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# Childhood Salivary Gland Tumors - An Update

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**Abstract**

Salivary gland neoplasms in children are rare. Salivary gland tumors in children and adolescents are extremely uncommon among these neoplastic disorders, with an estimated annual incidence of 0.08 per 100,000 children. Mucoepidermoid carcinoma is the most common malignant parotid tumor in children, followed by acinic cell carcinoma and lymphoma. Primary malignant lymphoma of the parotid gland affects around 1.7 percent of children with parotid neoplasms. This review lays emphasis on the various benign and malignant salivary gland tumors occurring in children along with clinical examination and management.

**Keywords:** Salivary Gland Tumors, Ranula, Pleomorphic Adenoma, Mucoepidermoid Carcinoma, Kuttner Tumor

## Introduction

Salivary gland neoplasms are usually rare in younger age group of less than 18 years; the morphogenesis is also controversial, but, the tumors arising from epithelial origin are considered exceedingly rare till adolescent years.[1] If we were to exclude the vascular neoplasms, then, percentage of malignant salivary gland tumors is approximately around 50%.[2] Pleomorphic adenomas, muco-epidermoid carcinomas and acinic cell carcinomas account for about 90% of all epithelial tumors in those individuals under 17 years of age.[3]

Salivary gland tumors are classified as benign, malignant, epithelial, soft tissue, hematolymphoid or secondary tumors. [3] Tumors of the salivary glands may be located in the parotid, submandibular, sublingual, or other minor salivary glands. The most common site of occurrence is the parotid gland, which accounts for 79-85% of cases. [4] Malignant tumors constitute 15-32% of all parotid tumors[3] and their clinical presentation is variable, where in few cases, there may be asymptomatic slow growing tumors and in others, there may be late recurrence after treatment. [5]

Salivary gland neoplasms can be considered “orphan” diseases, which implies [6]:

- There is a paucity of clinical and biological details about them and their clinical behaviour, and many pediatric oncologists and surgeons may be unaware of the treatment options available
- No specific clinical or scientific organizations have been established to support their clinical management and related research
- It is very difficult to conduct clinical trials on them, which makes it hard to develop evidence-based treatment guidelines, thus making their treatment individualized
- Dedicated financial resources are most of the times limited

**Anatomy of the salivary glands**

The parotid gland is the largest of all the salivary glands, has 5-6cm excretory duct, also known as parotid duct or Stensen’s duct; has an average diameter of 1.4mm at the hilum, 1.2mm as it runs through buccinators and 0.5mm at the ostium. [7] The body of the gland lies in the retromandibular fossa, above and posterior to the masseter muscle and the mandible. The seromucous submandibular gland is located between the anterior and posterior bellies of the digastric muscle on the hyoglossal muscle; the duct's typical width is 1.5mm over its whole length from hilum to ostium. [7] The sublingual gland is located in the sublingual fossa, just above the mylohyoid muscle, in submucosal tissue; its excretory duct empties into the submandibular duct or exits independently into the oral cavity on the salivary papilla. [8]

**Physiology of the salivary glands**

A normal adult healthy person produces 1.5 litres of saliva per day; 20-25% comes from the parotid, 70-75% from the submandibular gland and 5% from the sublingual gland. The viscosity of saliva is determined by the proportions secreted by the individual glands. Hidas et al. [9] studied salivary flow in children and adolescents with attention deficit hyperactivity disorder (ADHD) where the total salivation rate at rest was reported at 1.1 ml/min in the control group, in contrast to the flow rate in patients with ADHD with or without medication which was reported as significantly lower.

Saliva consists of 99.5% water and the rest 0.5% is made up of proteins, electrolytes and various antimicrobial and bactericidal factors. On a whole, there emerges two characteristically secretory compartments; the first being the fluid serous saliva which has copious amounts of bactericidal substances and the second is the mucous component of saliva, which also aids in sense of taste and clarity of speech. [10]

**Clinical examination and imaging**

Any symptom of a disease or an underlying pathology starts with a detailed and appropriate clinical examination and recording accurate history; a careful history is the first and decisive step which will finally aid in the diagnosis, prognosis and treatment of the underlying condition. Most conditions affecting the salivary glands results in either a painful swelling or a painless swelling, which may be coupled with xerostomia, sialorrhea, facial palsy and related generalized symptoms which accompany any infection. The differential diagnosis is of significance particularly in children such as lymphadenopathy which may be due to various reasons and origins, autoimmune and systemic diseases etc. Clinical examination may aid in assessment of Suppurative infections, salivary stones, tumors etc. Further, with advanced diagnostic aids such as Imaging, one can define the condition and confirm the diagnosis

- Colour Doppler helps to characterize lesions precisely, especially the haemangiomas and arteriovenous malformation
- Traditional Sialography and X-rays have no place in diagnostic investigations, especially in childhood, and Sialography must be conducted under sedation in young children.
- Magnetic Resonance Imaging and Computed Tomography are 'reserved' for certain space-occupying lesions that may exceed the ultrasonographic limits of salivary glands and further extends into the para-or retropharyngeal spaces or even to the base of the skull or mastoid. MRI focuses mainly on soft tissue and nerves, MR Sialography may be extended to examine the ducts or topography of vascular supply. CT Scans are used to assess bony structures which may aid in determining extent of abscesses, particularly in the para-or retropharyngeal spaces.
- Sialendoscopy, due to its direct visualization, can be of superior help in obstructive salivary gland diseases
- Fine Needle Aspiration Biopsy can be used to ascertain the exact nature of tumor
- Polymerase Chain Reaction is employed to detect pathogens
- Fine needle aspirations are limited in children and must be used very carefully due to further risks.

**Congenital salivary gland diseases/pathologies**

Hypoplasia or aplasia of the salivary glands is a rare occurrence, but can affect one or all of the salivary glands. Aplasia may occur in isolation or associated with other malformations of the first branchial arch; and are usually not recognized for many years as the patients seldom complain any related symptoms. But on occasions, patients usually present with large carious lesions associated with xerostomia, difficulty in swallowing and regurgitation of fluids. Early diagnosis can help in considerable relief to the patient.

Only 5% of all salivary gland tumors occurs during childhood; congenital tumors are also uncommon. [11] Tumors that arise mostly in the perinatal period can be easily identified on ultrasound scans during pregnancy; the parotid and submandibular glands being most affected. The treatment of choice is always resection, but may present difficult scenarios when there is possible infiltration into the surrounding structures. Chemotherapy is also a useful adjunct in many cases where the tumor residues respond well.[12] Scott et al [13] have reported from their series, that as far as rhabdomyosarcomas are concerned, they can be effectively treated with several rounds of Ifosfamide $3g/m^2$/day on days 1 and 2, Vincristine $1.5g/m^2$ on day 1; Actinomycin D $1.5g/m^2$ on day 1 and Doxorubicin $30mg/m^2$/day on days 1 and 2. It has to be kept in mind that radiotherapy at this age is associated with adverse effects of bone growth later on.

**Salivary gland infections/pathologies**

Children are mostly affected with inflammatory conditions; the most important viral pathogens being the mumps virus and cytomegalovirus [14] that affect the salivary glands. Further, there may be an interplay of acute bacterial infections, chronic inflammation and autoimmune diseases, coupled with the role that saliva itself plays in many bacterial, viral and other mycotic diseases.

Table1: WHO Histological Classification of Salivary Gland Tumors 2017 [15]

| Benign epithelial tumors | Malignant epithelial tumors |
|---|---|
| Pleomorphic adenoma 8940/0 Myoepithelioma 8982/0 | Acinic cell carcinoma 8550/3 |
| Basal cell adenoma 8147/0 | Mucoepidermoid carcinoma 8430/3 |
| Warthin tumor 8561/0 | Adenoid cystic carcinoma 8200/3 |
| Oncocytoma 8290/0 | Polymorphous adenocarcinoma 8525/3 |
| Canalicular adenoma and other ductal adenomas 8149/0 | Epithelial-myoepithelial carcinoma 8562/3 |
| Sebaceous adenoma 8410/0 | Clear cell carcinoma 8310/3 |
| Lymphadenoma | Basal cell adenocarcinoma 8147/3 |
| Sebaceous 8410/0 | Sebaceous adenocarcinoma 8410/3 |
| | Secretory carcinoma 8502/3 |
| Non-sebaceous 8410/0 | Intraductal carcinoma |
| | Oncocytic carcinoma 8290/3 |
| Ductal papilloma 8503/0 | Salivary duct carcinoma 8500/3 |
| Sialadenoma papilliferum 8406/0 | Adenocarcinoma, NOS 8140/3 |
| Cystadenoma 8440/0 | Myoepithelial carcinoma 8982/3 |
| Soft tissue tumors | Carcinoma ex pleomorphic adenoma 8941/3 Poorly |
| Hemangioma 9120/0 | differentiated carcinoma 8020/3 Carcinosarcoma 8980/3 |
| | Squamous cell carcinoma 8070/3 |
| Hematolymphoid tumors | Lymphoepithelial carcinoma 8082/3 |
| Hodgkin lymphoma diffuse large B-cell lymphoma 9680/3 | Sialoblastoma 8974/1 |
| Extra nodal marginal zone | Secondary tumors |
| B-cell lymphoma 9699/3 | |
| Hodgkin lymphoma | |

**Ranula**

Ranula is derived from the Latin word ‘Rana,' which means frog, since it resembles a frog's underbelly. It develops as a result of trauma due to obstruction of the excretory ducts of the salivary glands situated either in the submandibular or sublingual space. [16] Epidemiologically, these lesions occur most frequently in the first two decades of life, and more-often prompts a conservative management approach.

**Myoepithelioma**

Myoepithelioma is a rare, benign salivary gland neoplasm. It mainly occurs in the mid-adult years which is equally seen in men and women.[17] Salivary gland tumors are rarely seen in children and adolescent. Myoepithelioma constitutes 1.8-10% of all salivary gland tumors occurs in children. [18,19] It frequently occur in the superficial lobe of parotid gland compared to minor salivary glands or in oral mucosa. The lesion present as a firm, circumscribed mass with an intact overlying epithelium. If the lesion is in parotid gland, superficial parotidectomy is required for diagnosis. For management, the tumor requires excision with 0.5cm margins peripherally and deep margins of periosteum or muscle fascia as appropriate.[17]

**Warthin Tumor (Papillary Cystadenoma Lymphomatosum)**

Warthin tumor is a benign hamartomatous or reactive proliferation of ductal salivary cells and lymphoid elements. It is commonly seen between 30-70 years of age. Men are more frequently affected than women with 3:1 ratio respectively. It is present in the superficial lobe of the parotid gland as a firm to doughy painless mass. It is frequently seen in the "tail" of the parotid gland, which is the most inferior and posterior portion of the gland below the ear and angle of mandible. A CT scan is required for diagnosis and location of tumor in the gland. Along with this, FNAC is also performed for quick and confirm diagnosis. Warthin tumor is excised with a superficial parotidectomy. [17]

**Pleomorphic Adenoma**

Pleomorphic adenoma is a benign neoplasm which has the ability to regrow if not completed excised, but is incapable to metastasis. It commonly affects the superficial lobe of the parotid gland as a painless, firm mass. Mucosa over the posterior and anterior hard palate are the site of predilection, when pleomorphic adenoma arises in the oral mucosa. It is usually seen during $4^{th}$- $6^{th}$ decade of life, and are slightly common in females.[17] However, pleomorphic adenoma affecting minor salivary glands in children are rare and is seen in 11-18 years of age.[20] CT scan or MRI scan is required for confirmation of the pleomorphic adenoma. Superficial parotidectomy should be carried out for diagnostic biopsy and definitive treatment. After confirmation of the lesion, it should be excised with 1 cm clinical margins at its periphery along with the overlying surface epithelium and periosteum of the palate.[17]

**Basal Cell Adenoma**

According to WHO, a basal cell adenoma is a benign tumor comprised of basaloid cells arranged with a strong basal cell layer and characteristic basement membrane-like structure, but without the myxochondroid stromal component observed in pleomorphic adenomas.[21] They are more prevalent in the parotid gland rather than oral mucosa. When it arises in the oral mucosa, it affects upper lip and rarely palatal mucosa. The lesion are painless, well-circumscribed masses, which is often observed in men and older adults of approximately 60 years of age. FNA should be performed for diagnosis. If the lesion is on oral mucosa, excision is carried out with 0.5 cm margins. [17]

**Oncocytoma**

Oncocytes are benign epithelial tumors. They are very rare, benign salivary gland tumor which exceptionally occurs in an oral mucosa. It is frequently seen on the superficial lobe of parotid gland as a slow-growing, painless, firm mass that is freely movable. It affects individual older than 50 years of age with no gender predilection. FNA is performed to rule put malignancy. Superficial parotidectomy is a treatment of choice. [17]

**Canalicular Adenoma**

Canalicular adenoma is an uncommon benign neoplasm of oral mucosal minor salivary glands. It occurs more commonly occurs in the upper lip submucosa as a painless, freely movable mass. This mostly affects females older than 50 years of age. Excision of mass with 0.5 cm margin along with overlying epithelium and muscle is diagnosis and treatment of choice.[17]

**Sialadenoma Papilliferum**

Sialadenoma papilliferum is rare benign neoplasm of minor salivary gland. It affects male rather than female in a ratio 1.5:1 in 50 years of life. It is a slow growing mass which do not exceed more than 2 cm. Excision of mass is required for diagnosis and definitive treatment.[17]

**Inverted Ductal Papilloma**

Inverted ductal papilloma is a rare benign neoplasm derived from the duct system of minor salivary gland [22] and is a proliferation of ductal epithelium of the excretory duct. It produces 0.5-2cm raised, nodular mass in the submucosa.[17] The site of predilection is buccal mucosa and upper lip, predominance among middle aged men.[23]

**Kuttner's Tumor**

Kuttner tumor, commonly known as Chronic Sclerosing Sialadenitis (CSS), is a salivary gland inflammatory illness first described by Kuttner in 1896. This condition, which affects the submandibular gland, is very rare and under-recognized as a cause of salivary gland enlargement. It can affect anybody at any age, ranging from 12 to 83 years old, however the majority of patients are in their third to seventh decades of life; the average age is 42–44 years old, with a small male predominance. Although research in Western and Japanese populations have proven that this is an IgG4-related illness, the specific cause is unclear. [24]

**Mucoepidermoid Carcinoma**

Stewart et al identified Mucoepidermoid carcinoma as a distinct entity within salivary gland neoplasia in 1945. It involves 50% of all malignant salivary gland tumors in children. [25] The most common malignant tumor in the parotid gland (15%) and the second most common in the submandibular gland.[26] Although this is the most frequent salivary gland neoplasm in children, it is uncommon in children under the age of ten. The most common symptom of a salivary gland tumor is a firm to hard mass that grows slowly. Unless there is pain, regional lymphadenopathy, or cranial nerve involvement, signs and symptoms may not be able to differentiate between malignant and benign tumors. [25]

**Adenoid Cystic Carcinoma**

Adenoid cystic carcinoma is the second most prevalent malignant tumor of the salivary glands, affecting both the major and minor glands. It is a slow-growing tumor with a high tendency for local invasion, recurrence, and distant metastasis. It is predominantly seen among women in the fifth and sixth decade of life. It is very rare among children. This was first introduced by Robin et al in 1853. In 1859, Billroth used the word "cylindroma" to characterize the cribriform appearance formed by tumor cells around cylindrical pseudolumina. Spies, on the other hand, proposed the widely recognized and commonly used name ACC in 1930. It is one of the most frequent malignant tumors of the minor salivary glands, which accounts for 22% of all salivary gland malignancies, with the palate being the most common location. This tumor of the small salivary glands is one of the most biologically destructive and unexpected tumors. [27] The perivascular and perineural tendency for infiltration is a distinguishing feature of the, which renders prognosis less favorable, with a 5-year survival rate of 60% and a 10-year survival rate of 40% in all age categories. [28] Mucoepidermoid carcinomas, adenoid cystic carcinomas, and acinic cell carcinomas account for 80-90 percent of all malignant lesions of the salivary glands in children. [28]

**Acinic Cell Carcinoma**

Acinic cell carcinoma is a rare, generally low-grade tumor that most commonly affects the parotid gland, followed by the oral cavity's minor salivary glands. Originally thought to be a benign organism, it was reclassified in 1953 after investigations revealed that it may reoccur and spread. Acinic cell carcinoma of the parotid gland accounts for 3% to 4% of all parotid gland tumors in children, as well as 3% to 30% of malignant parotid neoplasms. After mucoepidermoid carcinoma, it

is the second most common parotid malignancy in children. Large tumor size, high histologic grade, histologic type, facial palsy during diagnosis, discomfort, local invasion, rapidly growing tumor, and presence of regional or distant metastases, as well as past radiation exposure and family susceptibility, are all associated with a poor prognosis. The specific pathogenesis, however, is unknown. [29]

**Epithelial- Myoepithelial Carcinoma (EMC)**

Intercalated duct EMC is a low-grade biphasic salivary gland tumor with a proclivity for local invasion, recurrences, perineural involvement, and distant metastasis in certain cases. It is seen in the parotid gland in 80% of cases. Donath et al. invented the term EMC in 1972. [30] Since 1991, EMC has been recognized as a distinct entity under the WHO classification of salivary gland tumors. [30,31] It represents 0.5 percent of salivary gland tumors in women between the ages **6.1 Ranula**

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**Polymorphous adenocarcinoma (PAC)**

PAC is mainly associated with minor salivary glands. Freedman et al. identified and termed it as lobular

carcinoma in 1983 because of its similarity to lobular carcinoma of the breast. [32]

Batsakis et al. coined the term Polymorphous low-grade adenocarcinoma (PLGA) in 1984. This carcinoma is most commonly detected in the palatal region, although it has also been observed in other intraoral regions such as the upper lip, buccal mucosa, posterior tongue, and retromolar trigone in exceptional cases. It is most common in the sixth to seventh decades of life, with a few cases occurring in adolescence. It is the second most frequent kind of small salivary gland malignant tumor, followed mucoepidermoid carcinoma. [32] According to the World Health Organization's histological categorization of salivary gland carcinomas, cribriform adenocarcinoma of the small salivary glands is a low-grade, polymorphic adenocarcinoma. It predominantly affects females (70%) who are generally older than 60 years old, and it affects Africans and African-Americans more frequently. [33]

**Squamous cell carcinoma (S.C.C)**

SCC is one of the rarest types of malignant salivary gland tumors. Primary squamous carcinoma is most often seen in the parotid gland and extremely rare in the submandibular gland. Some of the possible causes include metastasis from distant primary squamous malignancy, direct invasion from cutaneous or mucosal squamous carcinoma, a squamous component of mucoepidermoid carcinoma, or primary squamous cell carcinoma of salivary origin. The latter is by far the most unusual. The parotid gland is the most common location in the major salivary gland. It is seen in 9% of all malignant parotid tumors but just 2% of malignant submandibular tumors. [34]

**6.17 Sarcoma**

Non-lymphoid soft tissue sarcomas of major salivary glands are extremely rare, according to Kauffman et al (1963). Most children with rhabdomyosarcoma are under the age of ten at the time of diagnosis. There is no racial predilection and the gender incidence is equal. The annual incidence of rhabdomyosarcoma is estimated to be 4 per million youngsters (Young et al, 1975). Age, histology, original location, degree of illness, and therapy all have an impact on the prognosis of rhabdomyosarcoma in children. Patients who appear before the age of two years have less progressed illness. Rhabdomyosarcoma is divided into four pathological types: embryonal, alveolar, pleomorphic, and mixed. [35] Rhabdomyosarcomas of the parotid and submandibular glands have the histological appearance of a skeletal muscle tumor, although they can develop in tissue that lacks striated muscular components. The most fatal forms of salivary gland tumor in children are rhabdomyosarcomas (RMS) of the parotid and submandibular glands. [36]

**Lymphoma**

Lymphoma in the salivary glands has the lowest incidence, accounting for just 1.7 percent to 5% of all salivary neoplasms. Non-lymphoma Hodgkin's of the B-cell follicular type is the most common kind of lymphoma of the parotid gland. It mostly affects men in their forties and fifties, and it's quite unusual in children. When it occurs in children or adolescents, unlike the persistent disease course of adult parotid follicular lymphoma, the prognosis is quite excellent when it is first treated with surgical excision. As a result, pediatric follicular lymphoma is defined as a single follicular lymphoma in an extra nodal location that has occurred in children (PFL). In children, primary malignant lymphoma of the parotid gland is identified in around 1.7% of cases. [37]

1. **Management**

The following are the guidelines for children salivary gland carcinomas from the Fracture (French Pediatric Rare Tumors) group. If at all feasible, multidisciplinary team consultations between adult and pediatric physicians should take place before any surgery or biopsy. In the instance of a salivary gland tumor in a kid, a head and neck ultrasound and MRI should be performed initially, followed by an FNA evaluated by a pathologist who is familiar with pediatric SGC. Only in situations of uncertain diagnosis, large expansive tumors, or anticipated mutilating surgery (including facial nerve section) should an initial Tru-Cut or surgical tumor biopsy be done. In every patient should have a wide tumor excision with clean margins, as well as frozen section analysis to ensure full resection. Modalities of surgery for parotid tumors are guided by the location of the tumor and its extension on MRI (e.g., total or superficial parotidectomy, facial nerve section followed, if possible, by facial nerve reconstruction). In the absence of clinical or radiologic problems, no preventive cervical nodal dissection should be performed. In situations with first clinical/radiological node involvement, simultaneously related node dissection should be performed at the same time as tumor excision. When there are macroscopic or microscopic remains, immediate surgical re-excision should be done, preferably without damage. If histologic node involvement or a high-grade tumor is discovered, a neck dissection should be considered. Complementary treatment with radiation must be addressed in a multidisciplinary conference on a case-by-case basis, taking into consideration the patient's age, grade of malignancy, and unfavorable concomitant pathologic characteristics (lymph node involvement, vascular invasion, perineural tumor extension). In situations of full resection following a low intermediate–grade SCG with no cervical node expansion, no systematic adjuvant treatment is advised. Except in rare cases of metastatic tumors, there is little evidence to support the use of chemotherapy in juvenile SGCs. [38]

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