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(CASE REPORT)

Blindness in adolescents due to craniopharyngioma: A case report

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Abstract

Craniopharyngioma is a benign tumor in children that can lead to blindness often irreversible. Its diagnosis is essentially radiological but its management is multidisciplinary. We report the case of bilateral blindness caused by a craniopharyngioma in an 18-year-old adolescent.

Keywords: Craniopharyngioma; Blindness; Adolescents; Panhypopituitarism

1. Introduction

Craniopharyngioma is a rare benign epithelial tumor originating in the pituitary gland or pituitary gland and arising in the sellar and suprasellar region. Its incidence is 0.5 to 2 new cases per year and per million inhabitants. The clinical picture may include endocrine disorders suggestive of hypopituitarism, ophthalmologic disorders, signs of intracranial hypertension and related focal neurologic signs. Its treatment is surgical, its management should not be delayed at the risk of deteriorating the quality of life of the patient. We report the observation of a case.

2. Observation

An 18-year-old teenager, who has presented for 10 years with progressive temporal headaches with intermittent vomiting. He consulted different doctors and on several occasions with an initial cerebral scan showing a sellar lesion process. However, surgical treatment was unfortunately not performed.

Faced with worsening symptoms, in particular intense headaches and visual disturbances, initially predominantly on the left and then evolving towards bilateral blindness, the patient consulted us 8 years later.

Examination revealed: delayed stature and puberty with a height of 151 cm (-3-4DS), a Tanner G1P1 and a 4 cm penis, bilateral blindness.

Biological workup showed: prolactin: 0.37 ng/ml; testosterone: 0.03 ng/ml, FSH 0.5 IU/L, LH 0.5 IU/L, low cortisol, TSH: 0.16 IU/L, FT4: 14.7 pmol/L.

A brain MRI was performed showing a lesional process in the suprasellar region measuring 34x27x21mm (Figure 1).

The visual field was impossible in both eyes: no light perception.

The patient was operated with a morphological aspect compatible with a craniopharyngioma on histological examination.

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Moreover, the patient also has a panhypopituitarism for which he is under substitution.

The evolution at 6 months was marked by a recurrence with MRI: sellar and suprasellar lesion process with haemorrhagic changes measuring 27x22x19 mm. The patient was operated on again. However, his quality of life was impaired due to bilateral blindness.

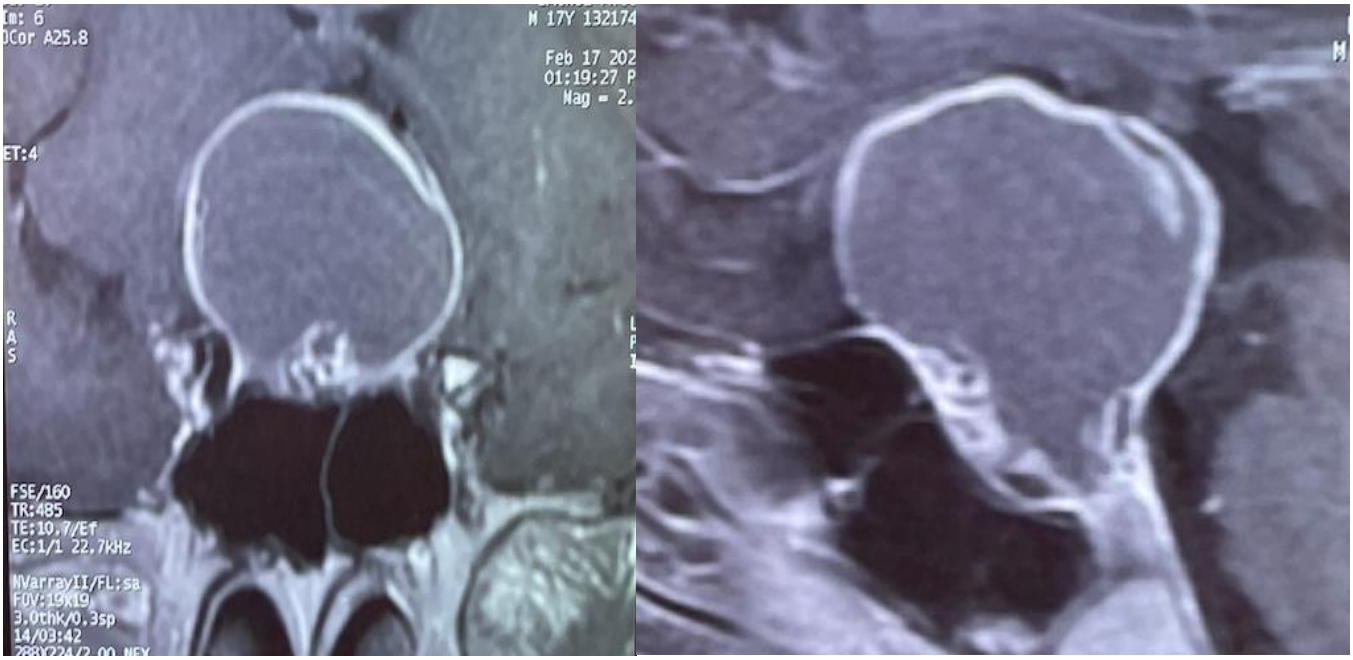

Figure 1 Sagittal and Coronal hypothalamic-pituitary MRI showing a sellar tumor process measuring 34x27x21mm

3. Discussion

Craniopharyngiomas are benign epithelial tumors arising from the pituitary gland or pituitary gland in the sellar and/or parasellar region [1]. Affecting mostly children, they represent 10% of all intracranial tumors [2].

The management of craniopharyngiomas in children is multidisciplinary and increasingly uncontroversial, but remains a surgical challenge[3]. It is, however, a major surgery due to the sensitive anatomical relationships.

The natural evolution of a craniopharyngioma can be towards a chiasmatic syndrome that can lead to bilateral blindness, thus deteriorating the quality of life [4].

Visual disturbances are very common and may be related to optic tract compression and/or secondary to chronic intracranial hypertension. In children, the first sign is visual in 30% of cases. Amblyopia is often very severe at the first examination, since visual acuity is already zero on one side in one child out of five. Even in countries with a high medical density, there is still a rate of 5 to 20% of children who are almost blind at the time of diagnosis [5].

Despite the benignity of these lesions, these tumors are accompanied by significant morbidity, related to the development of the tumor itself and/or its treatment. Recurrence is the main evolutionary risk of this disease [6].

Extensive tumor removal should be attempted in case of localized tumor, but there are increasing arguments for partial surgery, respecting the hypothalamus, associated or not with radiotherapy, in case of invasion of the hypothalamus by the craniopharyngioma [7]. Hormone replacement therapy may also be necessary in cases of endocrine damage [8].

4. Conclusion

Craniopharyngioma is a rare benign tumor that occurs in children more than in adults and can lead to serious visual complications, including bilateral blindness that is often irreversible. Management of this tumor should be

multidisciplinary, with regular ophthalmologic surveillance for visual deterioration. Surgical resection is the first-line treatment and may be followed by radiation therapy and hormone replacement therapy.

Compliance with ethical standards

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I thank my teachers for their help in developing this work.

Conflict of interest statement

I declare no conflict of interest.

Statement of ethical approval

The present research work does not contain any studies performed on animals/humans subjects by any of the authors.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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