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Craniopharyngioma: Epidemiological and Histological Profile About 22 Cases with a Literature Review

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Abstract

Craniopharyngiomas are benign epithelial tumors originating in the pituitary stalk or pituitary and developing in the sellar and supra-sellar region. The other names of these tumors: RATHKE pocket tumor, pituitary tract tumor, ERDHEIM tumor, adamantinoma, ameloblastoma, supra-sellative epithelial cyst, pituitary stem tumor, were abandoned in favor of the proposed craniopharyngioma term. by Mc KENZIE and SOSMAN in 1929, term taken up by Mc LÉAN in 1930, then CUSHING in 1932. The objective of this work is to study the clinical and pathological characteristics of these tumors. It is a retrospective study of 22 cases of craniopharyngiomas diagnosed at the pathology anatomy department of the Mohammed VI University Hospital of Marrakech between 2004 and 2017.These were 10 women and 12 men whose age ranged between 1 year and 50 years. Isolated intracranial hypertension was the main revealing symptom of the disease, isolated or associated The pathological study was performed on a fragmented material in all cases showing a craniopahryngioma adantinom in 90% of cases. Craniopharyngioma is a rare benign epithelial tumor that originates in the pituitary stalk or pituitary gland. Its incidence is 0.5 to two new cases per year and per million inhabitants. Of embryonic origin, it affects both adults and children. The clinical picture is very polymorphic. The diagnosis is based on MRI and CT scan, and anatomopathological study. Radical surgery makes sense in this benign extracerebral tumor. In case of incomplete excision, radiotherapy reduces the risk of recurrence. The evolution is finally favorable for 80% of the patients, allowing the return to a normal active life.

Introduction

Craniopharyngiomas are benign epithelial tumors originating in the pituitary stalk or pituitary and developing in the sellar and supra-sellar region. The other names of these tumors: RATHKE pocket tumor, pituitary tract tumor, ERDHEIM tumor, adamantinoma, ameloblastoma, supra-sellative epithelial cyst, pituitary stem tumor, were abandoned in favor of the proposed craniopharyngioma term. By Mc KENZIE and SOSMAN in 1929, term taken up by Mc LÉAN in 1930, then CUSHING in 1932 [1,2]. The objective of this work is to study the clinical and pathological features of these tumors.

Material and methods

This is a retrospective study of 22 cases of craniopharyngiomas diagnosed at the pathology anatomy department of the Mohammed VI University Hospital of Marrakech between 2004 and 2017.

Results

There are 10 women (45.5%) and 12 men (54.5%) with a sex ratio (M / F) of 1.2 (Figure 1) whose age ranged from 1 year to 50 years with an average age of 29 years.

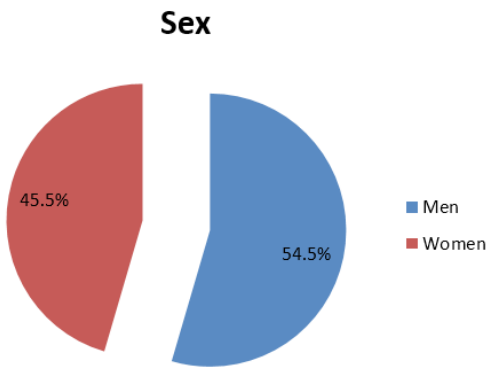

Figure 1: Distribution of patients by sex.

Most patients were polysymptomatic (Figure 2). Intracranial hypertension was present in 16 patients, was associated with visual disturbances in 5 patients, cerebellar syndrome in 4 patients, hemiparesis or facial palsy in 6 cases and adrenal insufficiency in 1 patient (Figure 3), and she was isolated in 6 patients.

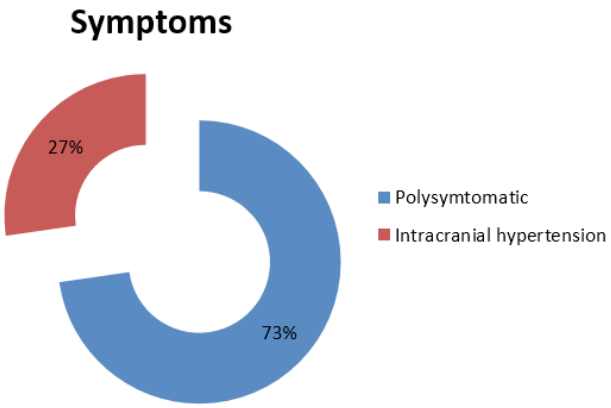

Figure 2: Symptoms.

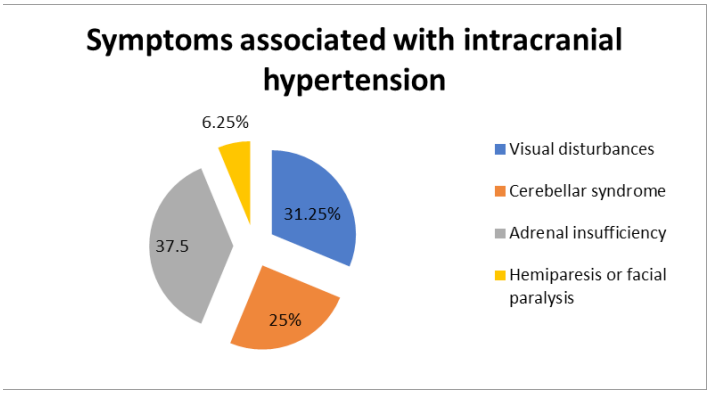

Figure 3: The main symptoms associated.

All patients underwent cerebral imaging such as computed tomography or magnetic resonance imaging, which looked like a craniopharyngioma (Figure 4). The anatomopathological study was performed on a fragmented material in all cases showing tumor proliferation arranged in thick trabeculae, cords, multistratified squamous epithelium bridges with peripheral palisades. Multistratified squamous cells are cylindrical or polygonal peripherally forming a peripheral palisade evoking an adantinomatous craniopahryngioma in 90% of cases, and squamous epithelium plaques forming pseudopapiles without palisades, nor calcifications, nor cholesterol crystals, absence of spongy center, the epithelial trabeculae rest on a fibrovascular stroma and form papillae in the papillary form 10% (Figures 5,6).

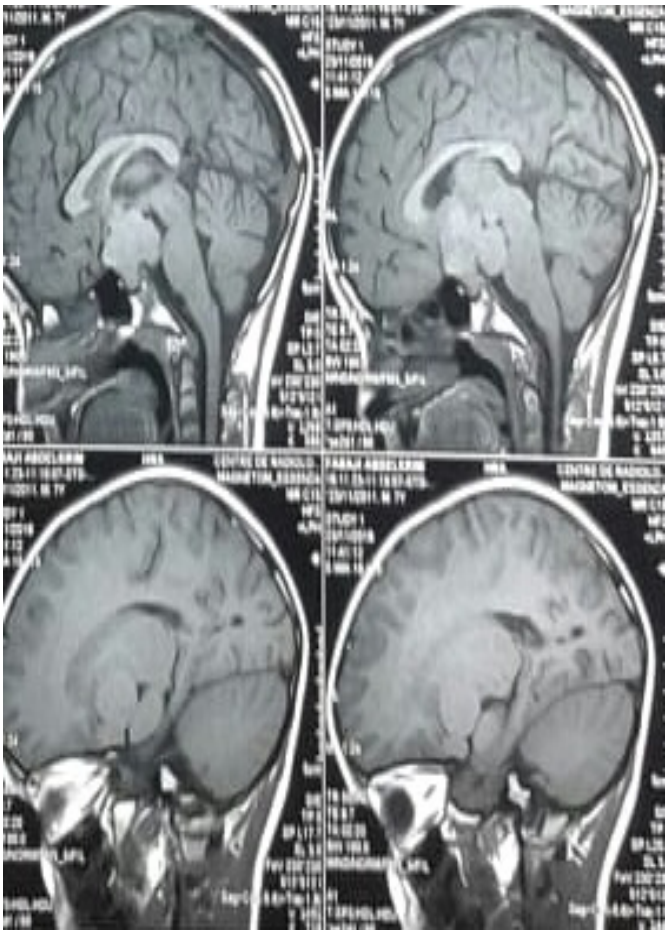

Figure 4: Sagittal magnetic resonance imaging :supra-sellar and sellral processes with fluid component and calcifications suggestive of craniopharyngioma.

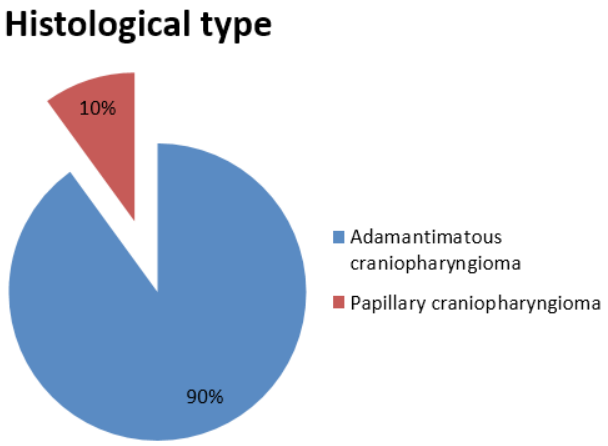

Figure 5: patient distribution by histological type.

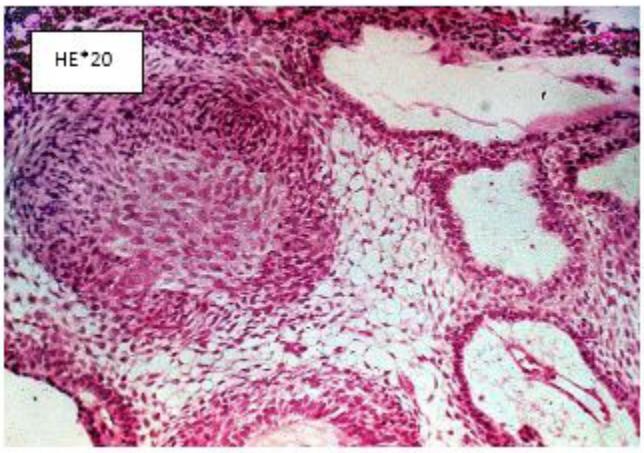

Figure 6: Microcystic proliferation bordered by squamous epithelium with palisades on the periphery suggestive of adamantinomatous craniopharyngioma.

Discussion

Craniopharyngioma is a rare benign epithelial tumor that originates in the pituitary stalk or pituitary gland. Its incidence is 0.5 to two new cases per year and per million inhabitants. Of embryonic origin, it affects both adults and children [3,4]. The prevalence of sex is variously appreciated according to the authors [5,6]. The majority of the series of the literature give a slight male predominance about 55% according to Choux [7] 67% for Cruz [5]. In our series, the male sex was predominant with a sex ratio (H / F) of 1.2. Age at diagnosis ranged from 1 to 50 years with an average age of 29 years. Like ours, the other series give a predominance of the disease in the second decade (29,2% for Cabezudo [8]. The relative proportions of the telltale signs are very variable according to the series of literature. The telltale signs are most often neurological and ophthalmological findings Endocrine signs are the third most frequently overlooked by patients and the clinical picture at the time of diagnosis in our series is similar to that described in the literature, with a combination of ophthalmological, endocrine and neurological deficits. Intracranial hypertension is the preferred mode of presentation of the disease in children, present in more than half of the cases.In the adult, it affects between 25 and 50% of the patients [9,10] The craniopharyngioma can take different In fact, this tumor contains three elements in variable proportion: a fleshy part, cysts more or less bulky and calcifications. Sometimes the tumor is essentially made of macro-cysts. These contain a yellow, brownish or green liquid, often flakes of cholesterin. Their surface is either smooth and translucent, or dotted with fine calcifications. The fleshy tumor containing microcysts or calcifications sometimes very extensive and one or more cysts of variable size. The fleshy tumor is frequently endo and immediate supersellar. The wall of the cyst or cysts infiltrates between the nerve and vascular formations of the supra, latero, pre and retro-sellar regions that it molds and represses and to which it often adheres. This adhesion may be related to the calcifications contained in the wall and which become encrusted in the nervous tissue and adventitious vessels. It can also be related to the intense reaction gliosis that occurs in adjacent nerve tissue. This glial reaction zone allows, at least theoretically, the anatomical cleavage of the tumor wall with respect to the functional nervous tissue.

This zone of gliosis, more or less extensive and thick, is not in fact constant and there are often places where the tumor appears really encrusted in the nervous system and where no cleavage plane can be found even under strong optical magnification [11-13]. Histologically The most common adamantinomatous form in children, representing 11% of intracranial tumors [8], consists of thick trabeculae, cords, multistratified squamous epithelium bridges with peripheral palisades. Multistratified squamous cells are cylindrical or polygonal periphery forming a palisade. Inside the cells are loose forming a spongy reticulum, Rosenthal fibers without keratohyaline granules. the papillary form consists of squamous epithelial plaques forming pseudopapiles without palisades, nor calcifications, nor cholesterol crystals, absence of spongy centers, the epithelial trabeculae rest on a fibrovascular stroma and form papillae. This form is especially solid, without calcifications, especially at the III level, they are more encapsulated, easier excision. This tumor is slow growing and is accompanied by significant reaction gliosis with many fibrosing Rosenthal fibers and marked inflammation with adherence to adjacent structures and vessels (resection difficulties). Intimate interdigitation and trapping of the vascularization of the Willis circle which explain the frequent recurrence despite the impression of complete excision. The treatment is based on complete excision, in case of incomplete excision, radiotherapy reduces the risk of recurrence. The evolution is finally favorable for 80% of the patients, allowing the return to a normal active life [11-14].

Conclusion

The craniopharyngioma is a rare tumor, relatively common in children.Although it is histologically benign, this tumor is the most formidable benign tumors given its endocrine consequences, its possible adhesions to neighboring neurovascular structures. The correct attitude requires a good correlation anatomoclinic to ensure proper care of patients.

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