than childhood ALL [4]. Prenatal features are non-specific, with hydrops and hepatosplenomegaly present in two-thirds of cases and intrauterine death complicating about half [2]. None of these signs were identified on the three ultrasounds performed during pregnancy. However, at fetal autopsy, a mild hepatomegaly was recorded.

Likewise intrauterine infections, genetic disorders (e.g.: Beckwith-Wiedemann syndrome) or maternal disease (e.g.: anaemia, diabetes), the congenital malignancies may cause placental enlargement [4,5]. Most frequent chromosomal abnormalities, including Down syndrome, more often associated with this diagnose, were excluded.

The diagnostic criteria for congenital leukaemia include proliferation of immature leucocytes, infiltration into the extra haematopoietic tissue and absence of diseases that can cause leukaemia or leucoerythroblastic reactions [4,5]. Moreover, genetic or chromosomal disorders that may be associated with unstable haematopoiesis, such as trisomy 21, leukemoid reactions and blood group incompatibility must be distinguished from acute leukemia [6].

Some authors have suggested a link between prenatal viral exposure and leukaemia risk but the only link found was the association between the Epstein-Barr virus (EBV) and ALL [7,8] and it was excluded in our case.

Few months later after fetal demise, the patient was diagnosed with a Large B-cell lymphoma. Metastases of maternal cancer to the placenta and fetus are rare in cases of maternal primary malignancy. Since malignant cells were never identified in placental interstitial space, placental and fetal metastasis from this maternal haematological cancer would be unlikely. An increased risk of non-Hodgkin lymphoma among individuals with a family history of hematopoietic malignancy (e.g.: chronic lymphocytic leukemia) was also reported [9].

Extensive stromal and vascular involvement of the placenta by leukemic cells was considered to be the probable cause of this IUGR and unexpected fetal death.

Conclusion

Congenital malignancy must be considered in the differential diagnosis of an abnormally large placenta.

The dissemination of fetal malignant disease involving the placenta, such as congenital acute lymphoblastic leukemia, and the fact that most congenital neoplasm are not apparent at birth still presents a considerable diagnostic challenge to healthcare professionals.

This case emphasizes the important role of careful histopathologic examination of the placenta which, combined with immunohistochemistry and clinicopathologic correlation, may establish the accurate diagnosis.

References