European Society of Pathology

30th European Congress of Pathology

Pathology: Path to Precision medicine

8 – 12 September 2018
Euskalduna Conference Centre, Bilbao, Spain

www.esp-congress.org

Abstracts

Conclusion: Despite obvious potential risks with Port-A-Caths, they are useful in many cases. With this case we would like to highlight the importance of having good 1) anatomy knowledge 2) dissection skills 3) taking good photographs to perform high quality paediatric autopsies. Pathologists should always be aware that findings during an autopsy may trigger legal proceedings.

PS-18-007

Gaucher's disease diagnosed on bone marrow biopsy performed for suspicion of malignant hemopathy

S. Chouchane*, M. Njima, N. Ben Abdeljelil, S. Ben Khalifa, R. Hadhri, A. Moussa, A. Zakhama, L. Njim
*Fattouma Bourguiba Hospital, Pathology, Ksibet El Mediouni, Tunisia

Background & Objective: Gaucher's disease (GD) is an autosomal recessive lipid storage disorder due to deficient or defective production of a lysosomal enzyme glucocerebrosidas. It is a rare genetic disorder. Here we report a case of a child in whom a blood cancer was suspected and the correct diagnosis of GD was based on bone marrow biopsy.
Method: This case was diagnosed at in our department of Pathology at of the University hospital of Monastir.
Results: A six-year-old female child with history of acute osteomyelitis of the distal extremity of the left femur presented with severe pain of the distal extremity of the right femur. On examination, the child had delayed milestones. The spleen was 11 cm palpable below the left costochondral margin. Haemoglobin was 8.2g/dl and Platelet count was 181,000/μL. An MRI of the right knee showed a diffuse infiltration of the bone marrow directing to a malignant hemopathy. A bone marrow biopsy was performed and showed sheets of Gaucher's cells seen as histiocytes with abundant granular and fibrillar cytoplasm. These cells had small eccentrically placed nuclei and had a crumpled tissue paper appearance. The diagnosis of GD was given and later confirmed by glucocerebrosidase levels estimation.
Conclusion: We report a patient with acute osteomyelitis as the single classical symptom, in whom Gaucher cells detected in bone marrow biopsy indicated the correct diagnosis of GD after clinical and radiological suspicion of malignant hemopathy. Our case might emphasize the utility of bone marrow study for GD, especially in cases where a blood cancer is suspected.

PS-18-008

Association between placental syncytiotrophoblast remodeling and angiogenic factor expression under hypoxia conditions

A. Savchenko*, O. Reshetnikova, L. Rudiuk, A. Bessarabova
*Baltic Federal University, Kaliningrad, Russia

Background & Objective: Gestation accompanied with maternal congenital heart disease (MCHD) is at risk with adverse pregnancy outcomes. We aimed to reveal the association of VEGF expression in placental syncytiotrophoblast (STB) and epithelial structural remodeling in cases of MCHD.
Method: 35 term placentas were divided into groups: I - 20 cases of MCHD and 15 cases of physiological pregnancy (control group). Chorionic villous morphology evaluated microscopically and by the computer morphometry. The standard immunohistochemical staining protocol with monoclonal mouse antibody to VEGF (RTU, Spring) for placental tissue samples has been developed. A standard single-stage protocol was performed with high-temperature antibody unmasking in 0.01 M citrate buffer (pH 7.6). Differences between groups' data were elucidated by non-parametric Wald-Wolfowitz test. Reliability established at $p<0.05$.
Results: Microscopic investigation revealed multiple foci of placental epithelial injury in cases of MCHD. Higher degree of pathologic changes discovered at peripheral and paracentral regions. Volume fractions of the terminal villi STB decreased in the central, paracentral and peripheral placental zones under MCHD conditions. These were accompanied with placental membrane thinning and higher expression of the VEGF within the epithelial layer.
Conclusion: The placental membrane thinning due to decreased STB volume fraction may accommodate fetal –maternal metabolism and gas exchange under hypoxia conditions in cases MCHD. The VEGF involvement in the control of fetal capillaries and syncytiotrophoblast remodeling discussed.

PS-18-009

Maternal factors and feto-placental remodeling in pregnancies with congenital heart disease

A. Bessarabova*, O. Reshetnikova, L. Rudiuk, A. Savchenko
*Baltic Federal University, Faculty of Medicine, Kaliningrad, Russia

Background & Objective: Number of pregnancies in women with cardiac disease is increasing worldwide. Heart distress during gestation always carries arduous challenge for physicians. The understanding of the adaptation mechanisms in 'mother-placenta-fetus' system may prevent maternal and fetal risk in pregnancy. To study maternal data, pregnancy outcomes and feto-placental peculiarities in operated vs non-operated cases of congenital heart disease (CHD) was the aim of present investigation.
Method: A total 39 patients of CHD were taken in this study. These were divided into two groups. Group 1- 20 patients who had no cardiac surgery and Group 2- 19- who had cardiac surgery to correct their cardiac defect before pregnancy. The control group (CG) included 15 cases of physiological gestation and childbirth. Medical records data, newborn - placental parameters, morphological and stereometric characteristics of placental villous chorion were compared in the two groups and CG using statistical analysis. Differences between groups' data were elucidated by non-parametric Wald-Wolfowitz test. Reliability established at $p<0.05$.
Results: Present study has shown that patients with CHD in pregnancy have a higher risk of obstetric complications. Mean newborns birth weight and body length, placental organometric parameters were smaller in cases of CHD. Cardiac pathology was accompanied with increased volume fraction of dystrophic processes in placental chorion structures. Nevertheless, the structural remodeling in placental fetal capillaries, villous membranes were very noticeable in operated vs non-operated cases of CHD.
Conclusion: Medical correction of hemodynamic disorders prevented placental insufficiency and thus contributed to successful maternal and fetal outcome of pregnancy.

PS-18-010

Features of the hidden immunodeficiency in a newborn baby whose mother suffered from leukemia

V. Sikora*, A. Romaniuk, M. Lyndin, O. Smiyan, O. Romaniuk, R. Moskalenko, N. Hyriavenko, L. Karpenko
*Sumy State University, Pathology, Ukraine

Background & Objective: To study the features of the pathomorphological changes in the intestines, spleen and thymus gland in a baby with the hidden congenital immunodeficiency provoked by the BCG vaccination.
Method: We present a case of the death of the newborn baby (14 days of life), whose mother suffered from acute leukemia in youth. The baby was born without pathologies. Therefore, she was vaccinated against hepatitis B (Engerix-B) and tuberculosis (BCG) according to the immunizations schedule. In a week the state of health of the baby became worse, the disease progressed despite the pharmacological treatment and led to multiple organ failures that caused the death. During the postmortem examination the changes in the intestines, spleen and thymus gland were observed and were studied in detail by using the histological-immunohistological (CD3/CD79α/CK-pan) methods.

Results: The histological study of the intestines revealed the diffuse foci of necrosis, strong leukocyte infiltration and vascular congestion with multiple hemorrhages. Perivascular T-cells hypoplasia was observed in the spleen. Thymus lobules were reduced, cortex thickness (lymphocyte loss and reticular stroma collapse) and proliferation of the epithelial reticular cells were observed. Numerous Hassall's corpuscles, different in their size, were observed. The results of the immunohistochemical study of the spleen showed the absence of the perivascular T-cells and the follicular hyperplasia of B-lymphocytes at the same time. Small number of CD3/CD79a-positive cells was observed in the thymus gland, CK-pan - significantly expressed in epithelial-reticular cells. The abovementioned changes indicate the development of the congenital T-cells immunodeficiency that led to the ulcerous-necrotic enterocolitis.
Conclusion: This clinical case demonstrates the importance of early diagnosis of immune status in the newborns, whose parents have leukemia in anamnesis. BCG-vaccination against the background of T-cell immunodeficiency may lead to the dangerous complications that threaten baby's life.

PS-18-011
The effect of anemia and malaria on placental vascularisation; a stereological analysis to identify the most vulnerable time point in pregnancy

K. Nielsen*
*Aarhus University Hospital, Institute of Pathology, Aarhus, Denmark

Background & Objective: This study aimed at investigating how placental vascularization is affected by anemia and/or malaria during pregnancy to identify the most vulnerable timepoints.
Method: The placentae were sampled by Systematic Uniform Random sampling and further sectioned by Isotropic Uniform Random sampling. Stained slides were analyzed by 197 placentae were collected in Tanzania assessing the volume, length and surface area of the vessels. The placentae were categorized as either control, anemic (Hb≤9g/dL, 9g/dL<Hb≤11g/dL) or malaria positive. The effect on transport and diffusion vessels and villi were investigated in four different intervals in pregnancy by gestational age (GA)
Results: Anemia: Only anemia after a GA of 28 weeks influences placental angiogenesis. Mild anemia reduced length of diffusion vessels but increased the length of transport vessels. Surface and volume were increase for both vessel types. Malaria Malaria: before a GA of 14 weeks increased surface of diffusion vessels and decreased the length of diffusion and volume of transport vessels. Malaria after a GA of 28 weeks increased all measured indices of angiogenesis except for surface of diffusion vessels. Malaria and/or anemia at GA 14+1-27+6 did not affect placental angiogenesis
Conclusion: Only malaria in early pregnancy has an influence on placental development whereas both anemia and malaria in the late pregnancy have a significant effect on the placental angiogenesis. After a GA of 28 weeks the placental demands are high due to fetal growth spurt. Increased surface, length and volume could be an attempt to compensate for the decreased oxygen due to anemia and disturbed diffusion surface due to malaria.

PS-18-012
ICAM1 and VCAM1 gene expression level in women with insufficient miscarriage

E. Valeeva*, E. Yupatov, O. Kravtsova, L. Maltseva, D. Valieva, E. Solgatova
*Kazan Federal University, Institute of Fundamental Medicine, Russia

Background & Objective: The physiological course of pregnancy in addition to external factors depends largely on the functional state of the vascular system. Endothelial dysfunction is usually accompanied by a systemic inflammatory process. It is believed that in the early stages of endotheliosis adhesion molecules ICAM-1 is strongly expressed, and VCAM-1 appears in acute endothelial dysfunction. However, despite all available information, the role of ICAM-1 and VCAM-1 in the pathogenesis of insufficient miscarriage still unclear. To evaluate the ICAM-1 and VCAM-1 genes expression level in women with insufficient miscarriage.
Method: Gene expression was measured by qPCR based on TaqMan technology in venous blood samples from 29 women with uncompleted pregnancy (UP) (I-II trimester) and 79 physiologically pregnant women (PP) collected during 22-24th, 32-34th weeks of pregnancy, separately in primapara and multigravidas.
Results: The mean level of ICAM1 activity was much higher in third trimester in PP women (primapara and multigravidas) compared to non-pregnant women (RQ 4.11 and 3.89 respectively ($p<0.05$)) but in UP group we detected significant decrease of its activity (RQ 0.0144, p 0.0453). Moderate increasing of VCAM1 gene activity also detected in PP women during gestation ($p>0.05$) but in UP group we showed almost 6-fold increasing of its expression (RQ 6.57, $p=0.037$).
Conclusion: Measurement of gene activity adhesion molecules ICAM1, VCAM1 is necessary for understanding the mechanisms of obstetrical pathology in pregnant women complicated with varicose veins of the pelvis.

This study was supported by Program of Competitive Growth of KFU.

PS-18-013
A new case of foetal fibrochondrogenesis – a diagnostic approach

V. Almeida*, C. Faria, J. Fraga, C. Cerdeira, B. Pimentão, A. C. Lai, H. Moreira, R. Almeida, R. Oliveira, P. Rodrigues, R. Pina, M. A. Cipriano
*CHUC, Pathology, Coimbra, Portugal

Background & Objective: Skeletal dysplasias are a rare and heterogeneous group of diseases that affects the development or growth of the chondro-osseous tissues. Presently, more than 450 skeletal dysplasias grouped by clinical, radiographic and molecular criteria are described. This work presents a case of a 21 weeks fetus with prenatal suspicion of skeletal dysplasia.
Method: Routine second trimester ultrasonography of an uncomplicated pregnancy revealed a male fetus with large head, frontal bossing and flat small nose. The long bones were short with normal echogenicity. The pregnancy was terminated due to the suspicion of skeletal dysplasia.
Results: The autopsy showed macrocephaly, prominent eyes with increased outer intercanthal distance and small nose. The most prominent feature was the markedly short limbs. Radiological examination confirmed rhyzomelic micromelia and showed metaphyseal flaring and methaphyseal spurs. The spine demonstrated platyspondyly and the ribs were short with metaphyseal cupping. Histophatological examination of the femur growth plate revealed a grossly disorganized hypercellular growth plate, with no demarcation between resting and proliferative zones. Chondrocytes of the resting zone were spindle shaped and intercellular matrix showed interwoven fibrous septae. Diaphyseal and metaphyseal trabecular bones were normal. Molecular studies identified one single mutation on gene COL11A1.
Conclusion: The evidence leads to the conclusion that the fetus had fibrochondrogenesis, a severe chondrodysplasia. A single mutation on gene COL11A1 is not sufficient to support the diagnosis of fibrochondrogenesis. However, the diagnosis is maintained by radiologic features and definitely confirmed by the unique histopathologic pattern. Genetic confirmation would however have been useful to improve genetic counselling.

PS-18-014
Adenoid cystic carcinoma of the lacrimal gland: a rare paediatric case

I. Franckevica*, S. Valeina, I. Strumfa, R. Kleina
*Children`s University Hospital, Dept. of Pathology, Riga, Latvia