83 *D. FLORET, D. ROSENBERG, G. HAGE, P. MONNET. Introduced by R.FRANCOIS. Clinique Médicale Infantile B. Hopital Edouard Herriot. Lyon FRANCE*

Hypothyroidism, diabetes mellitus and congenital rubella syndrome

The congenital rubella syndrome consists of miscellaneous clinical manifestations, some of which relate to the endocrine system. In this report, we describe a case of congenital rubella syndrome with late onset hyperthyroidism and diabetes mellitus. This male patient was born to a mother who developped rubella during the tenth week of gestation ; he presented a typical congenital rubella syndrome with mental retardation, neuro-sensory deafness, hypoplasia of the dental enamel and chorioretinitis.
Hyperthyroidism was diagnosed at the age of 3.10 years and was treated successfully with propylthiouracil during 4 years ; the course was complicated by premature craniosynostosis and a craniectomy was performed at the age of 7 years.
Overt diabetes mellitus developped at 17 years and was well controlled by insulin therapy.
Histocompatibility (HL-A) antigens were A2, B8, B40.
Although thyroid disorders and diabetes mellitus habe been previously described as late complications of the congenital rubella syndrome to our knowledge this is the first case presenting with an association of both endocrinopathies. It is possible that in this particular patient HLA B8 antigens might be responsible for an altered autoimmune function with increased susceptibility to rubella infections.

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Production of human chorionic gonadotropin(HCG)by a virilizing hepatoblastoma.

Malignant tumors with ectopic hormone production are extremely rare in childhood.Only a few cases of virilizing hepatic tumors have been described so far as causes of precocious puberty in boys.We have observed a 13 m old boy presenting with signs of isosexual precocious puberty and a palpable right upper abdominal mass.Preoperative concentrations of alpha-fetoprotein(125 mg/100 ml),HCG(ß-subunit)(73 uIU/ml) and testosterone(340 ng/100ml)were clearly elevated. A hepatic trisegmentectomy was performed; the tumor contained high concentrations of HCG.During the postoperative observation period of 4 months alpha-fetoprotein and HCG remained undetectable, testosterone normalized to 3.4 ng/100 ml.-Conclusions:Determination of ectopic hormones 1) can serve as a diagnostic clue to the presence of (un)suspected malignancy, and 2) may be useful in evaluating therapy and in detecting recurrence.

85 D.AYRAL, P. GILLET, and R. FRANCOIS. Service de Pédiatrie, Hopital Edouard Herriot and U.E.R. Alexis Carrel Lyon. France. Diencephalic syndrome with GH deficiency, hyperprolactinemia and neurovegetative and behavioral disturbances ; a case report.

We report the clinical history of a 13 year old girl with no familial pathology. The onset took place shortly after a non complicated small-pox vaccination (18 mo.). At completion it combined :

1 - An endocrine dysfunction : +5 SD obesity and polyphagia a complete GH deficiency treated on from age 7, a breast hypertrophy with normal estrogens but important hyperprolactinemia, easily blocked by bromocriptine. Other hypophyseal stimulines (ACTH, FSH, LH, TSH, ADH) were normal.

2 - Behaviour disturbances : retirement, multiple obsessions and fears being part of a marked psychotic behaviour causing a secondary deterioration.

3 - Signs of neurovegetative dysautonomia : absent tears, bouts of hyper -or- hypothermia (40° to 33° C), cyclic vomiting divergent squint, acrocyanosis...

This manifold semiology points out to a diencephalic localization. All neuroradiologic investigations failed to find any diencephalic tumour. Further investigations showed an abnormal EEG, diminished REM-sleep, lowered CSF-5HT turn-over under benemide but did not give a satisfactory pathogenic explanation for the child's disease.