

Management of a large craniopharyngioma in a child: A case report

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Abstract

We report the case of an 11-year-old girl who presents a huge sellar and suprasellar craniopharyngioma with cystic extension towards the two temporal fossae laterally, encompassing the two Sylvian arteries, towards the third ventricle and the frontal lobe above, towards the mesencephalon and the posterior cerebral fossa behind. The patient was successfully operated on but presented with panhypopituitarism post-op due to the sacrifice of the pituitary stalk which was invaded by the tumor.

Keywords: Craniopharyngioma; MRI; Child.

1. Introduction

Craniopharyngioma (CP) represent 1.2-4.6% of all intracranial tumors in children and carry a significant morbidity due to their lesional intimacy with structures involved in neurological, visual, and endocrinological functions [1].

2. Case report

We report the case of an 11-year-old girl, without any notable medical history, who consulted in the emergency department with headaches, vomiting, and gait disturbances with deterioration of vision. All are evolving over the past 3 months.

The clinical examination revealed a conscious patient with a Glasgow score of 15, intracranial hypertension syndrome, gait ataxia, and the ophthalmological examination: pupils in amaurotic mydriasis, negative light perception bilaterally, convergent strabismus and grade 2 papillary edema.

Note: no growth retardation or endocrine disorders and the hormonal balance was normal.

Brain MRI revealed a huge sellar and suprasellar solido-cystic tumor lesion with cystic extension towards the two temporal fossae laterally, encompassing the two Sylvian arteries, towards the third ventricle and the frontal lobe above, towards the mesencephalon and the posterior cerebral fossa behind. All suggesting a craniopharyngioma (Figure 1).

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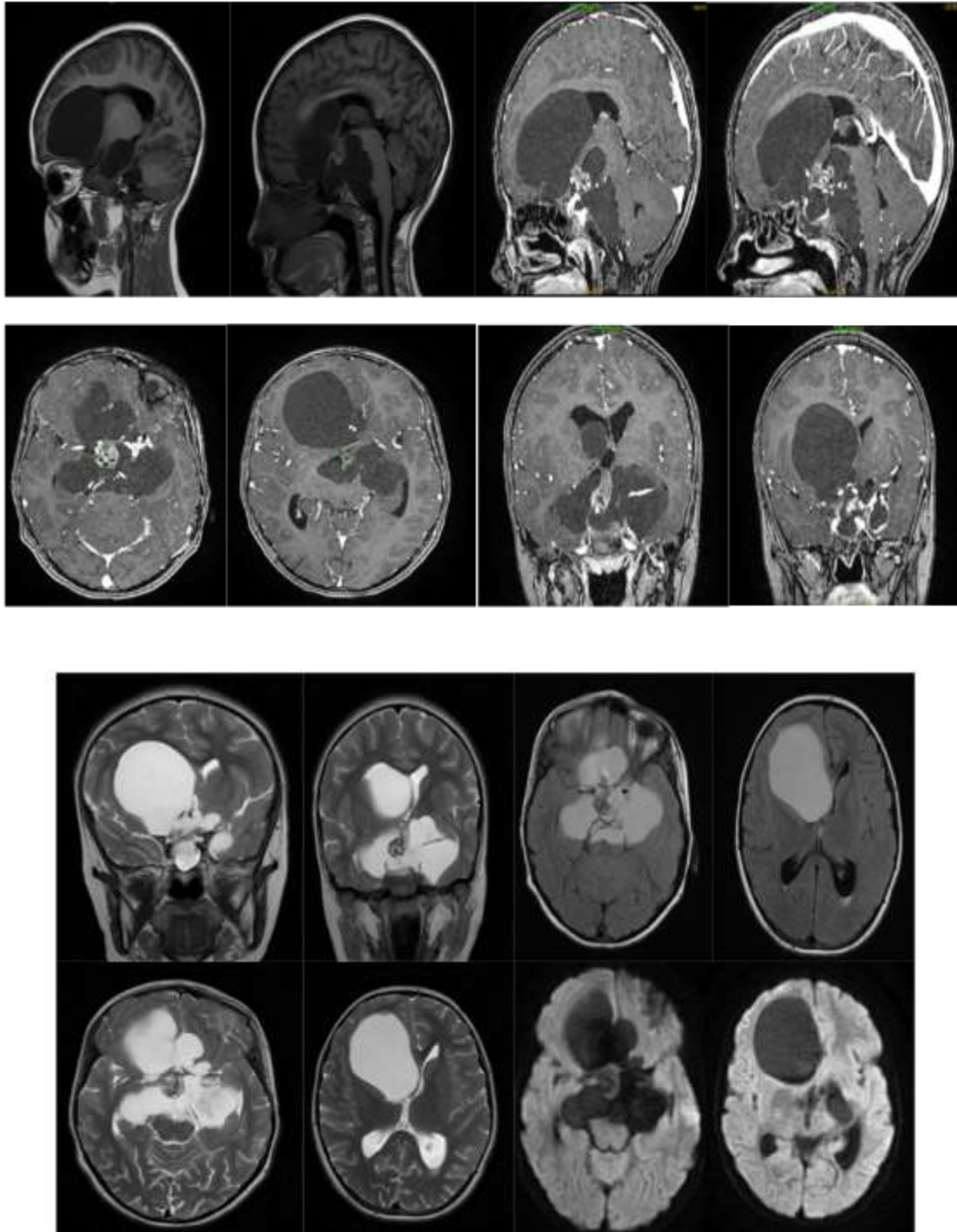


Figure 1 Preoperative brain magnetic resonance imaging.

The patient was operated on via the right pterion, Latero-frontal approach, and tumor resection was total, with preservation of the basal vessels. At the same time, the pituitary stalk was sacrificed as the tumor took it (Figure 2).

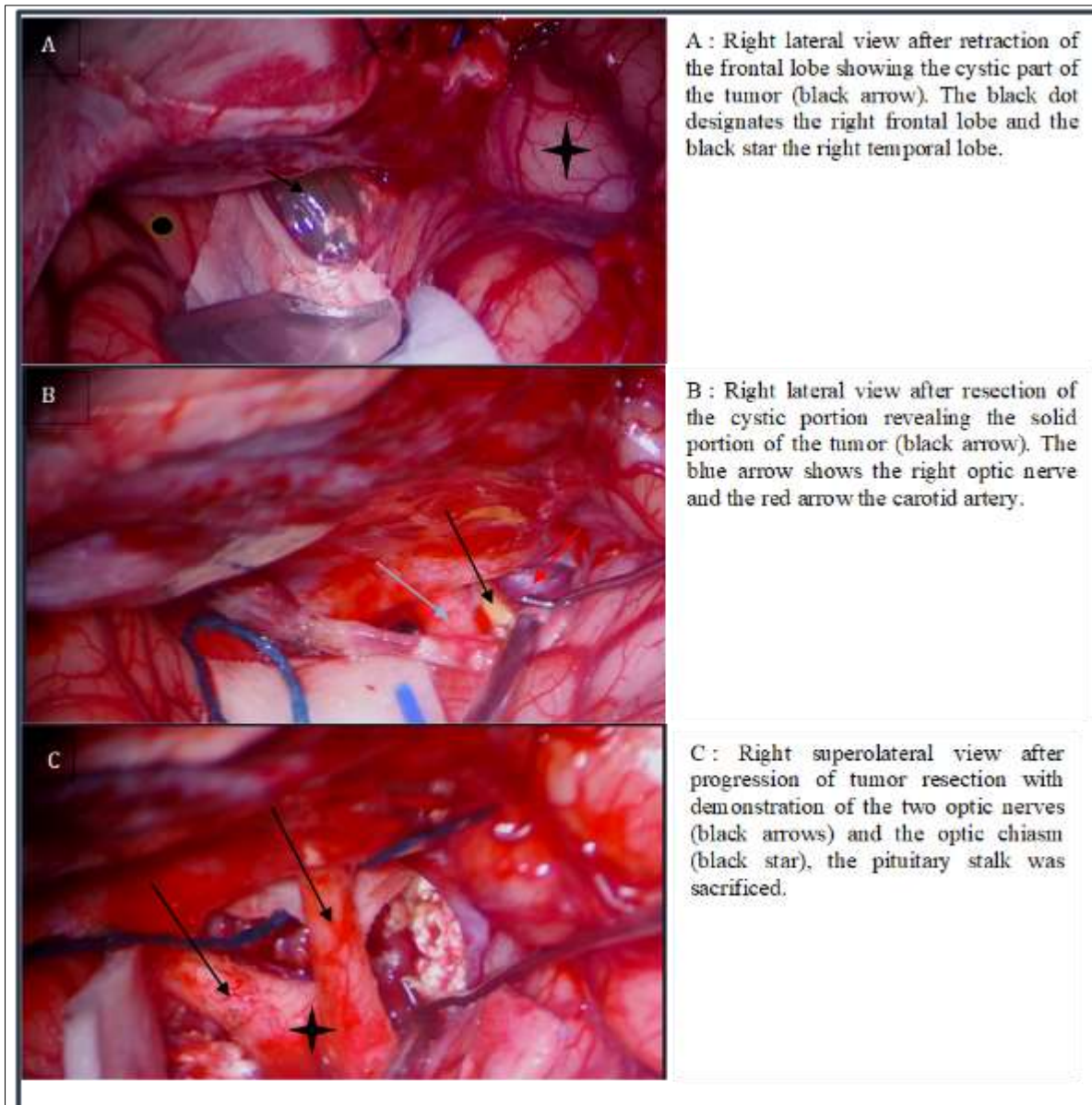


Figure 2 Intraoperative images

Immediate post-operative CT scan control confirms total resection (Figure 3).

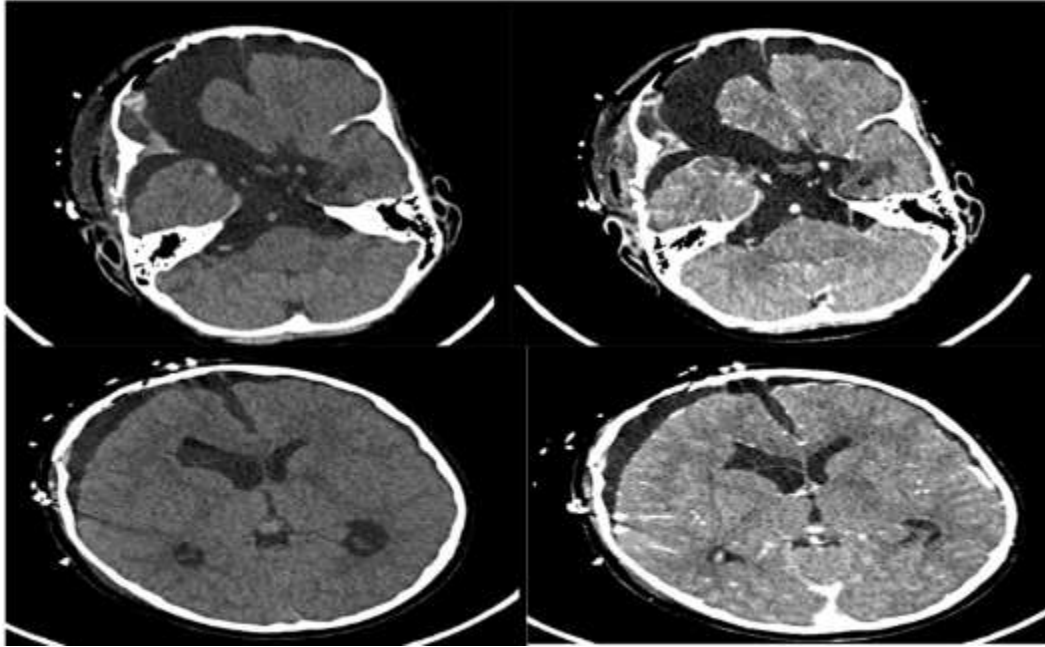


Figure 1 Postoperative axial section brain scan; uninjected on the left, injected on the right

The immediate post-operative evolution was marked by the development of diabetes insipidus (polyuria-polydipsia plus hypernatremia) treated with Minerin, thyrotropic insufficiency treated with Levothyrox, corticotropic insufficiency treated with hydrocortisone, epileptic seizure and meningitis treated with antibiotics, all with good clinical improvement. Brain MRI at six weeks was unremarkable and confirmed total resection of the craniopharyngioma (Figure 4).

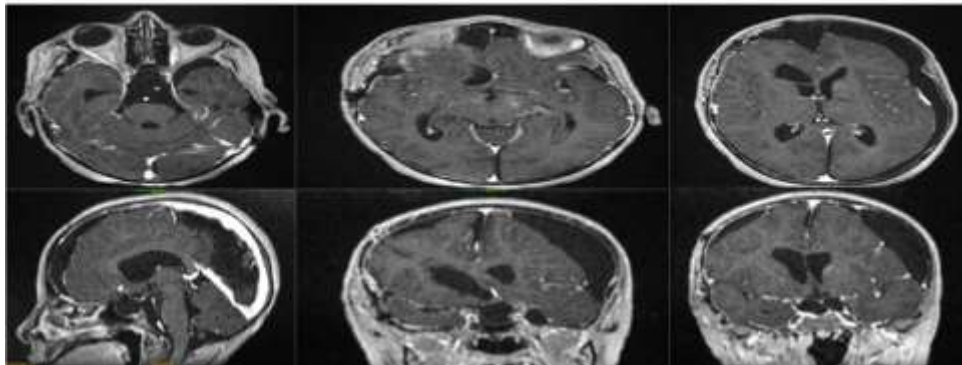


Figure 2 Brain MRI at six weeks post-op

Long-term evolution was marked by the reappearance of epileptic seizures revealing passive hydrocephalus treated by ventriculoperitoneal shunting.

3. Discussion

Craniopharyngiomas are rare, partly cystic and calcified embryonic malformations of the sellar/parasellar region with low histological grade (WHO I^o). A bimodal age distribution has been shown, with peak incidence rates in childhood-onset tumors at 5–14 years and adult-onset craniopharyngioma at 50–74 years [2]. Craniopharyngioma (CP) represents 1.2-4.6% of all intracranial tumors in children [1].

Clinical manifestations are related to hypothalamic/pituitary deficiencies, visual impairment, and increased intracranial pressure [1].

Variable treatment modalities being available, including surgery, radiation therapy, alternative surgeries, and intracystic therapies or combinations of them, their common goal is to reduce immediate and long-term morbidity while preserving these functions [1]. If the tumor is favorably localized, therapy of choice is complete resection, with care taken to preserve optical and hypothalamic functions. In patients with unfavorable tumor localization recommended therapy is limited hypothalamus-sparing surgical resection followed by local irradiation [2].

4. Conclusion

Cranio-pharyngioma is a low-grade tumor with locally aggressive behavior, leading to high morbidity in both children and adults. A multidisciplinary team of neurosurgeons, neuroradiologists, neuro-oncologists, pathologists, and endocrinologists should be involved in the diagnosis, treatment planning, and lifelong follow-up of these patients to achieve the best outcomes.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

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

References

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