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RESEARCH ARTICLE

ISOLATED INSIPIDUS DIABETES: AN UNUSUAL PATTERN REVEALING A PRIMARY EMPTY SELLA SYNDROME (CASE REPORT)

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ABSTRACT

The empty sella syndrome (ESS) or otherwise called "arachnoidocele" is a relatively rare syndrome, it occurs more frequently in young women, but hormonal dysfunction seems to occur more in men. There are two types of ESS: the primary and the secondary which is more frequent. ESS can be completely asymptomatic and discovered incidentally on imaging, or in the presence of endocrine, neurological or ophthalmological signs. Central diabetes insipidus (CDI) is a rare manifestation of ESS, it is usually associated with other pituitary disorders and may be due to compression of the pituitary stalk and/or post-pituitary gland. The treatment of ESS is symptomatic and relies on the administration of hormone replacement therapy in the case of hormone deficiency. Surgery is reserved for special situations.

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INTRODUCTION

Empty sella syndrome (ESS), is a condition in which the subarachnoid space herniates into the sella turcica, resulting in compression and flattening of the pituitary gland with stretching of the pituitary stalk (1).Central diabetes insipidus is a rare manifestation of ESS (2). We report a patient admitted for etiological investigation of a polyuropolydipsic syndrome and whose investigations concluded to isolated central diabetes insipidus on arachnoidocele. Objective of the work: describe the epidemiological, clinical, paraclinical and therapeutic aspects of empty sella syndrome.

OBSERVATION: Patient aged 39 years, with no notable history, admitted for a severe polyuro-polydispsic syndrome quantified at 8 litres per day with 3 nocturnal awakenings. The clinical examination revealed a patient of normal build, normotensive and not dehydrated. The biological assessment showed a plasma osmolarity of 298 mosm/l, a low urinary osmolarity of 125 mosm/l and a hypernatremia of 145 meq/l, contraindicating a water restriction test. A therapeutic test with Desmopressin was used, at the end of the test there was a decrease in the polyuropolydipsic syndrome and an increase in urine osmolarity. A pituitary MRI was done showing an enlarged empty sella turcica with loss of T1 hypersignal and a thinned pituitary gland flattened against the inferior wall (figure 1,2). The hormonal analysis showed the integrity of the other pituitary axes, the ophtalmological examination didn't find any anomaly. The diagnosis of isolated central diabetes insipidus on empty sella turcica was restrained and the patient was put on hormone replacement with Desmopressin.

DISCUSSION

The empty sella turcica syndrome is considered a rare entity. Its incidence increases between the fourth and sixth decade (1). Although more common in women (3), hormonal dysfunction is more frequent in mens (1). The term ESS is inappropriate: arachnoidocele seems to be more appropriate, since in reality the sella turcica is not empty, but rather filled with cerebrospinal fluid (CSF) (2).

There are two types of ESS; primary and secondary (3,4):
Primary ESS can be caused by intracranial hypertension and/or an abnormality of the sellar diaphragm in subjects without a pituitary history.

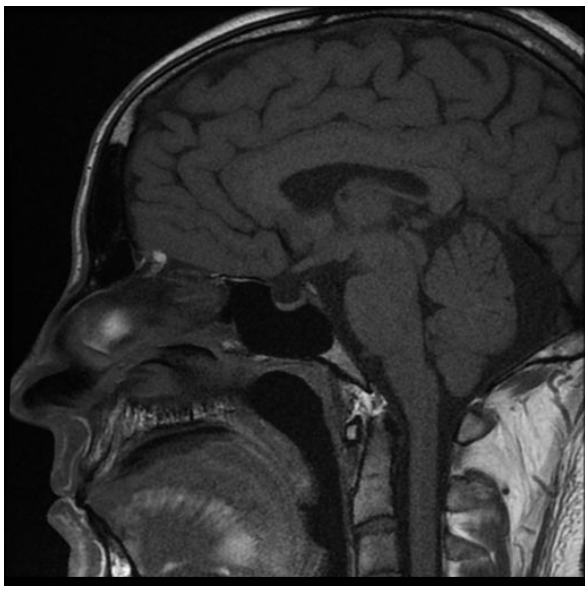

Figure N1: Sagittal section without injection showing the absence of T1 hypersignal of the post-pituitary

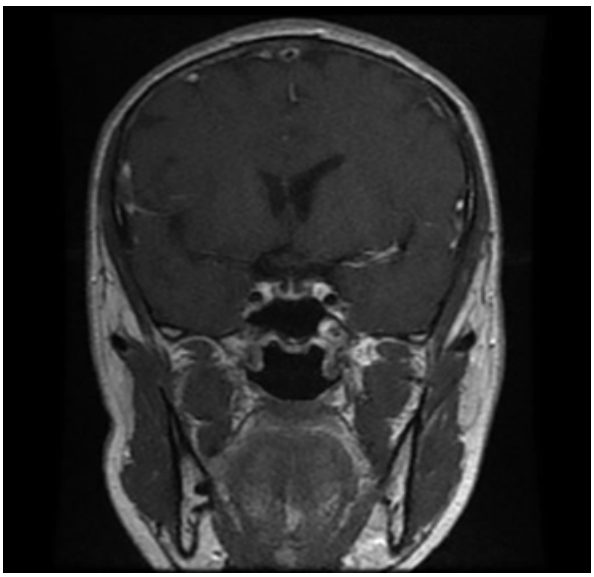

Figure N2. T1 coronal section with gadolinium injection showing enlargement of the sella turcica with thinning of the antehypophysis

In primary ESS, a deviation of the dura mater, which separates the suprasellar cistern from the sella, leads to unobstructed pulsatile movement of CSF into the chiasmatic cistern, causing the pituitary gland to flatten on the sellar floor. In extreme cases, bony erosion of the floor and CSF leakage (rhinorrhoea) may occur, increasing the risk of meningitis (3). Classic risk factors for primary EES are: female gender (F/M ratio is 5/1), multiple pregnancies, obesity, sleep apnoea, high blood pressure and middle age (4). Secondary ESS is more common. Its diagnosis is easy to establish when an underlying pituitary pathology is known. It may be secondary to: various pathological processes of the sellar region, apoplexy of a pituitary adenoma, postpartum pituitary necrosis, pituitary infections, iatrogenic (surgical, post-radiotherapy), late stages of hypophysitis and traumatic brain injury resulting in pituitary atrophy (3). Clinically, ESS may be completely asymptomatic and discovered fortuitously on imaging, or it may be discovered following endocrine, neurological, ophthalmological or psychiatric manifestations (5). Hormonal deficiency can range from panhypopituitarism to hypopituitarism or isolated hormonal deficiency with normal or elevated prolactin values. Somatotropic insufficiency is the most common pituitary deficiency in adults, occurring in 4-57.1% of patients. Corticotropic, thyroid and gonadal insufficiency vary between 2.3 and 32% depending on the series. Hyperprolactinemia is documented in 7 to 10% of patients. However, antidiuretic hormone deficiency is rarer, described in about 1% of cases. Pituitary hypersecretion is rarely associated with ESS and is thought to be secondary to ectopic secretions of GH and ACTH. Our patient has an isolated anti-diuretic hormone deficiency (4). The neurological signs of arachnoidocele are mainly headaches which are present in about 84-88% of patients, these headaches are typically described as lateral, persistent and lasting for several years. In a smaller percentage (about 20% of cases), headaches are accompanied by symptoms of intracranial hypertension such as papilledema and visual disturbances. Other neurological symptoms such as dizziness, syncope, cranial nerve disorders, convulsions or depression may be seen (4). The ophthalmological picture may include decreased visual acuity (37%), blurred vision (29%), diplopia (2%), oculomotor nerve defect (1%) and optic neuritis (1%) (4). Radiologically, the presentation of ESS on MRI is typical: enlargement of the sella turcica and flattening of the residual pituitary against the sellar floor. Finally, MRI will identify the infundibulum sign by showing a pituitary stalk crossing the sellar space, which is a crucial element in ruling out a cystic lesion (main differential diagnosis) (3,6).

Treatment of ESS is symptomatic: in patients with idiopathic increased intracranial pressure, osmotic diuretics or acetazolamide may be prescribed (7). Weight loss appears to be effective in obese or overweight patients, especially if accompanied by sleep apnoea. Symptomatic hyperprolactinaemia should be treated with dopamine agonists. In case of pituitary deficiency, hormone replacement should be prescribed depending on the area affected (3,2). Surgery should be recommended in cases of: severe symptoms of intracranial hypertension, severe headache, visual alterations and rhinorrhea (5). There is no way to prevent this condition, but clinical, hormonal and ophthalmological assessment at diagnosis is strongly recommended (7). However, due to the theoretical risk of progression (although minimal), regular endocrine and neuro-ophthalmological follow-up is necessary for asymptomatic patients (1).

CONCLUSION

ESS or arachnoidocele is a rare entity, may be primary or secondary. It is usually asymptomatic, but may be discovered by neurological, ophthalmological or endocrine signs. Nevertheless, central diabetes insipidus is rarely revealing of the disease, it is exceptionally isolated and often associated with other hormonal deficits. Our observation illustrates the rarity of this mode of revelation, since our patient presents an isolated CID with integrity of the other axes. Conflict of interest statement : The authors do not have any financial, real or perceived conflict of interest in the publication of this manuscript.

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