

Classification of Pediatric Oral and Maxillofacial Tumours: A Review

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ABSTRACT

Background: Cancer represents the second leading cause of childhood mortality. In pediatric tumours, head & neck tumours constitute 5-10% of all primary malignant lesions affecting the children. Various classifications exist which deal with pediatric tumours in the body. These are mainly based on the cell of origin or the morphology of the tumour. The ICCC deals with pediatric tumours affecting the body and is primarily based on morphology and secondly on topography. Clinical & post surgical TNM classifications are presented exclusively for certain tumours occurring in the children. Oral tumours make upto 3% of pediatric tumours affecting the body & are often seen by a pediatric dentist. Very few classifications exist in this area of pediatric oral & maxillofacial tumours. The oldest classification of pediatric jaw tumours was given by Choung & kaban in 1985 which was later followed by the classification of masses in the head and neck by Rapidis et al. As the incidence of tumours in the oral & maxillofacial area of the child is only increasing, it is rightfully needed that more light is thrown into this area & a proper classification is put forward to deal specifically with these tumours.

Keywords: Pediatric, oral tumours, maxillofacial tumours, classification.

INTRODUCTION

With growing awareness and newer trends in pediatric dentistry, children visit the dentist not only for dental problems but also for routine dental check-ups as early as 6-10 months age. Hence any mass suspected of a tumour occurring in the oral and maxillofacial area would be noticed immediately and when diagnosed early could save a great deal of trauma to the child.

Cancer ranks second to trauma as a cause of childhood mortality. It represents the leading cause of death from disease in children. Majority of childhood cancers are malignant solid tumours. 5-10% of primary malignant tumours in children originate in the head & neck area, & one in four malignant lesions occurring elsewhere in the body

eventually manifest in the head & neck region.^[1] Oral tumours in children constitute approximately 3% of all tumour like growths.^[2]

Early detection and correct diagnosis of tumors is desirable in any case but becomes especially important in pediatric cases as a child has much more to lose with an entire lifetime at stake. For the sake of convenience in both prompt diagnosis & proper treatment, American cancer Society gave the International Classification of Childhood Cancer (ICCC), which is primarily based on tumour morphology. This classification applies to malignant neoplasms affecting the child in any part of the body. Choung & kaban gave the oldest classification of pediatric jaw tumours in 1985. Since then various authors have used a broad categorization of pediatric oral tumours and tumour

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like lesions which is mainly based on convenience in undertaking studies or writing papers rather than a proper classification system being established to deal specifically with pediatric oral tumours.

Hence a review was undertaken by the author to assess the classifications and categorizations that exist in pediatric oral tumours .

Review

Table 1. International classification of childhood cancer (ICCC) [3]

ICCC GROUP	ICCC GROUP	ICCC GROUP
I. Leukemias, Myeloproliferative diseases, & myelodysplastic diseases a) Lymphoid leukemias b) Acute myeloid leukemias c) Chronic myeloproliferative diseases d) Myelodysplastic syndrome & other myeloproliferative diseases e) Unspecified & other specified leukemias	II. Lymphomas & reticuloendothelial neoplasms a) Hodgkin's lymphomas b) Non Hodgkin's lymphomas c) Burkitt's lymphoma d) Miscellaneous lymphoreticular neoplasms e) Unspecified neoplasms	III. CNS & Miscellaneous intracranial & intraspinal neoplasms a) Ependymomas & choroids plexus tumour b) Astrocytomas c) Intracranial & intraspinal embryonal tumours d) Other gliomas e) Other specified intracranial & intraspinal neoplasms f) Unspecified intracranial & intraspinal neoplasms
IV. Neuroblastoma & other peripheral nervous cell tumour a) Neuroblastoma & ganglioneuroblastoma b) Other peripheral nervous cell tumours	V. Retinoblastoma	VI. Renal tumours a) Nephroblastoma & other nonepithelial renal tumours b) Renal carcinomas c) Unspecified malignant renal tumours
VII. Hepatic tumours a) Hepatoblastoma b) Hepatic carcinomas c) Unspecified malignant hepatic tumours	VIII. Malignant bone tumours a) Osteosarcomas b) Chondrosarcomas c) Ewing tumour & related sarcomas of bone d) Other specified malignant bone tumours e) Unspecified malignant bone tumours	IX. Soft tissue & other extraosseous sarcomas a) Rhabdomyosarcomas b) Fibrosarcomas, peripheral nerve sheath tumours & other fibrous neoplasms c) Kaposi's sarcoma d) Other specified soft tissue sarcomas e) Unspecified soft tissue sarcomas
X. Germ cell tumours, trophoblastic tumours, & neoplasms of gonads a) Intracranial & intraspinal germ cell tumours b) Malignant extracranial & extragonadal germ cell tumours c) Malignant gonadal germ cell tumours	XI. Other malignant epithelial neoplasms & malignant melanomas a) Adrenocortical carcinomas b) Thyroid carcinomas c) Nasopharyngeal carcinomas d) Malignant melanomas e) Skin carcinomas f) Other & unspecified carcinomas	XII. Other & Unspecified malignant neoplasms a) Other specified malignant tumours b) Other unspecified malignant tumours

Google and pubmed search engines were used with key words of pediatric oral tumours, pediatric jaw tumours, classification of pediatric tumors and standard reference books used in pediatrics were referred to determine the classification systems used in pediatric tumors with emphasis on oral and maxillofacial tumors. The various classifications and categorizations are listed below.

d) Gonadal carcinomas
e) Other & unspecified malignant gonadal tumours

The International Classification of Childhood Cancer is primarily based on morphology and secondly on site. Each ICCC group is further divided based on morphology and topography and individual codes are given for each entity. For the sake of convenience only the names of the ICCC group are mentioned in the table and the code numbers for each subgroup are omitted. Readers are kindly requested to see the reference article for details.

2. Classification by (Rapidis et al, 1988) [4]

A. Classification of “Masses” in the head & neck

1. Malignant tumours
2. Benign tumours
3. Tumour like conditions
4. Dysplasias from embryonal remnants

Table 2: Classification by (Rapidis et al, 1988) [4]

A.1. Histological Classification of malignant tumours	A.2. Histological Classification of benign tumours	A.3. Histological classification of tumour like conditions	A.4. Histological Classification of dysplasias from embryonal remnants
1. Lymphomas	1. Hemangioma	1. Dermoid & epidermoid cysts	1. Thyroglossal duct cysts
2. Rhabdomyosarcoma	2. Papilloma	2. Nasal & sinus polyp	2. Branchial cysts
3. Squamous cell carcinoma	3. Lymphangioma	3. Histiocytosis X – Eosinophilic granuloma – Hand Schuller Christian diseases – Letterer Siwe disease	3. Teratomas
4. Neuroblastoma	4. Fibroma	4. Pigmented lesions of the skin	
5. Fibrosarcoma	5. Thyroid adenoma	5. Pilomatixoma (calcifying epithelioma of Malherbe)	
6. Thyroid carcinoma	6. Nasal glioma	6. Trichoepithelioma	
7. Malignant histiocytoma	7. Lipoma		
8. Reticulum cell sarcoma	8. Schwannoma		
9. Hemangiopericytoma	9. Mixed salivary gland tumour		
10. Mesenchymoma	10. Epulis		
11. Kaposi's sarcoma	11. Thymic cysts		
12. Mucoepidermoid carcinoma	12. Osteoblastoma		
	13. Myoblastoma		
	14. Myxoma		

15. Glomus tumour

Table 3. Classification of Jaw tumours in children (Choung & Kaban, 1985)^[5]

1. Classification of Non odontogenic Jaw tumours in children			
I. Benign Mesenchymal tumours 1. Giant cell lesions 2. Fibro-osseous lesions 3. Myxoma	II. Hematopoietic & Reticuloendothelial tumours 1. Langerhans cell histiocytosis 2. Burkitt's lymphoma 3. Lymphoma	III. Neurogenic tumours 1. Neurofibroma 2. Neurilemmoma 3. Neuroma 4. Ganglioneuroma 5. Neuroblastoma 6. Melanotic neuroectodermal tumour	IV. Vascular lesions 1. Vascular malformation (capillary, lymphatic, venous, arterial, combined) 2. Hemangioma 3. Aneurysmal bone cyst
V. Malignant Mesenchymal tumours 1. Osteogenic sarcoma 2. Chondrosarcoma 3. Fibrosaroma 4. Ewing's sarcoma	VI. Malignant Epithelial tumours 1. Squamous cell carcinoma 2. Mucoepidermoid carcinoma 3. Adenoid cystic carcinoma 4. Adenocarcinoma		
2. Classification of Odontogenic Jaw tumours in children			
I. Epithelial tumours 1. Ameloblastoma (Peripheral; Unicystic; Solid, multicystic) 2. AOT 3. Calcifying epithelial odontogenic tumour	II. Mesodermal tumours 1. Cementoma 2. Periapical cemental dysplasia 3. Cementifying fibroma 4. Cementoblastoma 5. Odontogenic fibroma	III. Mixed Tumours 1. Ameloblastic fibroma 2. Odontoma	

4. Common Categorization of oral lesions ^[6]

- I Tumors and tumor like lesions
- II Papillary lesions
- III Radiolucent lesions of the bone
- IV Radiopaque lesions of the bone
- V Mixed radiolucent and radiopaque lesions of bone

Table 4. Common Categorization of oral lesions ^[6]

I Tumors and tumor like lesions	II Papillary lesions	III Radiolucent lesions of the bone	IV Radiopaque lesions of the bone	V Mixed radiolucent and radiopaque lesions of bone
Irritation fibroma	Squamous papilloma	Ameloblastic fibroma	Odontoma	Calcifying epithelial odontogenic tumour

Peripheral ossifying fibroma	Verruca vulgaris	Ameloblastoma	Osteoma	
Peripheral giant cell granuloma	Condyloma acuminatum	MNET of infancy	Fibrous dysplasia	
Pyogenic granuloma	Giant cell fibroma	Central giant cell granuloma	Cementoblastoma	
Gingival fibromatosis	Focal epithelial hyperplasia	Cherubism		
Hemangioma	Inflammatory papillary hyperplasia	Burkitts lymphoma		
Lymphangioma		AOT		
Congenital Epulis		Ameloblastic fibroodontoma		
Neurofibroma		Ossifying fibroma		
Mucosal neuromas				
Pleomorphic adenoma				
Juvenile fibromatosis				

DISCUSSION

The principal difference between cancers of children & adults is that the former include several embryonic tumours & the latter are most often epithelial tumours. The most obvious difference between these two kinds of tumours is that the precursor target cell is a generative stem cell in one (e.g. retinoblast, neuroblast, nephroblast) & a renewal stem cell in the other. 80% of observed adult cancer cases are induced whereas pediatric cancer is caused by genetic mutations of somatic cells.^[7]

Various etiologies for different cancers in children have been shown which include: role of EBV infection in malignant lymphoma, presence of a primary immune defect in patients with leukemia, a specific genetic abnormality associated with retinoblastoma (deletion of chromosome 13) & the maternal ingestion of diethylstilboestrol during pregnancy resulting in carcinoma of the vagina at puberty in the child.^[8] Various Classifications are proposed which make a simpler categorization of a wide range of pediatric tumours.

The ICCC (table 1) describes tumors all over the body and is based on morphology & topography. It has no particular section for tumors

in the oral & maxillofacial area.^[3] Choung & kaban gave one of the oldest classifications of jaw tumours in children in 1985 (table 3).^[5] This classification deals with only intraosseus tumours affecting the jaws but tumours of the oral soft tissues and salivary glands are not included.

Another classification by Rapidis et al in 1988 (table 2)^[4] classifies masses in the head and neck as malignant tumours, benign tumours, tumour like conditions & dysplasias from embryonal remnants. In this classification both benign & malignant tumors are subdivided histologically with no consideration for site. Tumor like conditions is a separate sub group which includes dermoid cysts & nasal & sinus polyps. But these lesions are no longer considered as tumour like lesions. Odontogenic tumors occurring in the child are aptly classified as epithelial, mesenchymal & mixed tumors.

While the classification of choung & kaban is still followed, even in text books of pediatric dentistry, some authors follow a broad categorization of oral lesions occurring in the child rather than a classification system.^[6] This categorization is partly based on radiographic appearance or partly the morphologic appearance (table 4). But when radiographic & clinical appearance is kept at base to

categorize lesions, having a separate group as tumors & tumour like conditions seems to be inappropriate. The categorization of pyogenic granuloma under tumour like conditions no longer fits the criteria. Similarly squamous papilloma is included under papillary lesions while in the correct sense it belongs to the category of benign epithelial tumors. The radiolucent lesions include a wide range beginning from odontogenic tumors at one end to cherubism at the other, which is a developmental fibro-osseous lesion. Similarly the radiopaque lesions also include tumors and fibro-osseous lesions.

In a study done by Saxena et al, the aim was to assess all central jaw tumours that reported to the institute, but apart from odontogenic & non odontogenic tumours, reactive lesions, fibro-osseous lesions, salivary gland lesions & aggressive cysts were included.¹² Kadlub et al did a retrospective study to determine the specificity of pediatric jawbone lesions. They classified the lesions as odontogenic, non odontogenic & pseudo tumours. Fibrous dysplasia, central giant cell granuloma, simple bone cyst & aneurysmal bone cyst were included as pseudotumours.¹³

It is clear that the categorization followed is more based on convenience to perform studies or a random attempt without proper basis. 5% of salivary gland tumors occur in pediatric patients & most of them occur in the parotid gland. A greater percentage of malignant salivary epithelial tumors are seen in children than in adults. Pleomorphic adenoma & Mucoepidermoid carcinoma are commonly seen in children & mesenchymal tumors are also seen in greater frequency in children than adults. None of the classifications incorporate the salivary gland tumors occurring in the child. [9,10, 11]

Tumours in children occur primarily due to an underlying genetic defect & the precursor target cell is usually a stem cell. [7] Hence newer advances in biotherapies like gene therapy or stem cell therapy can be targeted more on pediatric tumors.

Although new biotherapies are explored based on the new biology of cancer, it is unfortunate that none of the classification systems have included these parameters. Data on classification of pediatric tumours in the oral & maxillofacial region is especially rare. A new classification covering the

tumours of jaws, soft tissues & salivary glands in the oral & maxillofacial area of a child which can include not just the cell of origin but also the site, its growth patterns while also considering the genetic changes & prognosis of these tumors, is needed in this area of paediatrics so as to benefit the suffering child and avoid delays in diagnosis and treatment planning.

One must remember 'Today's children are tomorrow's future'. Hence an exclusive classification of pediatric tumors in the oral & maxillofacial region could benefit the child today and the mankind in general tomorrow.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

REFERENCES

1. Cunningham MJ, Mc Guirt Jr WF, Myers EN. Cancer of the head & neck, 3rd edn Saunders; 1996. Chapter 29; p. 598-624.
2. Bluestone, Stool, Kenna. Pediatric Otolaryngology, 3rd edn. Saunders; 1996.
3. Foucher ES, Stiller C, Lacour B, Kaatsch P. International Classification of Childhood Cancer. Cancer 2005; 103: 1457-67.
4. Rapidis AD, Economidis J, Goumas DP. Tumours of the head & neck in children. J. CranioMax Fac Surg 1988; 16: 279-286.
5. Choung R, Kaban LB. Diagnosis & Treatment of jaw tumours in children. J Oral Maxillofac Surg 1985; 43:323.
6. Pinkham JR, Cassamassimo PS, Fields HW, Mc Tighe DJ, Nowak AJ. Pediatric dentistry- infancy through adolescence, 4th edn. Saunders 2005.
7. Alfred G. Knudson, Jr. Pediatric Molecular Oncology. Cancer 1993; 71: 3320-4.
8. Viswanathan J, Desai AB. Achar's textbook of paediatrics. 3rd edn Orient Longman Ltd; 1989.

9. Fonseca I, Martins AG, Soares J. Epithelial Salivary gland tumours of children & adolescents in southern Portugal. Oral Surg Oral Med Oral Pathol 1991; 72: 696-701.
10. Robert A. Ord. "Pediatric Oral & Maxillofacial Surgery" Saunders, 2004. Chapter 14; p. 202 - 211.
11. Callender DL, Frankenthaler RA, Luna, Lee SS. Salivary gland neoplasms in children. Arch Otolaryngol Head Neck Surg. 1992;118: 472-476.
12. Saxena S, Kumar S & Pundir S. Pediatric jaw tumors: Our experience. J Oral Maxillofacial Pathol 2012 Jan – Apr; 16(1): 27-30.
13. Kadlub N, Kreindel T, Belle MV, Coudert A et al. Specificity of pediatric jawbone lesions: Tumours & pseudotumours. Journal of Cranio-Maxillo-Facial Surgery 2014 (42) 125-131.