Case Report

Voluminous craniopharyngioma evolving since childhood revealed by a torticollis

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Abstract

In case of a torticollis or a stature-weight growth delay in a child, it is always necessary to eliminate a cerebral tumor in the first place, especially a craniopharyngioma. We report the case of a 22-year-old patient with a large craniopharyngioma probably evolving since childhood and revealed by a torticollis associated with decreased visual acuity and a delayed growth and puberty.

Keywords: Craniopharyngioma, stature-weight growth delay, torticollis

Introduction

Childhood craniopharyngiomas are rare embryonic tumors arising from Rathke's pouch that put at risk the vital and visual prognosis of the patients and pose a problem of therapeutic management. Primary care physicians should always rule out a brain tumor in a child with torticollis or growth delay.

Case Report

A patient aged 22 years consulted for torticollis and decreased visual acuity. On examination: a pale patient with a childish face, a torticollis with lateral tilt of the head on the left side, a staturo-ponderal delay: weight: 40 kg (< -4DS) height: 1,46m (< -4DS), an impuberism with small testicles and a micropenis [Figure 1].

The ophthalmological examination showed visual acuity in the right eye of 0/20 with optic atrophy and visual acuity in the left eye of 16/20 with a stage II papilledema.

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We performed a cerebral computed tomography (CT) scan, which showed a tumor process with three components: liquid, tissue, and calcified, with polylobed contours measuring 88 × 83 × 90 mm [Figure 2]. Magnetic resonance image (MRI) showed a large tumor measuring 58 × 102 mm intra and suprasellar that extended to the left temporal fossa and frontal necrosis evoking a craniopharyngioma [Figure 3].

An endocrine hormonal workup was performed showing global anteropituitary insufficiency with corticotropic insufficiency low cortisol at 1.1 ug/dl, thyroid insufficiency TSH (thyroid stimulating hormone): 3.56 mui/ml (0.27-4.20) FT4: 8pmol/l (12-22), hypogonadotropic hypogonadism Testosterone: 0.03ng/ml (2.70-17.34) FSH (Follicle-stimulating hormone): 0.5mui/ml (1.50-12.4) LH (Luteinizing hormone): <0.1 mui/ml (2- 12), and somatotropic insufficiency with low IGF1 (insulin-like growth factor 1): < 50ng/ml.

Therapeutically, the patient benefited from hormonal substitution of the corticotropic axis (hydrocortisone 20 mg/d) and substitution of the thyrotropic axis (levothyroxine 75 µg/d). The somatotropic and gonadotropic axis could be substituted only after complete surgical removal of the tumor due to the risk of volume increase of the tumor under growth hormone (GH) or androgens (testosterone).

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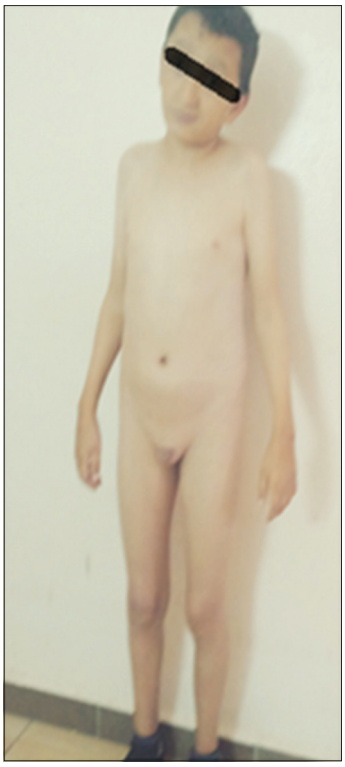

Figure 1: Image of the patient showing torticollis, impuberism, and staturo-ponderal delay

The patient underwent surgical excision via the right pterional approach with wide excision of the fleshy and calcic component of the tumor and evacuation of the cystic part. The pathological study of the surgical fragment showed a craniopharyngioma in its adamantinoma form.

The postoperative course was simple, with no onset of diabetes insipidus (no polyuro-polydepsia syndrome) or hypothalamic syndrome (no hyperphagia, correct natremia). An ophthalmological checkup was performed, which showed improvement of the papilledema in the left eye with a visual acuity of 20/20 and no recovery of the right eye since it was already at the stage of optic atrophy.

The MRI follow-up at 1 month showed the persistence of a tumor residue, and the patient was referred to radiotherapy for further management.

Discussion

Torticollis is a contracture of one or more muscles of the neck. There are many etiologies. Their severity is variable, including muscular pathologies, traumatic, inflammatory, and infectious or more serious causes such as revealing a brain tumor.

Torticollis in a brain tumor manifests as a lateral tilt block, not a rotational block. It reflects an antalgic posture due to an engagement of the cerebellar tonsils[1] as in the case of our patient who presented with a torticollis that revealed a large craniopharyngioma.

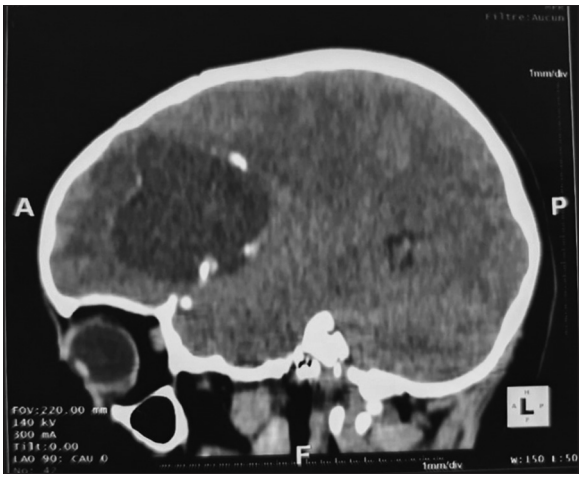

Figure 2: CT sagittal section showing a large cerebral tumor. CT = computed tomography

Craniopharyngiomas are benign tumors by their histology, but malignant by their functional and daily life quality repercussions: important risk of visual and hormonal disorders, school difficulties, and failure to thrive. Mortality is very low and is not due to the tumor itself, but essentially due to its hormonal impact. In our patient, the hormonal repercussions caused a severe delay in stature and a delayed puberty, and the functional repercussions of the tumor caused blindness in one eye.

The craniopharyngioma develops from Rathke's pouch, a remnant of the pharyngeal–pituitary canal. The adamantinomatous form, the most frequent in children[2] as in our patient's case, consisting of an epithelium derived from the dental enamel, often with calcifications and cysts. The tumor develops from the pituitary stalk, which can very rarely be preserved during resection. It, therefore, develops in the suprasellar region, either below the sellar diaphragm (subdiaphragmatic form) or above it (supradiaphragmatic form) or on both sides.[3]

The visual presentation, the patient presents a reduction of the visual field, often a decrease of vision whose diagnosis is often late, sometimes an oculomotor disorder related to the intracranial hypertension or to the infiltration of the cavernous sinus.[4] In our patient, the tumor caused optic atrophy in the right eye and papilledema in the left eye.

He endocrine presentation of craniopharyngioma, staturo-ponderal stagnation is more chronic, it can be a diabetes insipidus with nocturnal rising to urinate and drink, a growth delau because of GH deficit, an excessive weight gain linked to the infiltration of the hypothalamus.[5]

Imaging shows a suprasellar tumor, often calcic and cystic, and helps in studying its relationship with the hypothalamus, the visual pathways, and structures of the cavernous sinus.

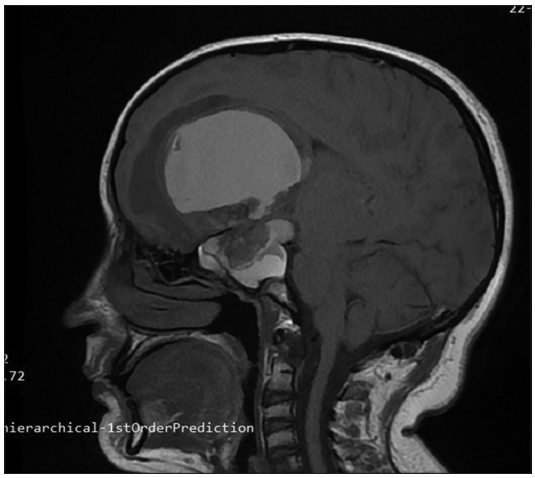

Figure 3: MRI sagittal section showing a large intrasellar and suprasellar craniopharyngioma. MRI = magnetic resonance imaging

The CT scan[6] allows visualization of the tumor, but does not always confirm its nature. MRI is the examination of choice for craniopharyngiomas. The cystic portion has a more intense signal on T1 sequences as the cyst contains more cholesterin or methemoglobin (some craniopharyngiomas may undergo hemorrhagic remodeling). Calcifications are not visualized on MRI, but the absence of a localized signal allows to guess their presence or even their outline. MRI is sufficient to confirm the presence of a tumor and to appreciate its relationship with all the vascular and nervous elements of the suprasellar and hypothalamic region.[6]

Surgical resection followed by radiotherapy is still considered the gold standard for craniopharyngiomas. Beyond providing rapid relief of symptoms, surgery allows tissue collection for histological diagnoses.[7] However, because of the infiltrating nature of the tumor to the hypothalamus and the cavernous sinus, it is often incomplete. Moreover, it is rarely possible to preserve the pituitary stalk and the patient will be dependent for life on a substitute treatment, as in the case of our patient in whom it was not possible to carry out a complete excision of the tumor and who retained a global anterior pituitary insufficiency for which he was put on substitute treatment.

Conventional radiotherapy[8] is an effective treatment, but has certain disadvantages which are as follows: aggravation of existing endocrine disorders, late cognitive disorders, and radiation-induced tumors which seem to be particularly worrisome in patients irradiated for craniopharyngioma. Gamma-knife radiosurgery[9] is an alternative to surgery for a small and hardly accessible fragment.

Medical treatment by anterior and posterior pituitary endocrine balancing is a major problem during the pre-, peri-, and postoperative period, with the need to substitute the endocrine axes, in particular, the corticotropic axis. Treatment with GH is usually necessary, but often delayed, especially since puberty is also delayed after complete resection of the tumor. However, these treatments must be given with close monitoring because craniopharyngiomas in children are aggressive tumors with a high rate of recurrence, even after apparently complete resection.

The evolution of the disease is marked by endocrine sequelae, which are almost universal, followed by visual sequelae, in particular, the reduction of the visual field. Cognitive and psychobehavioral sequelae[10] also have a major impact on the quality of life and integration. On the whole, the functional prognosis is mediocre, as in the case of our patient, who has retained blindness in one eye and a global anterior pituitary insufficiency. Our case aims to educate primary care physicians to eliminate cerebral tumors in front of torticollis or growth delay as soon as possible in children to allow an early management and to avoid hormonal and visual sequels.

Conclusion

In case of a torticollis or a stature-weight growth delay in a child, it is always necessary to eliminate a cerebral tumor in the first place, especially a craniopharyngioma, to allow an early management and to avoid hormonal and visual sequels.

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Conflicts of interest

There are no conflicts of interest.

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