Case report :

Severe hyponatraemia after plastic surgery in a girl with cleft palate, medial facial hypoplasia and growth retardation.

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SUMMARY

A 10 year-old girl with middle face hypoplasia, cleft lip and palate developed severe hyponatraemia on the first day following surgery. Final diagnosis was inappropriate secretion of antidiuretic hormone (ADH) whereas complete hormonal investigation revealed partial deficit in growth hormone secretion. The incidence of hormonal deficiency associated to midline facial malformations is discussed.

Hyponatraemia is one of the most common postoperative electrolyte disturbance which may be due to several causes. This case report describes the difficulties in assessing the exact cause of severe postoperative hyponatraemia in a girl with middle face hypoplasia, cleft lip and palate.

Case report

A 10 year-old girl, weighing 30 kg, was scheduled for maxillary reconstruction with bone grafting to correct superior maxillary hypoplasia. She had middle face hypoplasia, cleft lip and palate, and she underwent three reconstructive surgeries at the age of 3 months, 6 months and 5 years. She had also growth retardation (- 2.5 SD) but brain ultrasonography performed in the neonatal period did not revealed any medial cerebral abnormalities. She was taking no medication preoperatively. No laboratory testing was ordered during the preoperative examination as she had three previous surgeries in the same hospital and coagulation tests were normal on two occasions previously. Surgery lasted 4 hours and was uneventful. Anaesthesia was induced with halothane and endotracheal intubation was facilitated with fentanyl and atracurium. During surgery, 750 ml of lactated Ringer with 1% dextrose were administered to compensate for fasting deficit (8 hours) and to provide for maintenance hourly fluid therapy. Blood loss was minimal during surgery (less than 5% of total blood volume). The trachea was extubated at the end of surgery and the girl returned to surgical wards two hours later. An IV line was left in place for standard fluid therapy (2 l/24h of D5 in 0.33 NS). Amoxicilline and clavulanic acid were continued for 48 hours. Postoperative pain relief was achieved with i.v. propacetamol (900 mg q6h) and non steroidal anti-inflammatory drugs (niflumic acid 400mg q6h).

The following day, she became confused and developed headache, vomiting, without fever. Laboratory testing evidenced severe hyponatraemia (117 $mmol.l^{-1}$), hypochloraemia (83 $mmol.l^{-1}$), whereas BUN were within normal ranges (creatinine 38 $mmol.l^{-1}$, urea 1.7 $mmol.l^{-1}$). Serum potassium was normal (4.6 $mmol.l^{-1}$), as well as total protein (73 $g.l^{-1}$). Initial treatment consisted in sodium supplementation (10 $mmol.kg^{-1}.d^{-1}$). On the second day, clinical improvement was noticed, but plasma sodium concentration was still low (120 $mmol.l^{-1}$). Urinary output was 800 ml for the last 12 hours. Urinary sodium was high (80 $mmol.l^{-1}$) and urinary potassium was 32 $mmol.l^{-1}$. Plasma osmolality was 255 $mosmol.kg^{-1}$, whereas urinary osmolality was 342 $mosmol.kg^{-1}$ and urinary density 1.011. Three hypothesis were proposed to explain such severe postoperative hyponatraemia : 1) dilutional hyponatraemia due to infusion; 2) pituitary insufficiency and 3) inappropriate secretion of antidiuretic hormone (ADH). No overinfusion was evident by reviewing the patient's chart. In order to evaluate a possible hormonal deficit, blood was taken for hormonal dosages before starting i.v. hydrocortisone. This treatment led to a dramatic improvement of plasma sodium values which rose to 132 $mmol.l^{-1}$ on day 2 and 142 $mmol.l^{-1}$ on day 4. Blood urea, creatinine and total protein remained unchanged. Hormonal investigations revealed normal values for ACTH, cortisol, thyroid hormones, and IGF_1. Dynamic testing showed a partial growth hormone (GH) deficiency after two stimulation tests (ornithin and arginin-insulin) without dysfunction of the cortisol axis. The brain magnetic resonance imaging was normal. The antidiuretic hormone (ADH) level was within the normal range (5 $pg.ml^{-1}$, mean of the series 4.1 $pg.ml^{-1}$), but inappropriately high in relation to the hypoosmolality of body fluids. This strongly suggests that hyponatraemia was due to an inappropriate secretion of ADH. The diagnosis of inappropriate secretion of ADH is also supported by severe hyponatraemia, associated to normokaliemia, hypochloraemia, preservation of potassium and sodium excretion, and urinary to plasma

osmolality ratio greater than 1. Plasma sodium concentration was regularly followed for the next six months and remained normal throughout this period.

Comment

This report describes severe unexpected postoperative hyponatraemia due to an inappropriate secretion of ADH in a 10 year old girl who presented a cleft lip and palate associated with undiagnosed growth hormone deficit without cerebral morphological abnormality.

Our patient had complex midfacial anomaly combining hypoplasia of middle face and medial cleft. Malformations of this type are frequently associated with cerebral malformations and pituitary deficiencies [1-8]. Transfontanellar ultrasound is usually performed in the neonatal period in those patients in order to assess the presence or not of cerebral abnormalities. A defect in the number of the neural crest cells during embryogenesis produces a defect in forebrain, and in bony and skin structures of the midface [9]. Development of those neighbouring structures takes place between the 4th to the 8th weeks of gestation [1,3,9]. The earlier the malformation is induced, the more serious it is. Inducers are alcohol, toxic, diabetes, viral infections and genetic causes.

Growth hormone deficit is frequently observed in patients with cleft lip and palate [7,10,11]. The prevalence of GH deficit in those patients is estimated between 4 and 10% [10,11], whereas that of extremities malformations is 29%, that of cardiac malformations 15% and that of other facial malformations is 13% [12]. The overall incidence of associated malformations is three fold greater in patients with clefts than in the general population

The diagnosis of inappropriate secretion of ADH was unexpected in this particular case. Indeed, diabetes insipidus is more likely associated to midline facial malformation than paradoxical increased ADH secretion. However the association of intermittent inappropriate secretion of ADH and diabetes insipidus had been reported by Hasegawa and coll. [13] in a patient with holoprosencephaly. Moreover, inappropriate secretion of ADH due to chronic reset of osmoreceptor response had been described in children with agenesis of corpus callosum [14]. No cerebral anomaly and no chronic hyponatraemia were present in our patient. However, failure to maximally suppress ADH when plasma osmolality falls to the normal « threshold » level (270 mosmol.kg^{-1}) supports the hypothesis of fluctuent default in hypothalamic osmoreceptors which may occur in association of a variety of diseases [15]. This could be due to a defect in neurosensory cells or to a selective loss of inhibitory input from the osmoreceptor. Anaesthesia and surgery are usually described among the classical aetiologies of inappropriate secretion of ADH. Although most of the cases have been reported following major surgical procedures, inappropriate secretion of ADH has also been reported after minor surgery [16]. The implication of anaesthetic drugs is not well supported. Drugs such as diclofenac may however induce an inappropriate secretion of ADH [17]. In our case, the association of a triggering event (anaesthesia and surgery) together with a hypothalamic osmoreceptor dysregulation will likely explain the occurrence of inappropriate secretion of ADH.

In conclusion, inappropriate secretion of ADH may occur in every surgical patient. This case report stresses the frequency of hypothalamic dysfunction in children with midface malformations and will encourage the anaesthetists to monitor plasma sodium concentrations after surgery in those children. These patients should be managed by experienced paediatric anaesthetists.

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CEREBRAL OEDEMA: SWELLING OF THE BRAIN

HYPONATRAEMIA: LOW SODIUM (NORMAL RANGE 135-145mmol/l)

RENAL FAILURE: FAILURE OF THE KIDNEYS

OBSTRUCTIVE UROPATHY: OBSTRUCTION TO THE OUTFLOW FROM KIDNEYS

CONGENITAL: BORN WITH

POLYURIA: PRODUCTION OF LARGE VOLUMES OF DILUTE URINE

URETERS: CONNECTING KIDNEYS TO BLADDER

FUNDOPLICATION: OPERATION TO STOMACH

GASTRO-OESPHAGEAL REFLUX: STOMACH CONTENTS REGURGITATING INTO THE GULLET (oesophagus)

ORCHIDOPLEXY: OPERATION TO FREE UNDESCENDED TESTICLE

PERITONEAL DIALYSIS: THE REMOVAL OF WASTE AND TOXIC PRODUCTS FROM BLOOD BY THE USE OF THE PERITONEUM AS A SEMIPERMEABLE MEMBRANE

GASTROSTOMY: ARTIFICIAL HOLE IN STOMACH CREATED BY SURGERY

CVP: CENTRAL VENOUS PRESSURE

CT SCAN: COMPUTERISED AXIAL TOMOGRAPHY

PULMONARY OEDEMA: FLUID ON THE LUNGS

EPIDURAL: INJECTION OF ANALGESIC INTO THE SPACE AROUND THE SPINAL CORD

LUMBAR: LOWER BACK

ELECTROLYTES: IN THE MAIN SODIUM AND POTASSIUM

A.D.H.: ANTI-DIURETIC HORMONE

ALDOSTERONE:	HORMONE CONTROLLING SODIUM BALANCE
HYPOXIA:	DIMINISHED AMOUNT OF OXYGEN IN TISSUES
HAEMORRHAGE:	BLEEDING
ISOTONIC:	SAME CONCENTRATION AS PLASMA i.e.. normal saline is isotonic with plasma
HYPOTONIC:	A SOLUTION HAVING A LOWER OSMOTIC PRESSURE THAN ANOTHER ONE
CEREBRAL PERFUSION:	THE PASSAGE OF BLOOD THROUGH THE BRAIN

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