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Sellar Arachnoidocele: An Unusual Evolution in a Case Report

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Abstract

The sellar archnoidocele is a herniation of the subarachnoid space within the sella turcica which is often associated with a variable degree of flattening of the pituitary. Its clinical presentations range from an asymptomatic radiological finding to endocrine and neuro-ophthalmological manifestations. Its management depends on the initial symptomatology and requires long-term follow-up. We report here the case of a young child followed for a statural delay on a GH deficiency secondary to a sellar arachnoidocele, and who presents a rather unusual evolution.

Keywords

Sellar Arachnoidocele, GH Deficiency, Growth Hormone, Prognosis

1. Introduction

Recently, the incidence of sellar arachnoidocele or so-called empty sella turcica has been increasing after improvements in neuroradiological imaging techniques. Overall, estimates range from 2% to 20% [1]. This incidence remains lower in the pediatric population compared to adults.

The etiopathogeny remains poorly understood, despite the multitude of hypotheses suggested. Generally, several factors may be involved: a congenitally missing (deficient) sellar diaphragm, the involvement of Upper-sellar factors (such as CSF pulsatility, stable or intermittent increase in intracranial pressure), and the occurrence of pituitary factors (such as changes in pituitary volume) [2].

Generally, the asymptomatic form is the most common clinical form in adults. However, in children, an empty sella turcica is frequently associated with hypothalamic-pituitary dysfunction, as well as neuro-ophthalmological manifestations [3].

Management is based on an endocrine and ophthalmologic evaluation, which will help guide subsequent therapy, which is based on replacement therapy for the various deficits, and sometimes surgery in specified indications [3].

The main objective is to focus on the management of intra sellar arachnoidocele and its evolution through a clinical case of a child followed for a GH deficiency due to an intra sellar arachnoidocele.

2. Report Case

Patient currently 17 years old, with no specific pathological history, including no history of trauma, surgery or cranial radiotherapy, who consulted at the age of 13 years for a severe statural delay at −3 SD. The clinical examination didn't note any abnormalities, the size was 138 cm (−3 SD), the weight was 38 kg (−1 SD), and the BMI was 19.9 kg/m^2. Genital examination revealed Tanner stage II testicles, Tanner stage II pubic hair, and no micropenis, the rest of the somatic examination was normal. The first-line assessment of the statural delay was without abnormalities except for low IGF1 at 86.6 ng/ml (119 - 511) (Z-score: −4.1 DS). The rest of the hormonal evaluation: Cortisol at 8 h: 14.9 ug/dl, Testosterone: 1.11 ng/ml (0.2 - 2), FSH: 1.53 mUI/ml (0.5 - 3), LH: 0.97 mUI/ml (0.5 - 3), LT4: 0.98 ng/l (0.70 - 1.48), Prolactin: 5.69 ng/ml (2.58 - 18.12). The bone age is estimated at 11 years. We completed by 2 stimulation tests, the combined Glucagon/Propanolol test showed a GH peak at 6.02 ng/ml, and the insulin hypoglycemia test showed a GH peak at 5.89 ng/ml. The diagnosis of GH deficiency was confirmed. We completed the explorations by a hypothlamo-hypophyseal MRI which demonstrated an intra sellar arachnoidocele with a laminated pituitary parenchyma (**Figure 1**). The ophthalmological assessment reveals an inferior temporal scotoma of the left eye (**Figure 2**). Management consisted of growth hormone replacement therapy, with clinical, biological and radiological monitoring. Over a period of 4 years, the patient was able to reach his target size with an estimated total statural gain of 33 cm (**Figure 3**). Control hypothalamic-pituitary MRI showed a reduction in the size of the arachnoidocele with a larger volume pituitary parenchyma compared to the initial imaging (**Figure 4**).

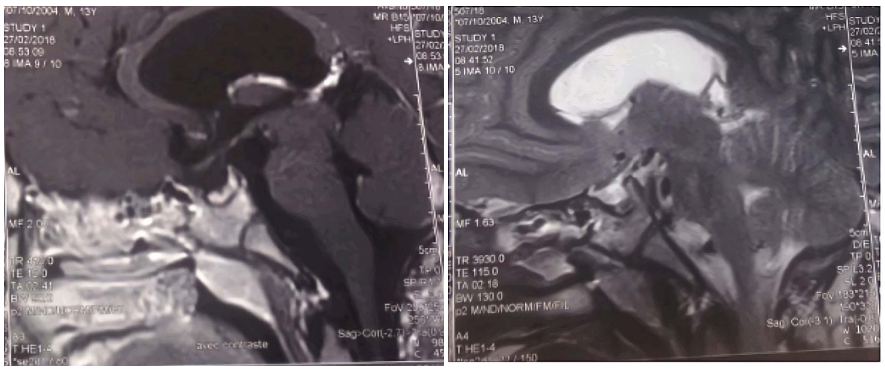

Figure 1. Hypothalamic-pituitary MRI, sagittal section in T1 and T2 weighted showing an intra sellar arachnoidocele with a laminated pituitary parenchyma.

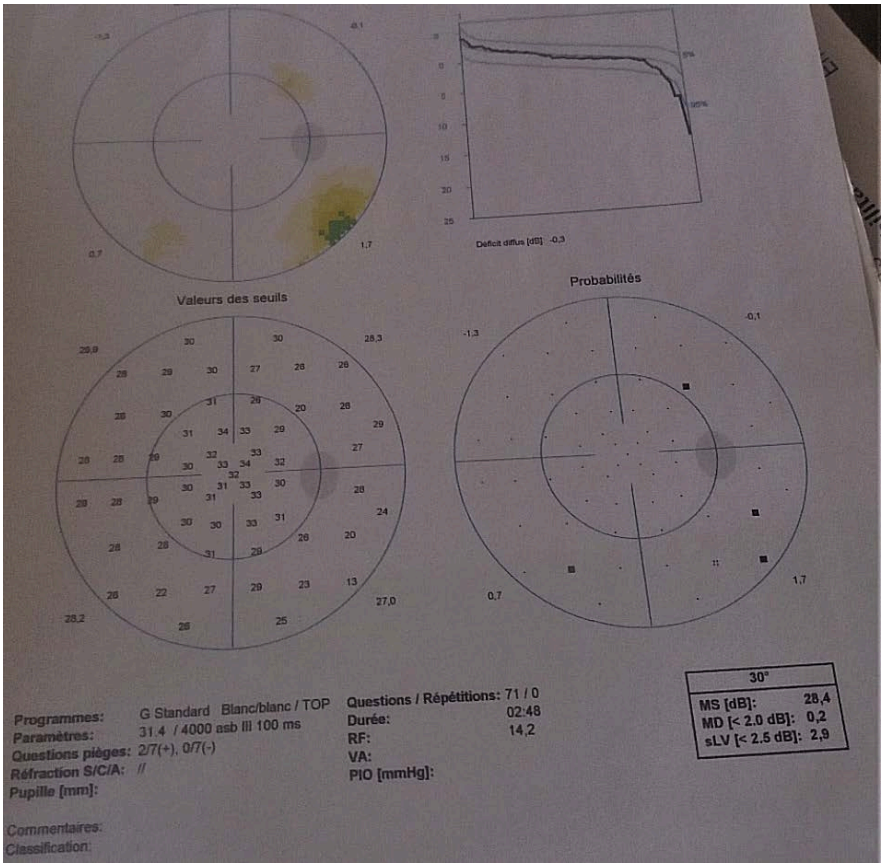

Figure 2. Goldmann visual field showing an infero-temporal scotoma of the left eye.

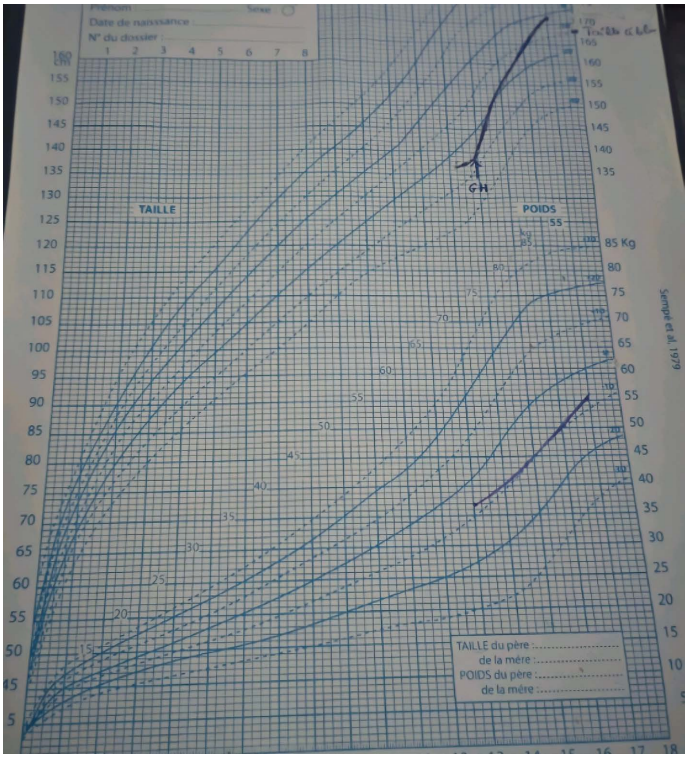

Figure 3. The Patient's growth curve.

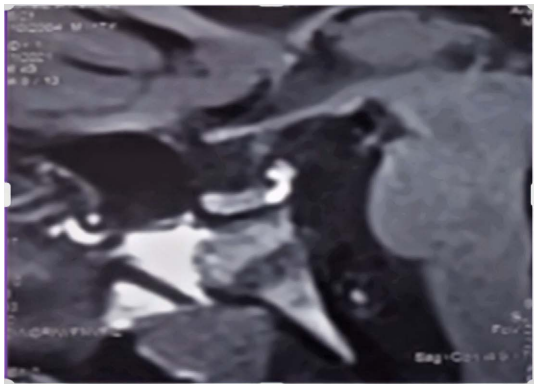

Figure 4. Control hypothalamic-pituitary MRI showing a reduction in the size of the arachnoidocele and a paradoxical increase in pituitary parenchyma.

3. Discussion

Primary empty sella turcica is characterized by the absence of any history of pathology, trauma, surgery or radiotherapy of the hypothalamic-pituitary region [4], as reported in our case. In most cases, the finding of an empty sella turcica may be incidental on radiological imaging without any clinical implications. However, when it is associated with clinical manifestations such as endocrine, neurological and ophthalmological, it is referred to as empty sella turcica syndrome [5]. Endocrine abnormalities are documented in around 19% of patients [6]. Anteropituitary insufficiency can be explained by the compression of the pituitary gland against the sellar cavity walls, associated with a posterior stretch of the pituitary stalk. The insufficiency can be of variable degree, ranging from a simple isolated hormonal deficit to a true panhypopituitarism. GH deficiency is the most common hormone deficiency in adults as well as in the pediatric population. According to the different series, this frequency varies between 4% and 57.1% [7] [8] [9]. In our case, the hormonal evaluation showed an isolated GH deficit. The mechanism responsible is still unclear, however, it is well established that GH is the most vulnerable pituitary hormone and its secretion is early disturbed in case of pituitary gland damage. This can be explained by the fact that somatotropic cells occupy almost 50% of the normal volume of the pituitary gland. In addition, the mechanisms of neuroregulation of GH secretion can be significantly influenced by the increase in intrasellar pressure [9].

The frequency of visual disorders varies according to the series but seems to be visual field alteration (bitemporal hemianopia or even superior bitemporal-quadranopsia) is the most frequent symptom; papilledema and optic atrophy are more exceptional [10]. Our patient had an inferior temporal scotoma of the left eye, and the rest of the ophthalmologic examination was normal.

In case of symptomatic empty sella turcica, the management consists of hormonal treatment of pituitary deficits systematically. Surgical treatment is reserved for patients with worsening visual alterations requiring peritoneal-ventricular surgery, in order to avoid serious complications of intracranial hypertension. On the other hand, in asymptomatic patients, simple endocrine, neuro-ophthalmo-

logical and regular radiological monitoring is recommended in the literature [5].

In our case, the management consisted of growth hormone replacement therapy at a dose of 0.025 mg/kg/d with clinical, biological and radiological monitoring. The evolution was good with an estimated statural gain of 33 cm over 4 years, and the patient reached his target size. Radiologically, a reduction in the size of the arachnoidocele was noted with an increase in the volume of the pituitary parenchyma compared to the initial imaging. The mechanism explaining the reduction in size of the arachnoidocele remains to be studied.

4. Conclusions

Empty sella turcica is a very heterogeneous condition: ranging from an occasional neuroradiological finding without any clinical involvement, to empty sella syndrome, characterized by the occurrence of endocrine, ophthalmological and neurological symptoms or a combination of these. Its management must be modulated according to the clinical context.

A multidisciplinary approach is strongly recommended with the integration of endocrine, neurological and ophthalmological experts.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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