SEVERE HYPERNATREMIA REVEALING A ROHHAD-NET SYNDROME.

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BACKGROUND

R	Rapid-Onset
O	Obesity
H	Hypoventilation
H	Hypothalamic dysfunction
AD	Autonomic dysregulation
NET	NeuroEndocrine Tumors

Rapid-onset Obesity with Hypoventilation, Hypothalamic dysfunction and Autonomic Dysregulation (ROHHAD) first described by Ize-Ludlow then recently named ROHHAD-NeuroEndocrine Tumors (ROHHADNET) is a rare cause of obesity in children. The diagnosis is extremely challenging as there is no single confirmatory diagnostic test. Mortality rate can go up to 50 to 60 % due to cardiorespiratory arrest, therefor early diagnosis may minimize mortality

CASE REPORT

We report a case of a six years old boy, referred to our clinic for hypernatremia.
Six months ago, he started to present episodes of acute respiratory distress diagnosed as asthma. Few months later and following an acute respiratory distress, he was admitted for coma due to a severe hypernatremia reaching 200 meq/l complicated by renal failure. He was successfully managed and left the intensive care unit with a normal electrolyte balance and a normal renal function. Parents reported progressive weight gain without polyphagia. ROHHAD-NET Syndrome was suspected, Table 1 summarize the clinical features and investigations leadimg to the diagnosis.
Fluid balance is well controlled with oral hydration and low sodium diet. Obesity is managed by food dietary alone since exercising remains limited by the respiratory distress episodes.

Rapid Onset Obesity	**BMI=23 (> 97th Centile)**
Autonomic dysregulation Excessive sweating Cold hands and feet Raynaud phenomenon	
Respiratory manifestations Recurrent respiratory distress Obstructive Sleeping Apnea Alveolar hypoventilation	
Hypothalamic-pituitary disorders Hypernatremia HyperProlactinemia FT4 TSH IgF1 8AM Cortisol	200 meq/l 91.4 ng/ml (3.7-17.9) 10.45 pmol/l (10-17.1) 3.5 µUI/ml (0.6-4.84) 53.78ng/ml(57.7-434)
NeuroEndocrine Tumors Thoraco-abdominal CT Scan VMA SDHEA Δ4 Androstenedione Testosterone	Enlargement and calcifications of the right adrenal (Fig.1) 3.98µmol/mmol (N<10) 0.28µg/ml (0.24-2.1) 0.3 ng/ml (0.01-1.31) <0.05 ng/ml (0.39-2.01)
Megaloblastic anemia	**Hb: 11g/dl, MCV: 110 fl**

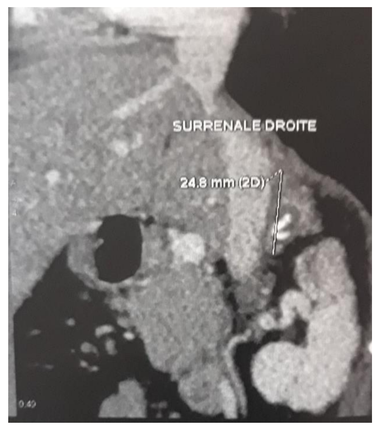

Fig 1: Right adrenal calcifications

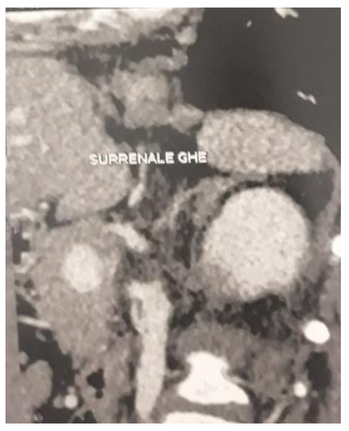

Fig 2: Left adrenal

DISCUSSION

Hypernatremia is the life thretening symptom in our patient. It was present in all 6 patients described by Bougnère (1) and 7/15 patients in Ize-Ludlow 's serie (2)

Ganglioneuroma suspected upon adrenal calcifications needs a multidisciplinary team discussion in order to indicate adrenal resection.

respiratory manifestations are associated with a high mortality and need specific management.

CONCLUSION

Our patient management requires a multidisciplinary team collaboration and his prognosis relies on the severe hypernatremia episodes, the sleep apnoea disorder and the development of neuroendocrine tumours.

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