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CONGENITAL ADRENAL CORTICAL NODULAR HYPERPLASIA - CUSHING`S SYNDROME, CASE REPORT

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Introduction: Adrenocortical tumors are very rare in children, 0,3-0,4% of all age dependent tumors. Clinically they may manifest as Cushing`s Syndrome.
Methods: The aim of the report is to describe the rare disorder in a preterm girl from 36 gestational weeks, born by Caesarean Section with severe growth retardation (1054 g). Hypertension (89/64 – 147/97 mmHg) and hyperglycaemia (7-11 mmol/l) from the 1st day of life were present. Other features were :"moon face", hirsutism, thin skin, muscle wasting and weakness. Biochemical tests showed: plasma cortisol level > 1380 nmol/l at 8.00 and 24.00, but normal ACTH level. Serum DHEAS (>27,1 □mol/l), testosterone (41,4 nmol/l), androstendione (223 nmol/l) were all increased. After 48 h dexamethazone supression test (20 □g/kg) plasma cortisol decreased by 35%, 24h urinary free cortisol level by 72% and ACTH from 27,5 to 8,1 pg/ml. Ultrasound scan and MRI showed slightly enlarged adrenals (21x10mm). Neonatal diabetes and pheochromocytoma were excluded. Bilateral adrenectomy at the age of 6 weeks was performed. Histological findings showed bilateral multinodular (up to 5 mm) adrenocortical hyperplasia (5,7 and 4,2 ml). The course of disease was complicated by repeated infectious episodes and multiorgan failure. The baby died at the age of 3 months.
Conclusion: nodular adrenal hyperplasia in neonatal period may present with severe IUGR, hyperglycaemia, hypertension.