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Craniopharyngioma: Epidemiological, Clinical and Therapeutic Aspects at the Central Hospital and the General Hospital of Yaoundé

Haman NO[1, 3], Ndome Toto O[2, 4], Djientcheu VP[1, 3]

[1] Department of Surgery and Specialties, Faculty of Medicine and Biomedical Sciences, University of Yaounde 1 (Yaounde, Cameroon)

[2] Department of Surgery and Specialties, Faculty of Medicine and Pharmaceutical Sciences, University of Douala (Douala, Cameroon)

[3] Yaounde General Hospital (Yaounde, Cameroon)

[4] Douala General Hospital (Douala, Cameroon)

Corresponding author: Haman Nassourou Oumarou.
Mail: haman06[at]yahoo.fr
Tel: +237 694194727
Department of Neurosurgery, General and Central Hospitals, Yaounde, Cameroon

Abstract: ***Introduction: Craniopharyngiomas are benign, partially cystic epithelial tumors of the sellar or supra sellar region, presumably derived from the epithelium of Rathke's pouch. They represent 1.2 - 4.6% of all intracranial tumors. Their age representation is bimodal. The classic clinical presentation is composed of signs of intra - cranial hypertension, endocrine, visual disorders. Their management is a great challenge in the neurosurgical departments. Objectives: The aim of our study was to investigate the epidemiological, clinical and therapeutic aspects of craniopharyngioma in two referral Hospitals (Yaoundé Central and General Hospital). Methodology: This was a cross sectional descriptive study with retrospective data collection in the neurosurgical departments of HCY and HGY. It covered a 10 years period from January 2011 to January 2021. All patients with primary tumor of the sellar and/or supra sellar region with presumption of diagnosis on imaging and/or histological confirmation of craniopharyngioma were included in our study. Data were collected using a pre - established data sheet; variables were recorded and analyzed by SSPS 23.0 software. Results: At the end of our study, 27 cases of craniopharyngioma were recorded. CP represented 12.3% of primary intracranial tumors. The sex ratio M/F was 0.8. The mean age was 16.3 years with extremes of 01 year and 52 years. Patients under 16 years of age were the most affected (70.4%). The average time of first consultation was 2.26 months. Patients came to the hospital for signs of raised ICP (51.9%), ophthalmological (33.3%) and endocrine (14.8%) involvement. The diagnosis on imaging was made mainly by CT scan (63.0%). Hydrocephalus was associated with the tumor in 55.5% of cases. Surgical treatment was the main therapeutic modality (81%). Early mortality was 14.8%, with 60% survival at 5 years. The adamantinomentous histological type was the most common in our environment (63.6%). Conclusion: Although craniopharyngioma a rare tumor, it accounts for one third of tumors of the sellar and/or supra sellar region. It is more frequent in children. The clinical presentation is classic with a predominance of advanced forms. The management is essentially surgical and the adamantinamentous histological type is the most frequent.***

Keywords: craniopharyngioma, epidemiology, clinical, treatment, Yaoundé.

1. Introduction

Craniopharyngiomas (CP) are rare tumors of embryonic origin classified according to the World Health Organization (WHO) among tumors with low histological grade malignancy (WHO 1). The WHO defines them as "benign, partially cystic epithelial tumors of the sellar or suprasellar region, probably derived from the epithelium of the Rathke pouch [1]. Craniopharyngiomas represent 1.2 to 4.6% of all intracranial tumors, which represents 0.5 to 2.5 new cases per million inhabitants per year worldwide. With 30 to 50% of cases presenting during childhood and adolescence and a higher incidence in Japan, West Africa and Nigeria [2, 3]. There is a well - recognized bimodal age distribution, with the first pic occurring at school age (5 - 14 years) and a second pic in middle or late adulthood (45 - 65 years) [3, 4]. It's a non - glial intracranial tumor derived from a malformation of embryonic tissue whose pathogenesis is still poorly understood. With mainly as theories encountered: embryo - genetics and metaplastic put forward [5]. Due to their sellar and suprasellar location, the clinical manifestations are essentially endocrine, ophthalmic and related to intracranial hypertension [6]. The paraclinical diagnosis of CP uses computed tomography (CT) and especially magnetic resonance imaging (MRI). These imaging examinations thus allow the direct visualization of the tumor, the study of its extension, the preoperative exploration and the post - therapeutic follow - up. Also, multiple endocrine explorations will be carried out in relation to the disorders caused by the tumor [7]. There are many controversies surrounding the optimal management of CP. Currently, surgical excision followed by irradiation by external beam, in the event of residual tumor, constitutes the main therapeutic option. Intracystic irradiation or bleomycin, stereotactic radiosurgery or radiotherapy and systemic chemotherapy are alternative approaches that are still being tested [1]. The major risks after surgical treatment are recurrence, as well as serious ophthalmic and endocrine sequelae, in particular diabetes insipidus, weight gain and pituitary insufficiency [8]. With a high survival rate (87 to 95% in recent series) [7]. Despite their benign histological appearance and high survival rate, they very often present

with an unpredictable growth pattern, combined with the lack of randomized studies, poses significant difficulties in establishing an optimal therapeutic protocol. Besides the choice of treatment, it is difficult to identify the clinical and imaging parameters that will make it possible to predict patients with an unfavorable prognosis [7]. These elements are largely the reason for the interest in this primary brain tumor, which is generating enormous controversy and therapeutic challenges around the world. We find very few studies on CP in Africa and particularly in our environment. This is why we considered it appropriate through our study to study the epidemiological, clinical and therapeutic aspects of CP in the two reference hospitals in the city of Yaoundé where this pathology is managed.

Patients and Method

We conducted a cross sectional descriptive study with retrospective data collection in the neurosurgical departments of HCY and HGY. Patients diagnosed with brain tumors and regularly followed at the neurosurgical department of two university teaching hospitals (HCY HGY) from January 2011 to January 2021. The medical records of all patients with primary brain tumors and histological evidence or diagnosed with CP were reviewed. Patients with primary sella and supra sella tumors without histological confirmation and those with incomplete files were excluded. The following data were collected: epidemiologic features (age, gender), clinical features (diagnostic delay diagnostic circumstance, signs of raised intracranial pressure, endocrine disorders, ophthalmologic signs, neurological and extra - neurological signs), investigations (computed tomography scan, brain magnetic resonance imaging,), treatment (surgical, quality of tumoral resection radiation and chemotherapy treatment), evoultive (post - operative complication) outcome (death, survival). The data collected were entered, analyzed with SPSS version 23.0and Microsoft Excel software. Ethical permission to conduct this work was obtained from Ethical Committee of the University of Douala and administrative authorization obtained from the different university Teaching hospital.

2. Results

We reviewed 235 medical records of patients with brain tumors, and only 27 showed histological evidence of craniopharyngioma. The frequency of CP was 12.3% with respect to primary tumor. The mean age was 16.3 years. There was a female predominance (56%). the average time to consultation after onset of symptoms was 67.8 days (2.26 months) Clinical signs found were: Intracranial hypertension syndrome (headache, vomiting, blurred vision) (92.6%), neurologic (11.1%), endocrine (37.0%), ophthalmic (81.4%). Brain CT scan was performed in (63%) of patients and brain MRI in 37% of patients. As for craniopharyngiomas, location in the supra sella and sella were more predominantly found (55.6%), isolated sella in 11.1% and 33.3% for suprasella locations. Sixty percent 60% of tumor were both cystic and fleshy in nature 22% cystic and 18% flesh the average tumoral volume was between 4 to 6cm3 with the average being 4.62cm.55% of patient had hydrocephalus on imaging. The histological type of craniopharyngioma was adamantine (63.6%) and papillary (36.4%).). All the patients had surgical interventions in this case, it was total surgical extirpation in 44.4%, 37.0% had DVP before surgical extirpation. In our study, 14.8% of the patients died post operatively, 14.8% presented with diabetes insipidus 7.4 post - operative meningitis.60% had five – year survival. None of the surviving patients received additional treatment after surgery.

Clinical presentation

Duration to consultation (in days)

Average Neurosurgical consultation time after onset of symptoms: 67, 8 days.

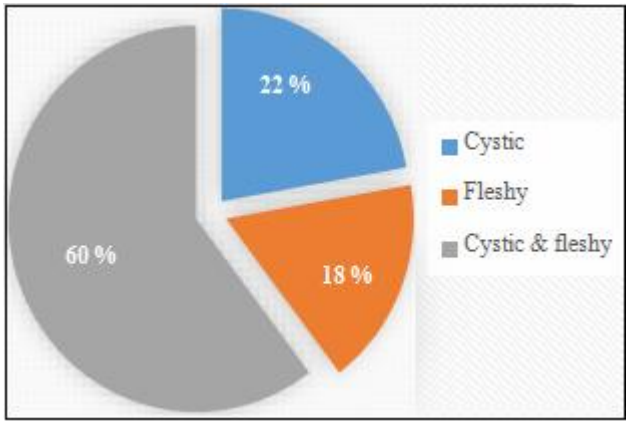
Circumstances of discovery

Table 1

Circumstances of discovery	Number (n=27)	Percentages (%)
Signs of intracranial hypertension:	14	51, 9
headache	12	44, 4
Vomitting	2	7, 5
Ophthalmological signs:	9	33, 3
Decline in visual acuity	6	22, 2
blindness	3	11, 1
Endocrine signs:	4	14, 8
Polyuro polydipsic Syndrome	2	7, 4
Amenorrhea	2	7, 4

Table 2: Distribution of complication

Types of complications	Numbers (n=27)	Percentages (%)
Cerebrospinal ascites	2	7, 4
Postoperative meningitis	2	7, 4
Diabetis insipidus	4	14, 8



On medical imaging, in most patients the craniopharyngiomas were both cystic and fleshy (60, 1%).

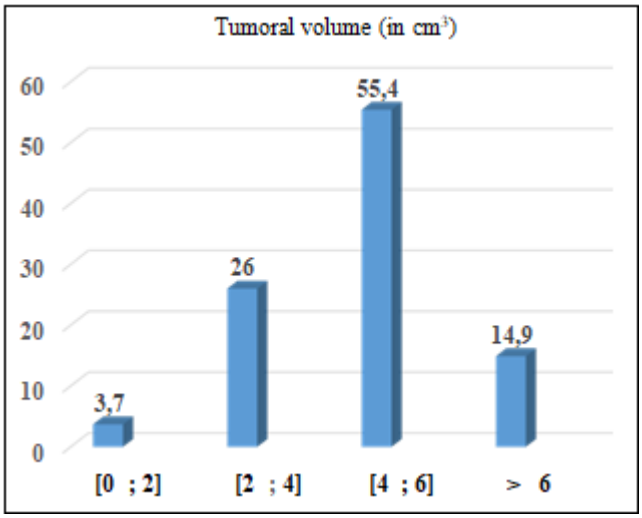


Figure 2: Breakdown by volume

The tumor volumes most often varied between 4cm^3 and 6cm^3, with an average volume of 4, 62 cm^3.

3. Discussion

In our study, the mean duration to consultation was 67.8 days (2.26 months). This duration was similar to that of Amayari et al in 2017 who found an average of 2.5 months. These rather long delays could be explained by the wandering of patients suffering from brain tumors in different specialties and health facilities [2, 4]. The frequency of craniopharyngioma in our series was 12.3% of intracranial tumors this is higher than the frequency Eyenga et al found in 2008 with a frequency of 4.8% [13] this could be due the increase frequency of this tumor in our environment. In our series, the female predominance joins the results found by some authors [6, 18]. Other authors reported a male predominance [10, 11], while other studies did not find any difference in sex predominance [2, 4, 19]. These conflicting results could suggest that there is a variable sex predominance in the CP. The mean age of patients with CP was 16.3 years old, this results is similar to that found by Elabadi et al in Morocco in 2013 who had 16.5 years [20]. Children were the most affected with 70% of patient being under the age of 16 and more specifically, 62.9% of the children were in the age group of 5 to 14 years. In our environment, CP would be a predominantly infantile tumor, in accordance with many authors who nevertheless describe a bimodal age distribution with two pics one in children aged 5 to 14, other in adults between 50 to 74 years old [1, 2, 7, 13]. The majors symptoms of CP were represented by the triad; neurological symptoms (symptoms of raised intracranial hypertension), visual disorders and endocrine disorders [6, 7, 9, 10, 12, 14, 15, 20]. In our study, the signs of raised intra cranial pressure were the most frequent with headache in 51.9% and 7.4% presented with vomiting. This results were similar to many other studies in the literature [1, 9, 12, 15, 20]. Visual disturbances represented the second mode of revelation of CP in our series, 33.3%, similar to other studies [12, 14, 15]. On the other hand Bouzzekar et al 2019 in Morocco found visual disturbances as the foreground with 51% cases [10] next to visual disturbance were endocrine disorders 14.8% with amenorrhea and polyuro polydipsic syndrome at 7.4% each [12, 14, 15].

In our series, 92.6% of patients with CP presented with signs of raised intracranial pressure, where the most revealing presentations were headaches (59.2%); macrocrania in infants (14.8%) and alterations on funduscopy examination in particular at 55.5% distributed as follows: papillary blurring (25.9%), papillary edema (18.5%) and finally optic atrophy (11.1%). These results are close to those found by several authors [10, 12].

81.4% presented with visual impairmentdistributed as follows: a decrease in visual acuity (37%); visual blurring (25.9%), blindness (11.1%), hemianopsia and diplopia in 3.7%. These data correlated with those of the literature [10, 12]. On the endocrine level, we found failure to thrive (37.0%) and weight related to a disorder of GH secretion (22.2%), obesity (14.8%) and polyuro - polydipsia syndrom in 14.8%. Muller et al in 2019 in Germany found similar data [21].

Neurological signs like seizure at 3.7%; consciousness disorder at 3.7% and motor deficit at 3.7% were described by Adeloye in Nigeria. In our series, the most performed examination was cerebral CT in 63.0% of patients. This result are similar with that found in Morocco by Elabadi et al in 2013. Limited number and difficult access to MRI in low - income countries [20] can explain these results.

In our series 55.6%of patients had a mixed tumor location (supra sellar and sellar) and suprasellar location in 33.3 %. These findings are similar to studies in the literature [17, 20]. In addition, 60.0% of patients presented with both cystic and fleshy component and 22.0% of patients presenting with a purely cystic component. These results are similar to those of Sadik et al 2005as well as Elabaddi et al 2013 in Morocco respectively [20, 22].

The average tumor volume in our series was 4.62 cm³. Shi et al in China in 2008 revealed an average tumor volume of 3.45 cm³ with volumes varying from 2 cm^3 to 9 cm^3 [11]. Sadik et al in 2005. Elabaddi et al in 2013 both in Morocco had respectively 45.0% and more than 50.0% of patients with a tumor volume greater than 4cm^3 [20, 22]. Karavitaki et al in the United Kingdom in 2006 showed respectively that 58.0 - 76.0% of tumor volumes in their series were between 2cm and 4cm^3 [1]. These results suggest the variability of tumor volumes in the different series in relation to diagnostic delays as well as a different size of the target populations. Hydrocephalus was associated with 55.5% of patients with craniopharyngioma in our series: this result is close to that of Elabaddi et al in 2013 in Morocco, which was 54.0% [20]. Karavitaki et al in the United Kingdom in 2006 found that 20.0 - 38.0% of cases were associated with hydrocephalus [1]. This difference may be related to the proportion of the pediatric population in the series; hydrocephalus being most often associated with craniopharyngioma in children [1]. Faced with the therapeutic challenge represented by craniopharyngioma; different therapeutic modalities were found in our series, in particular tumor excision surgery alone was performed in 44.4%; DVP followed by tumor resection was performed in

37.0% of cases. None of our patients had total tumoral excision and 91% subtotal excision, Shi et al 2008, in China had total tumoral excision in 89.3% and subtotal excision in 6.5%. This difference could be explained by the difficulties encountered in tumor excision surgery because of the close relationship of the tumor to important structures: anterior pituitary, hypothalamus and carotids [11]. Tumor excision requiring an excellent technical platform and a quality health system facilitating the follow - up of patients who would present heavy postoperative sequelae very often absent in low - income countries. None of the patients received adjuvant treatment, in particular radiotherapy. This in connection with the difficulties of access to radiotherapy and the absence of radiotherapy intended for children in our environment

The two histological subtypes of craniopharyngioma: adamantine craniopharyngioma (CPA) and papillary craniopharyngioma (PCP) differ in their genesis and age distribution. The adamantine type Craniopharyngioma were found at 63.6% and the papillary type at 36.4%. This result can be explained by the presence of many more children than adults in our series.

Postoperative complications in our series were represented by of diabetes insipidus in14.8%, postoperative meningitis in 7.4% are complications found in several other series at different proportions [10, 12]. It is important to specify that 7.4% (2) patients presented with refractory cerebrospinal fluid ascitis, a complication not found in other studies in the literature we reviewed. The latter being observed only in our environment in patients who have suffered from craniopharyngioma. The outcome of this complication being the death of patients two months after diagnosis. Postoperative mortality was 14.8%; this mortality was favored by the importance of intraoperative bleeding, infection and postoperative metabolic disorder. The 5 - year survival was 60.0% survivors; this result can be explained by the improvement of the therapeutic strategies.

4. Conclusion

Craniopharyngiomas are rare tumors among primary intracranial tumors. However, their frequency is increasing in our environment: moreover, they represent a third of primary tumors of the sellar and/ or suprasellar region. They are more common in children with a slight female predominance. The initial reasons for consultation are mainly headaches and vomiting in patients with a classic clinical picture. The latter generally appearing with advanced forms at the entrance. Computed tomography is the most requested examination for radiological diagnosis with tumors most often of mixed component and sellar and suprasellar location. Surgery is the main therapeutic modality in our environment with good postoperative results. Mortality is quite high compared to developed countries. Five - year survival is 60%; and the adamantinomatous type is the most common in our environment.

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