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Management of Pediatric Orbital Tumors in the Maxillofacial Department of Casablanca Morocco

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Abstract **Original Research Article**

Pediatric orbital tumors are a rare entity; they differ in their clinical and imaging appearance as well as in their histological nature. In this study, we report a retrospective analysis of a series of 21 cases of orbital tumors in children collected in the Maxillo-Facial surgery department at IBN ROCHD University hospital center in Casablanca during 9 years, from 2015 to 2023. The most affected age group is between 10 and 15 years, with a male predominance and a sex ratio of 3.2. Clinically, proptosis was the primary symptom in 76% of the cases. Fourteen patients underwent orbital and cerebral CT scans, which revealed various radiological features of each tumor, while MRI was used for 9 patients (43% of cases). While the majority of the lesions were benign (57%), orbital retinoblastoma was the most common tumor in 19% of cases, followed by rhabdomyosarcoma (14.28%) and orbital cysts (14.28%). The therapeutic management was multidisciplinary. All the patient underwent a surgical procedure; chemotherapy was needed in 33% of cases and radiotherapy in 4.76% of cases. This study aims to document and share the experience of the maxillofacial surgery department of Casablanca with pediatric orbital tumors.

Keywords: Pediatric orbital tumors, Proptosis, Retinoblastoma, Maxillofacial surgery, Casablanca, Histopathology.

INTRODUCTION

Orbital tumors are relatively rare in children; they are characterized by a great histological diversity, with a predominance of benign forms, particularly cystic ones. The clinical presentation is typically stereotyped, dominated by unilateral proptosis. CT, MRI, and other modern imaging techniques play a crucial role in guiding the etiological diagnosis, which is subsequently confirmed by histopathological analysis. Therapeutic management relies on three modalities: surgery, chemotherapy, and radiotherapy. These are selected based on the lesion type, its extent, and whether it is benign or malignant.

In this study, we report our experience in managing pediatric orbital tumors at the maxillofacial department 20 Août 1953 Hospital, outlining the epidemiological, clinical, radiological, histological, therapeutic, and prognostic characteristics of these cases.

MATERIALS ET METHODS

This is a retrospective, descriptive study involving 21 children under the age of 15 with orbital tumors who were hospitalized in the Department of Stomatology and Maxillofacial Surgery at the 20 Août 1953 Hospital in Casablanca between January 2015 and December 2023. Data collected from their medical records included demographic and clinical information, results of paraclinical investigations, therapeutic approaches adopted, and clinical outcomes.

RESULTS

Over a nine-year period, from January 2015 to December 2023, we received 21 child with orbital tumors, representing an incidence of 2.33 patients per year. Patient ages ranged from 13 months to 15 years, with a mean age of 8 years; the most affected age group was 10 to 15 years, accounting for 10 cases (47.61%). There was a male predominance, with a male-to-female ratio of 3.2. proptosis was the primary symptom, observed in 76% of the cases; it was unilateral in all patients. The right orbit was affected in 52% of cases. The onset of exophthalmos was gradual in 90.47% of patients, and the displacement was nonaxial in 52% of cases.

Figure 1: Clinical appearance of a non-axial proptosis of the right eye with chemosis

Figure 2: Clinical appearance of a non-axial proptosis of the right eye with ptosis

The other accompanying signs were primarily decreased visual acuity (67%), periorbital swelling (14%), blindness (14%), ptosis (9.5%), chemosis (9.5%), elevated intraocular pressure (4.7%), and limited eye movement accompanied by diplopia (4.7%). Fundus examination revealed stage I papilledema in 2 patients (9.5% of cases) and a retinal mass with vitreous seeding in a single patient (4.75%). Physical examination revealed nasal obstruction in one patient and cervical lymphadenopathies in another. Craniofacial CT scanning was performed on 67% of the patients. It enabled the characterization of the lesions' semiological features and revealed intracranial extension in two patients and sinonasal involvement in four. Nine patients (43%) underwent orbital and cerebral MRI, which allowed for a better topographic and vascular analysis of the lesions. Imaging techniques enabled us to determine the exact location of the tumor (Table 1) and guide the diagnosis in 86% of cases.

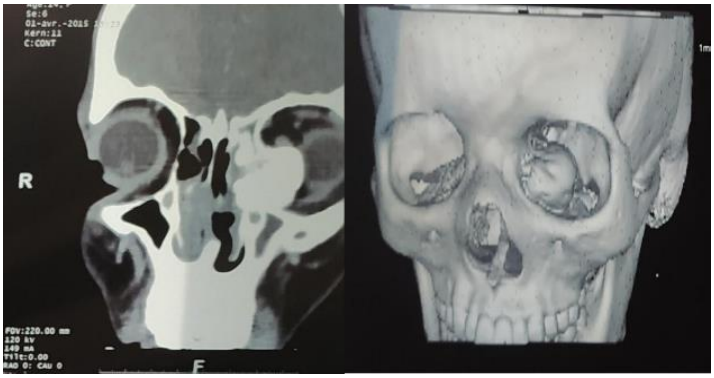

Figure 3: CT scan of a patient with an osteoma of the left orbit

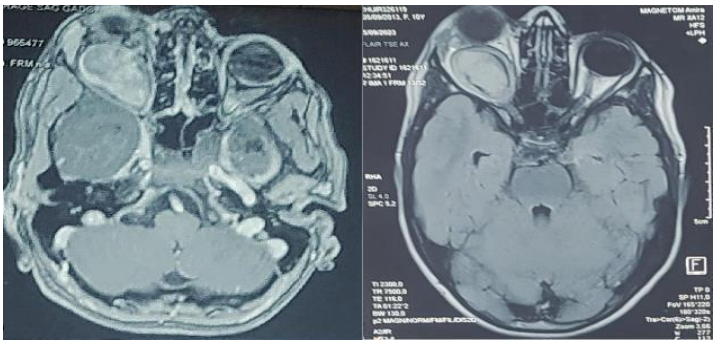

Figure 4: MRI of a patient with an optic nerve glioma of the right orbit

Table 1: Distribution of the location of the orbital tumors

Tumor location	Number	Percentage
Extraconal	9	43%
Intraconal	5	24%
Extra + Intraconal	4	19%

All patients underwent surgical exploration for diagnostic and curative purposes, and the choice of approach depended on the location of the tumor, the tumor volume, and the suggested histological type. An orbitotomy was necessary in 12 patients (57%).

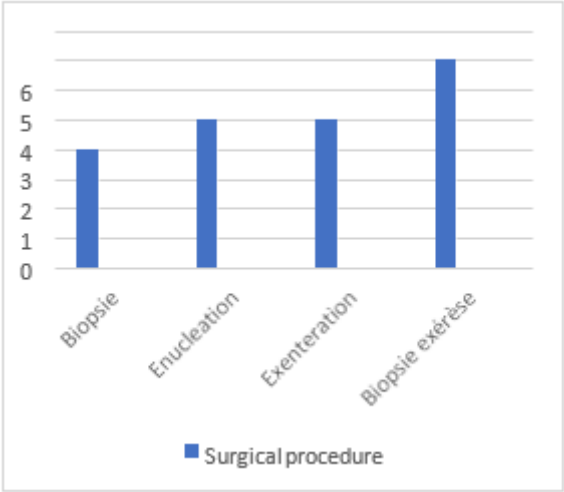

Figure 5: Distribution of the surgical procedures

Table 2 shows that in a total of 21 cases, 9 cases were of malignant tumors (43%), 12 cases of benign tumors (57%), the most common malignant tumor was found to be retinoblastoma with 4 cases (19%) cases and the most common benign neoplastic lesion was the dermoid cyst with 3 cases (14,28%).

Table 2: distribution of the orbital tumors based on the histological nature and on the gender

Tumor	Histological type	Age	Percentage	Male	Female
Benign Tumors 12 (57%)	Dermoid cyst	2 y	14,28%	2	1
	lymphangioma	8 y	9,52%	1	1
	hemangioma	4 y	4,76%	0	1
	Osteoma	14y	9,52%	2	0
	Ossifying fibroma	15 y	4,76%	1	0
	Shwannoma	8 y	4,76%	0	1
	Glioma	10y	4,76%	1	0
	Inflammatory pseudotumor	3 y	4,76%	1	0
Malignant Tumors 9 (43%)	Retinoblastoma	10y	19,04%	3	1
	Rhabdomyosacroma	6 y	14,28%	3	0
	Neuroectodermal tumor	7 y	4,76%	1	0

Surgical treatment was supplemented by additional management in 9 patients (42.8%), consisting primarily of chemotherapy, radiotherapy and intravenous corticosteroid therapy.

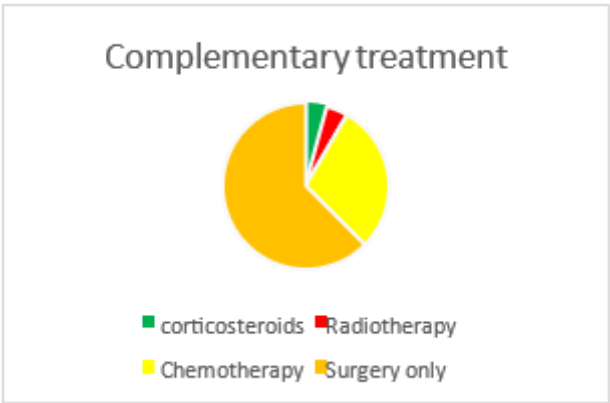

Figure 6: Distribution of the patients based on the received treatments

DISCUSSION

Orbital tumors are uncommon lesions within the spectrum of orbital pathology. Modi *et al.,* report an incidence of orbital tumors ranging from 3.5% to 4% of all orbital disorders [1]. Their frequency ranges from 5.6 to 13 cases per year [2–5], with a mean age at presentation between 3 and 7.5 years [2–6].

The clinical presentation of orbital tumors is typically dominated by proptosis, observed in 61.5% to 93% of cases [1, 3–8]. This proptosis is most often unilateral; it may be axial in 22% to 67% of cases [3, 9], suggesting a retroocular location for the lesion. An acute onset is noted in 28% of cases [10] and is most frequently seen in cases of rhabdomyosarcoma [11].

Other clinical signs—such as decreased visual acuity (found in 2.5% to 41% of cases [2, 3, 7, 8], inflammatory signs [7, 12], leukocoria [8, 13], diplopia or oculomotor paralysis [2, 8], and trigeminal hypesthesia [8]—may accompany the exophthalmos. Masses located in the anterior part of the orbit are usually palpable [7]. The main ophthalmoscopic signs of an orbital mass are chorioretinal folds [7], retinal vascular abnormalities [14], and optic disc edema and atrophy [3,7]. CT scans confirm the proptosis and quantify its grade. They also allow confirmation of the tumor's presence, location, and boundaries, as well as any intracranial extension. They also allow visualization of the various orbital structures and their contents, the paranasal sinuses, the nasopharynx, and the brain [15]. The presence of calcifications suggests certain pathologies such as retinoblastoma, histiocytosis, and dacryocystocele [16].

MRI of the orbit is the standard method for assessing endophytic (into the vitreous body) and exophytic growth patterns, possible infiltration of the optic nerve and extraconal structures, as well as intracranial extension, with superior sensitivity compared to CT scans [17, 18]. It can also distinguish lymphomas from pseudo-inflammatory tumors [17]. L Tumor location was specified in 66.66% of cases; an extraconal location was the most common, accounting for 28.57% of cases. Elyamouni *et al.,*specified the location in 73.2% of cases [7], while Vijay *et al.,*documented a high frequency of extraconal lesions (66.25% of cases); lesions involving both intraconal and extraconal spaces accounted for 22.5% of cases and were all malignant [5]. In the study by Yacoubi *et al*., an extraconal location was the most frequent, observed in 56.57% of cases [19].

Benign tumors were the most common in several studies (63% to 89%) [2–5, 19]. Conversely, malignant tumors predominated in others (59% to 65%) [1, 7, 20]. Surgery for orbital tumors may be solely diagnostic (surgical biopsy, percutaneous trocar biopsy, or fine-needle aspiration biopsy) or serve a dual diagnostic and therapeutic purpose, as in excisional biopsy. In certain cases, exenteration is necessary: for tumors poorly responsive to chemotherapy or radiotherapy, primary malignant orbital tumors, orbital metastases, intraocular orbital tumors with scleral breakthrough (such as retinoblastoma), and secondary malignant orbital tumors invading the paranasal sinuses or the cranium [8, 21].

CONCLUSION

Orbital tumors in children are rare and diverse. They threaten both functional and vital prognoses, necessitating early diagnostic and therapeutic management. Surgical procedures are becoming less invasive and increasingly precise through the adoption of conservative methods, and in some cases, they may be supplemented by chemotherapy, radiotherapy, or corticosteroid therapy.

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