G119 Sudden Death in the First Year of Life: The Importance of Pancreatic Histomorphological Analysis

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After attending this presentation, attendees will be able to better understand the importance of pancreatic histomorphological examination after a complete autopsy in sudden infant death, to discriminate between the explained natural sudden death and natural idiopathic sudden death (SIDS).

This presentation will impact the forensic science community by discussing the role of endocrine/metabolic pancreas disease as cause of sudden infant death, providing valid evaluation parameters to the diagnosis.

Material and Method: From 1990 to 2009, 175 autopsies (137 males and 43 females) were performed on infants suddenly dying of natural causes within the first year of life. The diagnosis of sudden natural death has been established through a complete autopsy and investigation including scene examination, review of family, social and medical history and toxicology studies. The age ranged between 2 and 273 days (median 42 days).

In 19 cases, death was unexplained (SIDS) while in 156 cases, it was due to congenital or acquired diseases (explained sudden infant death), primarily involving different systems: cardiovascular (121 cases); respiratory (15 cases); endocrine/ metabolic (12 cases); gastrointestinal (4 cases); central nervous system (4 cases).

Autopsy protocol were based on Perinatal Autopsy Manual. Washington, D.C.: Armed Forces Institute of Pathology, 1983, and on Paediatric Autopsy Techniques - Enid Gilbert-Barness and Diane E. Debich-Spicer in Handbook of Pediatric Autopsy Pathology, Humana Press pp.7-74, 2005. From a histological point of view, according to 2,875 fetal and pediatric autopsies, the following morphological pancreatic parameters were examined: (1) lobular architecture; (2) interstitial thickness; (3) number, branch and volume of ductular-acinar units; (4) number, size and cytology arrangement of islet (quantitative relation between α, β, and δ cells); (5) inflammatory infiltrates; and, (6) heterotopic erythropoiesis.

This analysis was performed on serial sections stained with hematoxylin-eosin, Alcian-PAS, Mallory's trichrome and Giemsa and partly investigated immunohistochemistry using antibodies anti-insulin and anti-glucagon.

Results: In the context of explained sudden natural death in the first year of life, pancreatic histological examination has allowed us to identify 11 cases related to endocrine/metabolic disease, of which, in nine cases, were interested the islets of Langerhans (endocrinous pancreas), and in two cases ductular-acinar units (exocrine pancreas).

The endocrine/metabolic diseases involving endocrinous pancreas were: glycogenosis (type 1b and 2)(five cases); maternal diabetes (2 cases); nesidioblastosis (2 cases (1 case in monochorionic twin)). The endocrine/metabolic diseases involving the exocrine pancreas were: cystic fibrosis (2 cases) macronesia and polynesia were observed in pancreas of both patients with glycogenosis and in children of diabetic mothers, these aspects were due to hyperplasia of the α-cells in patients with glycogenosis, and β-cell hyperplasia in children of diabetic mothers.

In these cases also present were cytoatipism of β-cell and eosinophilic granulocyte infiltration of the islets. In subjects with nesidioblastosis there was only a diffuse polynesia neoformation of islet from duct epithelium.

The cystic ectasia of the ductular-acinar structures associated with pink inspissated secretion was observed in cystic fibrosis.

Discussion and Conclusion: A complete autopsy is essential to establish the causes of sudden explained death in the first year of life. This approach allows to sample, for histomorphological examination, organs such as the pancreas that are almost always free of macroscopically visible changes.

The results of the study show that, in addition to the consolidated sampling of pancreas in autopsy, a complete and focused histomorphological study as suggested, allowing the identification of endocrine-metabolic anomalies, such as glycogenosis, nesidioblastosis, and cystic fibrosis, only rarely reported in the literature as a cause of sudden death in the first year of life.

This research demonstrated that, glucose postmortem levels in plasma and vitreous are not reliable for identifying potential endocrine- metabolic diseases, certainly the histomorphological data of the pancreas is the

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most reliable.

Glycogenosis, Nesidioblastosis, Sudden Infant Death

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