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RESEARCH ARTICLE

CRANIOPHARYNGIOMA REVEALED BY PAPILLEDEMA IN A CHILD: A CASE REPORT

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Abstract

Craniopharyngiomas are rare epithelial tumors, arising from the pituitary gland or pituitary gland and developing in the sellar and supra sellar region. We present an unusual case of a craniopharyngioma in a 9-year-old child reveled by a bilateral papilledema, with strabismus and extraocular neurological signs. The diagnosis of this tumor is often late and is based on the triad of neurological, ophthalmological and endocrine signs. Their treatment is multidisciplinary and includes surgical and hormonal treatment. Their prognosis depends on the precocity of the diagnosis, the location and the size of the tumor. Our aim is to describe a clinical case with bilateral papilledema revealing a craniopharyngioma in a child.

Introduction:-

Craniopharyngioma in children is a rare benign embryonic epithelial tumor that affects both children and adults [1], with an estimated 0.5 to 2 new cases per year and per million inhabitants [2], Their treatment is based on whether or not surgery is followed by a hormonal treatment. [3]

Case presentation:

We report the case of a nine-year-old child, without any notable pathological history, who presented to the ophthalmologic emergency room for a bilateral visual acuity decrease with a divergent strabismus installed over two weeks, associated with a paresis of the right lower limb, a urinary leakage with vomiting and an alteration of the general state preceded two months earlier by intense and rebellious headaches.

The ophthalmological examination revealed that the best visual acuity was to 2/10 in the right eye and counting fingers in the left eye,examination of the anterior segment found an anisocoria, the intraocular pressure was normal in both eyes.

The fundus examination after dilatation demonstrated a bilateral papilledemastage two, peri-macular exudates (figure 1) without inflammation signs.

The examination of the oculomotor nerves showed an incomplete paralysis of the extrinsic third cranial nerve.

A neuro-pediatric examination was performed, which revealed an intracranial hypertension syndrome and an opto-chiasmatic syndrome, completed by a cerebral magnetic resonance imaging (MRI) showing a sellar and supra sellar process evoking a craniopharyngioma with an active left hydrocephalus (figure 3).

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Fluorescein angiography was performed, which revealed papillary diffusion of fluorescein confirming the papillary edema of stasis (figure 2), a visual field was also done agonized.

The child was referred to the neurosurgery department for immediate decompression with surgical removal.

The rest of the check-ups including hormonal assessment were normal, and the histopathological examination confirmed that the tumor was a craniopharyngioma.

The evolution was characterized by a tumor recurrence that necessitated intra-tumoral bleomycin treatment; and a tumor regression with a control at first and second years without anomalies.

Discussion:-

Childhood craniopharyngioma is a rare histologically benign embryonal tumor, that arises from Rathke's pouch remnants in the hypothalamic-pituitary axis.All age groups can be affected,their diagnosis is frequently delayed. [1] after the appearance of the first sign which is not always recognized: a slowing of growth [4]. The most frequent signs leading to consultation are; the triad comprising (1) signs related to intracranial hypertension due to hydrocephalus (headache, nausea/vomiting, macrocrania in the smallest) (2), visual signs (visual field amputation, decreased acuity) which may be revealing as in our case and (3) hypothalamic-pituitary dysfunction with endocrine deficiency, overweight, hyperphagia... [5]

The optimal management of these children with craniopharyngioma involves a multimodal strategy by a combination of surgery(s) and sometimes radiotherapy, whose goal is to control the disease and limit morbidity [6].

The strategy should therefore take into account the degree of neurosurgical urgency, the age of the child [7] and the degree of hypothalamic invasion estimated on preoperative MRI (grades 0, 1 and 2) [3].

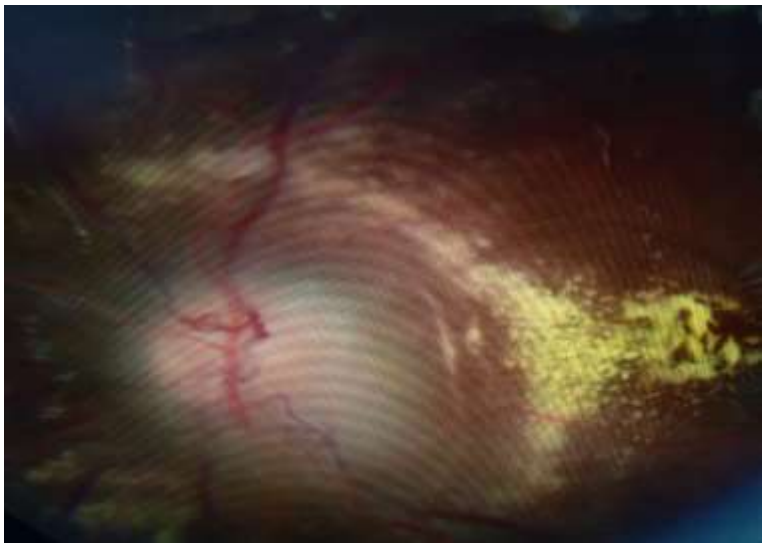
The localization and the origin of tumors effects the choice of surgical approach and prognosis. (8)

Craniopharyngiomas always remain severe lesions due to their potential for local recurrence, the neuropsychological consequences they can induce, particularly significant disorders of satiety, and consequently obesity.[9]

Conclusion:-

Craniopharyngioma remains a relatively common tumor among children and adolescents. Itsextension, the precocity of the diagnosis and the possibility of complete surgical resection govern the prognosis. The treatment remains difficult not only because of the need for a neurosurgicalintervention, but also by the complexity and heaviness of the hormone replacement therapy.

Figures:



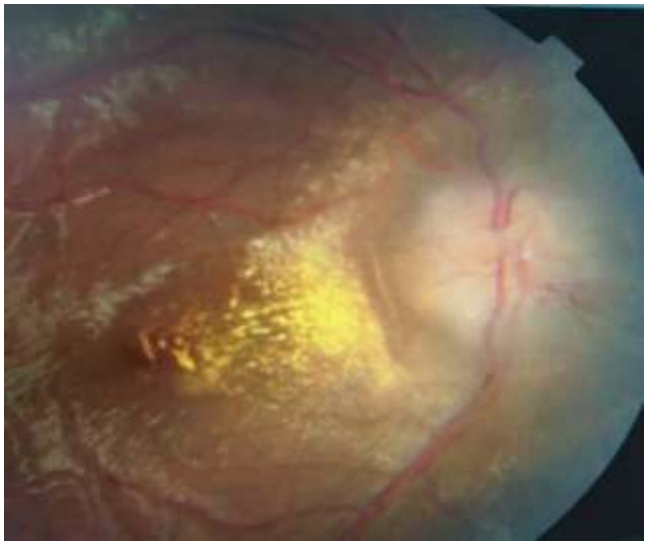
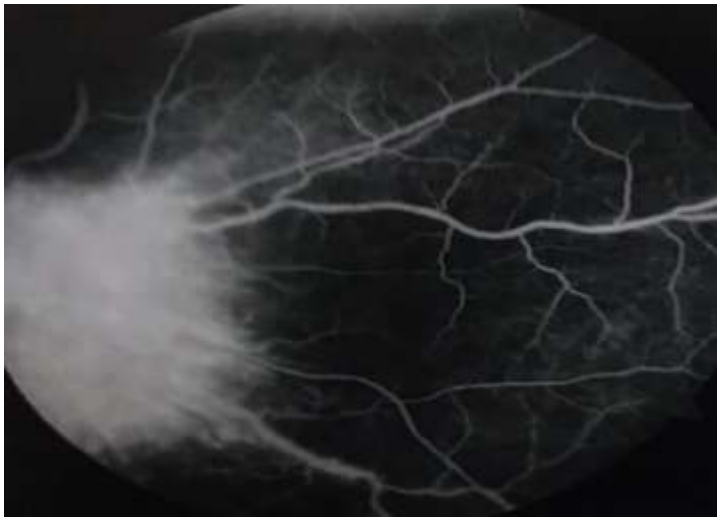


Figure 1:- bilateral papilledema stage two with macular exudates.



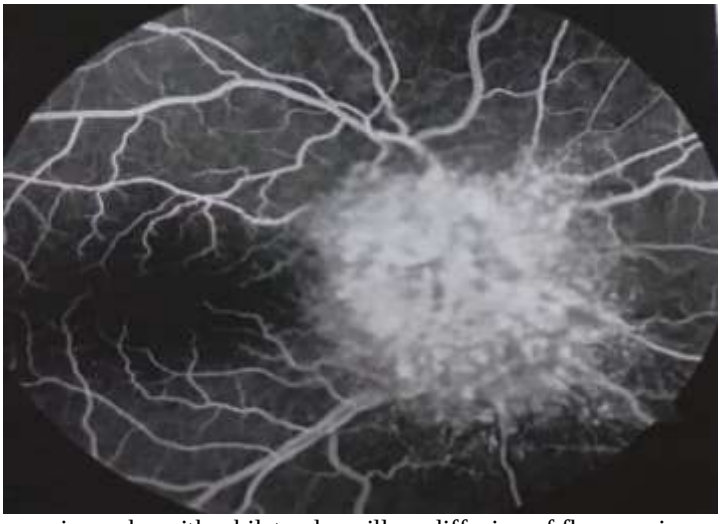


Figure 2:- Fluorescein angiography with a bilateral papillary diffusion of fluorescein confirming the papillary edema of stasis.

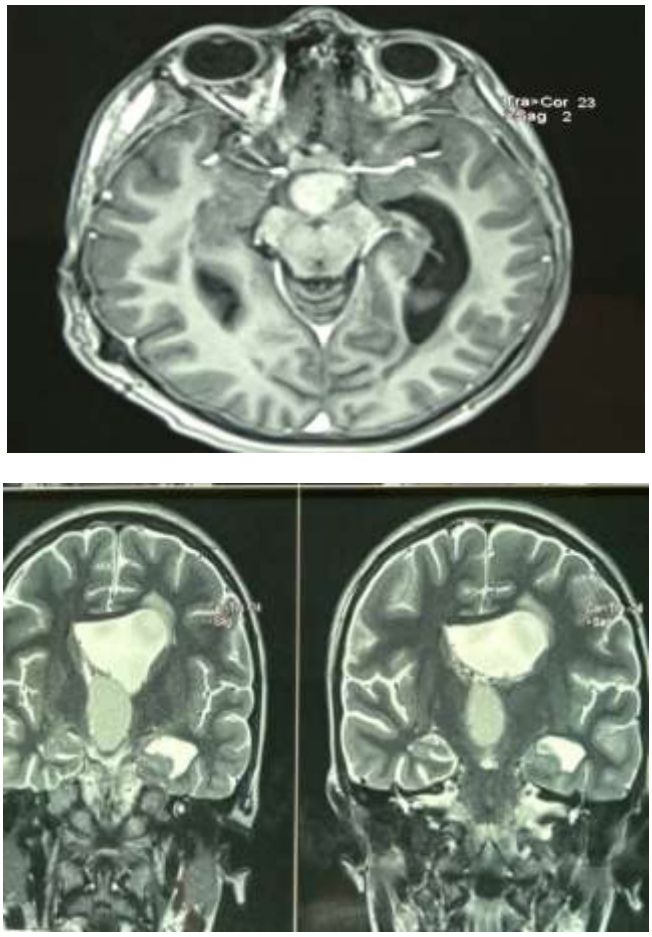


Figure 3:- Cerebral IRM showing a craniopharyngioma with an active left hydrocephalus.

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