HYPOTHALAMIC DISTURBANCES CAN MASK PITUITARY PATHOLOGIES: A CASE OF ADIPSIC CENTRAL DIABETES INSIPIDUS, GROWTH HORMONE DEFICIENCY AND HYPERPROLACTINAEMIA

1R Nichols, 2R Kumar, 3E Crowne, 4C Burren. 1Child Health, Royal Cornwall Hospital, Truro, UK; 2Paediatric Endocrinology, Bristol Royal Hospital for Children, Bristol, UK

Background This six year old girl unusually has central diabetes insipidus without polydipsia; with accompanying growth hormone deficiency and hyperprolactinaemia. She presented with 2 years of worsening breathlessness on minimal exertion, weight gain, poor linear growth and marked sweating in all weathers. Initial assessment revealed short stature (Height SDS -3.0), obesity (BMI 98th centile) and mild dehydration.

Methods Investigations demonstrated hypernatraemia (sodium 168 mmol/L), hyperprolactinaemia (prolactin 3341 mu/L), normal thyroid function and low Insulin-like growth factor-1. MRI pituitary demonstrated no microprolactinoma or other CNS lesion. Extremely low peak GH 0.5 ug/L (normal >7 ug/L) on Insulin tolerance testing indicated growth hormone (GH) deficiency, but normal cortisol. Inpatient assessment demonstrated absence of thirst despite persisting hypernatraemia, high serum osmolality (356mosmol/kg), inappropriately dilute urine (79mosmol/kg) and urine output 4ml/kg/hour. Low serum vasopressin 1.4 pmol/L. Negative tumour markers.

Results GH replacement was extremely effective with improvement in height and normalisation of BMI. Managing her adipsic DI was challenging. Increasing oral intake of 1800 ml/day was insufficient. DDAVP IM achieved urinary concentration (79mosmol/kg), but Oral DDAVP was ineffective, despite high daily dose 1000 ug. Normalisation of serum sodium and urine output was finally achieved with nasal DDAVP, although slight hyperosmolality persists. The patient has continued to experience excessive sweating. Serial MRIs continue to be normal.

Conclusion This unusual case of adipsic diabetes insipidus with excessive sweating is likely to represent hypothalamic dysfunction although no structural lesion is identified on MRI brain. Case series of adipsic diabetes insipidus have identified a high frequency of hypothalamic abnormalities associated with significant morbidity and mortality.

DIFFERENT ASPECT OF PRECOCIOUS PUBERTY: CLINICAL EXPERIENCE AND OUTCOME (8 CASES)

1S El Hilal, 2D Dhahiri, 1H Ajmi, 3S Mabrouk, 1K Kaddi, 1J Chemli, 1N Zouari, 3 Hassayoun, 4K Ttil, 5S Abroug. 1Pediatrics, University Hospital Sahloul, Sousse, Tunisia; 2Radiology, University Hospital Sahloul, Sousse, Tunisia

Background The term of precocious puberty signifies the onset of secondary sexual characters before the age of 9 years in the boys and 8 years in the girls. The first step in the clinical reflexion is to decide whether the child has central or peripheral form of precocious puberty.

Objective Identify some causes of precocious puberty, expose difficulty of management.

Methods A retrospective study of medical records of children hospitalised in the Paediatric unit at Sahloul hospital in Tunisia. Within a period of 6 years, eight patients were diagnosed with precocious puberty (4 boys and 4 girls). The mean age was 4 years and 3 months. The precocious puberty was isosexual in all cases with accelerated growth velocity. The PP was identified as central in four cases, the investigations showed: hypothalamic hamartoma (2 cases), pituitary adenoma (1 case) and arachnoid cyst (1 case). Peripheral precocious puberty included: congenital adrenal hyperplasia (3 cases) and adrenal cortical carcinoma (1 case). The children with central precocious puberty were treated with gonadotrophin releasing hormone agonists, those with congenital adrenal hyperplasia received steroids replacement therapy and surgery was performed for the boy with adrenocortical carcinoma. The clinical and biological evolution of our patients was favourable with a regression of signs of puberty and normal hormone levels after a mean of 5 years.

Conclusion Precocious puberty needs a systematic approach with detailed history and clinical examination. Early treatment contributes to optimise adult height potential. There are still unresolved questions that future studies will need to address. Finally, the behavioural and psychological outcomes of precocious puberty were still poorly evaluated.