Tumors of the maxillofacial region in children: retrospective analysis and long-term follow-up outcomes of 90 patients

Rezzan Tanrıkulu[1], Behçet Erol[1], Kenan Haspolat[2]

[1]Department of Oral and Maxillofacial Surgery, Faculty of Dentistry and [2]Department of Pediatrics, Faculty of Medicine, Dicle University, Diyarbakır, Turkey

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The aim of this study was to carry out a retrospective analysis of maxillofacial tumors in children and to present the lnog-term follow-up results.

Our study was performed with a retrospective analysis of 90 patients under the age of 15 years with maxillofacial tumor treated in our clinic between 1985-2002. In addition, treatment modalities and long-term follow-up results of these patients were evaluated.

According to our results, it was established that maxillofacial tumors were mostly observed in the 11-15 age group (39 cases, 43.3%) and on the mandible (48 cases, 53.3%). There were 21 (23.3%) odontogenic, 63 (70%) benign non-odontogenic and 6 (6.7%) malignant non-odontogenic. Mixed tumors were the most common type of the odontogenic tumors, and mesenchymal tumors were the most common non-odontogenic tumors.

Surgical excision, curettage or en bloc resection were adequate for treatment of these tumors.

Key words: tumor, maxillofacial, child.

A tumor is defined, in brief, as abnormal growth of tissue, and tumoral formations are classified under two main headings, benign and malignant.

A number of retrospective studies have been done on tumors of the maxillofacial region[1-5]. While pediatric tumors are far from uncommon, few studies on these have included retrospective analysis, demographic distribution, histopathologic spectrum, and treatment and follow-up outcomes[6-8].

The aim of the present study was to investigate the distribution of pediatric odontogenic and non-odontogenic tumors of the maxillofacial region according to age, sex, biological behavior, histopathologic spectrum, and location, as well as to evaluate treatment modalities and long-term follow-up outcomes.

Material and Methods

The present study was carried out on 90 patients who attended our clinic between 1985 and 2002 who were 15 years old or younger at the first visit, had healthy medical files, were radiographically and clinically diagnosed with odontogenic or non-odontogenic tumors, and were given appropriate treatment.

Seventy-five cases of pyogenic and peripheral giant-cell granuloma determined in our survey of medical records were excluded since they fell under the classification of reactive hyperplasia; only neoplastic formations were evaluated.

Tumoral formations were grouped under three main headings: odontogenic, benign non-odontogenic, and malignant non-odontogenic. Distributions according to age and sex, as well as histopathologic spectrum and location, were determined. In addition, distribution according to location was investigated for the subgroups of odontogenic and non-odontogenic tumors (epithelial, mesenchymal and mixed, fibrous lesions, vascular neoplasms, and neurological tumors).

Finally, the treatment modalities and long-term follow-up outcomes were assessed.

Results

Ninety children attending our clinic between 1985 and 2002 at ages ranging from 0 to 15 years with tumoral masses located in the maxillofacial region were included.

Age distribution was as follows: 15.6% (14 patients) were 0-5 years old, 41.1% (37 patients) were 6-10, and 43.3% (39 patients) were 11-15 (Table I). There was no noteworthy discrepancy in sex distribution, with the numbers of female and male patients being similar (43 girls, 47 boys) (Table I).

Of the 90 tumoral masses, 21 (23.3%) were odontogenic, 63 (70%) were benign non-odontogenic, and 6 (6.7%) were malignant non-odontogenic (Table I).

With regard to distribution according to tumoral mass location, the mandible was most frequently affected (48 patients, 53.3%), followed by the maxilla (27 patients, 30%) (Table II).

The location and frequency of the 21 odontogenic tumors indicated that the most frequent tumor type was mixed (12 patients, 13.3%) (Table III).

Our assessment of non-odontogenic tumors based on biological behavior, histopathologic spectrum, and location showed that slightly more than half of the tumors in this group (49 patients, 54.4%) were of mesenchymal origin, and that the majority of these were giant-cell neoplasms (36 patients) (Table IV).

Table I. Distribution of Tumors According to Age, Sex, Biologic Behavior and Tissue Origin

Age	Female	Male	Total	%	Tumor	Number	%
0-5	6	8	14	15.6	Odontogenic	21	23.3
6-10	17	20	37	41.1	Benign non-odontogenic	63	70
11-15	20	19	39	43.3	Malignant non-odontogenic	6	6.7
Total	43	47	90	100	Total	90	100

Table II. Distribution of Tumors According to Location

Location	Number	%
Mandible	48	53.3
Maxilla	27	30
Oral Mucosa	7	7.8
Tongue	2	2.2
Submandibular Area	2	2.2
Mandible+Maxilla	4	4.4
Total	90	99.9

Table III. Distribution of Odontogenic Tumors According to Location

			Location		
		Tumor	Mandibula	Maxilla	Total
Epithelial	2 2.2%	Ameloblastoma	–	2	2
Mesenchymal	7 7.8%	Odontogenic Fibroma	1	2	3
		Cementifying fibroma	3	1	4
Mixed	12 13.3%	Odontoma	5	3	8
		Ameloblastic Fibroma	4	–	4
	23.3	Total	13	8	21

Table IV. Distribution of Non-Odontogenic Tumors According to Biological Behavior and Location

		Tumor	Location						
		Benign	Mandible	Maxilla	Oral Mucosa	Tongue	Sub-mandibular	Disseminated	Total
Epithelial	5 5.6%	Verruca vulgaris	–	–	4	1	–	–	5
Mesenchymal	49 54.44%	Fibromatosis	–	1	–	–	2	–	3
		Fibroma	4	1	1	–	–	–	6
		CGCG	21	8	–	–	–	–	29
		GCT	2	5	–	–	–	–	7
		Osteoma	2	–	–	–	–	–	2
		Congenital epulis	1	1	–	–	–	–	2
Fibrous Osseous	4 4.4%	Ossifying fibroma	1	–	–	–	–	–	1
		Fibrous dysplasia	1	1	–	–	–	–	2
		Cherubism	–	–	–	–	–	1	1
Vascular	3 3.3%	Hemangioma	1	–	1	–	–	–	1
		Lymphangioma	–	–	–	1	–	–	1
Neurological	1.1%	NT	–	1	–	–	–	–	1
	Malignant								
R.E.S.	3 3.3%	Burkitt's lymphoma	1	–	–	–	–	2	3
Mesenchymal	3 3.3%	ERMS	–	–	–	–	–	1	1
		Round-cell sarcoma	–	2	–	–	–	–	2
	76.6%	Total	34	20	6	2	2	4	69

CGCG : Central giant-cell granuloma.
GCT : Giant-cell tumor.
NT : Neuroectodermal tumor.
ERMS : Embryonal rhabdomyosarcoma.
R.E.S. : Reticuloendothelial system.

The malignant non-odontogenic tumors in this study were determined to be Burkitt's lymphoma (3 patients), round-cell sarcoma (2 patients), and embryonal rhabdomyosarcoma (Table IV).

Treatment Modalities and Follow-up Outcomes

The majority of the 90 children with tumoral masses were treated with surgical excision, en bloc resection, and curettage. Some patients, however, received treatment specific to criteria such as the clinical behavior and extent of the lesion.

In one of the 36 patients with giant-cell lesions, involvement of the entire left mandible was observed, and, after hemimandibulectomy, the mandible was reconstructed with iliac bone graft, costochondral graft, and reconstruction plate; there was no recurrence on two-year follow-up (Figs. 1a-1b, 2). In five patients, giant-cell lesions (in the mandible in 2 and the maxilla in 3) exhibited aggressive behavior, causing destruction to the cortical bone, and were large enough to cause facial deformities; these patients were treated with surgical curettage and en block resection (Figs. 3a-3b, 4). Follow-up periods ranged from three months to 16 years. No recurrence was observed in any of the patients with giant-cell lesions, including the 16-year-old. The other giant-cell lesions were small and, after curettage under local anesthesia, follow-up was recommended, but none of these patients' follow-up periods exceeded one year.

One of the two patients with fibrous dysplasia, classified as a fibrous lesion, refused treatment, while the other, a 12-year-old girl, underwent staged surgery in three different periods: excision, en bloc resection, and osteoplastic contouring. She suffered no recurrence during 12-year follow-up. One patient (age 7) with cherubism, a fibrous lesion characterized by extensive involvement of the jaw and facial bones, has been under our supervision for approximately five years, and remission is expected during puberty.

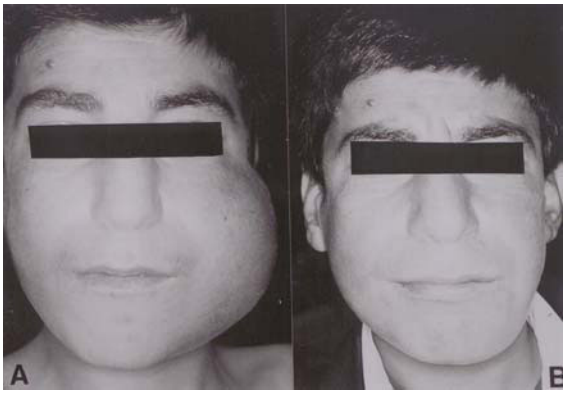

Fig. 1a-b. The patient with aggressive giant-cell lesion, preoperative and postoperative view.

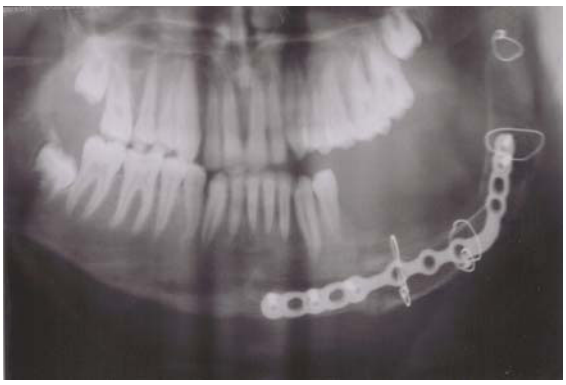

Fig. 2. Postoperative panoramic view of the same patient.

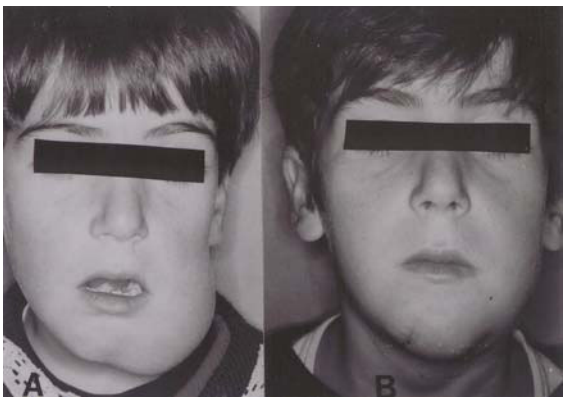

Fig. 3a-b. The aggressive giant-cell lesion on a seven-year-old girl. Preoperative and postoperative view.

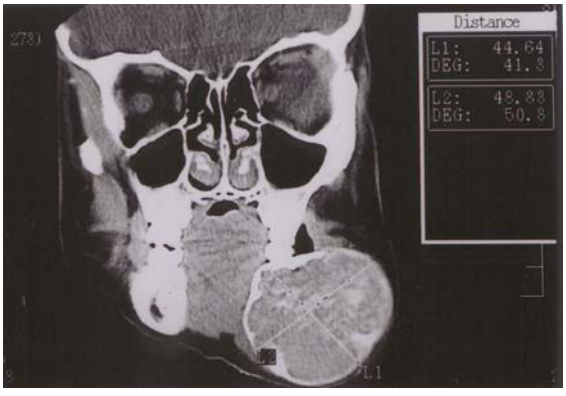

Fig. 4. Preoperative computerized tomography (CT) view.

Despite being categorized as benign tumors, ameloblastomas have a high rate of recurrence, and there is a risk of malignant transformation. One of the two patients with ameloblastoma underwent surgery approximately seven years ago, and thus far has not experienced recurrence. The other patient did not attend postoperative follow-up and examinations. Ameloblastic fibroma, a tumor of odontogenic origin, was determined in four patients (Table III). Two of these lesions were observed to have caused widespread destruction, affecting almost the entire hemi-mandible, and local exicision and curettage were performed. There was no recurrence during a mean follow-up period of seven years, and, with new bone formation in the region, the mandibular bone was reshaped in both patients.

The six-month-old patient with a neuroectodermal tumor underwent tumor excision, and recidivation was not observed during four years of follow-up. This patient's follow-up and supervision are still in progress.

Appropriate chemotherapy or radiotherapy were recommended in the oncology centers for three patients with Burkitt's lymphoma, a malignant non-odontogenic tumor, for one patient with embryonal rhabdomyosarcoma, and for one patient with round-cell sarcoma. While remission after chemotherapy was observed in one patient with Burkitt's lymphoma, the other four of the six malignant cases were not followed up. Another patient with round-cell sarcoma underwent surgery in our clinic approximately seven years ago, and is currently in good health.

Discussion

The majority of tumors of the mouth and jaw in children are benign[7,9]. Tanaka et al.[7] reported that only 3% of their cases were malignant in nature. In another study, benign tumors composed 93% of cases[9]. The present study, in parallel with the above-mentioned studies, showed a significant proportion (93.3%) of cases to be benign, with only six out of 90 (6.7%) being malignant. The reason for this ratio being less may be related to the smaller number of malignant tumors cases who applied to our clinic.

In contrast, studies performed in Nigeria have yielded malignancy rates of 40% or more[8,10]. Of Arotiba et al.'s malignant tumors[8] 22.4% were Burkitt's lymphoma, as were 44.8% of Asamoa's[10]. A high-grade non-Hodgkin's lymphoma, this tumor was first described in 1958 by Dennis Burkitt[11]. It is a prevalent neoplasm in children, and is endemic in Africa, although there is also a non-endemic form (North American Burkitt's lymphoma)[11]. The high incidence of malignant tumors in these studies may be accounted for by the endemism in Africa.

Tanaka et al.[7] reported that pediatric tumors occur most frequently in the 6-11-year age group (43.8%), followed by the 12-15-year group (31.4%). In a 102-patient series, they reported that 28 of 33 odontogenic tumors were in the 6-11 group, attributing this to the fact that crown formation of the permanent teeth is usually completed at 4-5 years of age[7].

A number of other researchers have reported higher incidences of tumor in the 11-15 age group[8,12]. The incidences for girls and for boys are reported to be approximately equal[7,8]. In the present study, in agreement with the literature, maxillofacial tumors occurred most frequently at 11-15 years of age (43.3%), while the rates for girls and for boys were similar.

In various studies on tumors, the mandible is reported to be the most frequently affected area[7,8]. In the present study, 53.3% of cases had mandibular involvement.

The great majority of pediatric jaw tumors are non-odontogenic[6,8,13,14]. Choung and Kaban[6] reported one ameloblastoma and odontomas of small diameter, as opposed to 47 non-odontogenic tumors. In a 46-patient series assessing benign jaw tumors, Dehner[13] found only four odontogenic tumors. In our series, non-odontogenic tumors accounted for 76.7% of tumoral formations, a considerable proportion.

Of all odontogenic tumors, ameloblastomas are the most controversial in terms of treatment[11,15]. Treatments range from surgical curettage to bloc excision or resection[11]. In planning treatment for pediatric tumors, authors stress the importance of the growth development of the jaw, and of esthetics and functional concerns in later periods of life[16,17]. In line with this view, with a single exception, we avoided radical resection in the treatment of all tumors, whether they were ameloblastomas or other benign odontogenic or non-odontogenic tumors. In addition, it has been reported that pediatric ameloblastomas are generally unicystic and do not extend beyond the cystic wall of the tumor cell[16]. In the present study,

there was no recurrence in the case of the cystic ameloblastoma that was located in the maxilla and exhibited growth into the sinus, which we were able to follow-up in the long-term.

Of benign non-odontogenic tumors in our series, tumors of mesenchymal origin were the most common (49 cases). This is in agreement with the literature data[6,8].

Of tumors mesenchymal in origin, giant-cell lesions had the highest incidence (36 cases). Choung and Kaban[6] reported that, a in their series, giant-cell lesions were the most common tumors of mesenchymal origin. Clear histopathologic distinction is not possible between central giant-cell granuloma and giant-cell bone tumor, both giant-cell lesions[6]. The histopathologic criteria to be considered in the diagnosis of real giant-cell tumors have been described, but the distinction between these two lesions cannot be made by histopathologic findings alone[6]. Therefore, in the diagnosis of cases we reported as giant-cell bone tumor and central giant-cell granuloma, in addition to histopathologic evaluation, intraoperative evaluation and he tumors's macroscopic appearance were important diagnostic criteria. The fact that the preliminary diagnoses we made based on our surgical experience were confirmed histopathologically suggests to us that, in giant-cell tumors, a specimen's microscopic appearance is more hemorrhagic, fragile, and liver-tissue-like in appearance than in central giant-cell granulomas, and that in central giant-cell granulomas, a tumoral tissue of solid, fibrous structure is dominant in the periphery of the surgical specimen; hence the curettage and enucleation of central giant-cell granulomas are easier. As a result of this observation, the following factors were determined to be criteria that must be considered in intraoperative evaluation and in the tumor's macroscopic appearance: the fragility, color, and consistency of the tumor tissue; whether or not it is hemorrhagic; and the ease of curettage and enucleation. Furthermore, the literature indicates that giant-cell lesions of the jaw may exhibit a variety of behaviors, and that central giant-cell granulomas may have as much changeability as aggressive lesions or malignant giant-cell tumors[6,18]. In giant-cell bone tumors in particular, recurrence is more expected due to aggressive clinical characteristics, and treatment consists of a range of surgical methods, from surgical curettage to hemimandibulectomy and reconstruction with bone graft[6]. There was no recurrence in any of our seven patients with giant-cell bone tumors. There was also no recurrence requiring a second operation in the six patients with giant-cell granuloma, which exhibited aggressive behavior and caused widespread bone resorptions and cortical perforations in places. One of these patients was treated with hemimandibulectomy, and the others with enucleation and curettage. Erol and Özer[19] reported that a central giant-cell granuloma in a six-year-old patient had caused widespread bone destruction in the corpus and ramus and that, after surgical curettage, there was no recurrence during long-term follow-up.

Another pathology that is histopathologically indistinguishable from giant-cell lesions is cherubism[6,11], a hereditary disease exhibiting autosomal dominant transfer[11]. It generally begins before the age of two, and spontaneous regression is expected after puberty. Choung and Kaban[6] followed up two cherubism patients, ages two and four, for 38 and 41 months, respectively, and determined minimal change. Our patient who attended our clinic at age seven and was diagnosed with cherubism has been followed up for approximately five years, and regression in puberty is expected.

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