CHAPTER 39
Salivary Gland Diseases in Children and Adolescents

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Introduction

Salivary glands are found in and around the oral cavity, and they are divided into major and minor salivary glands. The major salivary glands are the parotid, submandibular, and sublingual glands; the minor salivary glands are located in the lips, buccal mucosa, palate, and throat. Generally, salivary gland diseases are not common in the paediatric population. The classification of salivary gland diseases is very complex because it encompasses different entities; however, precise classification and terminology are necessary for accurate diagnosis and management. As in adults, diseases of the salivary glands may be nonneoplastic or neoplastic (tumours) (Table 39.1). The pattern of incidence in the paediatric population differs greatly from that in the adult group. Most salivary gland lesions in children are either inflammatory or vascular in origin. Of the developmental salivary gland diseases, haemangiomas are the most common. In the African paediatric population, mumps is the most common in the inflammatory/infection group, but in the developed world, only sporadic cases of mumps are now reported, and rhabdomyosarcomas are the most common nonodontogenic mesenchymal tumours in children.

Neoplastic changes in the paediatric population are very rare compared to the inflammatory groups. In the population as a whole, salivary gland neoplasms constitute 2.8% of all head and neck tumours, but in children it accounts for about 10% of all childhood neoplasms and between 3% and 22% of epithelial salivary gland neoplasms. The majority (88.5%) of salivary gland tumours are benign; the remaining 11.5% being malignant. In children, the most common benign epithelial tumour is pleomorphic adenoma, and the most common malignant tumour is mucoepidermoid carcinoma.

Salivary gland tumours in children have the same clinical and biologic behavior as those in the adult. The majority (76.7%) occur in the major glands, with the remainder in the minor glands, a ratio of 3.3:1. The ratio of occurrence of parotid to submandibular to sublingual tumours in the major salivary glands is 30:6:1. Globally, these tumours occur predominantly in girls and at any childhood age. A detailed clinical history with imaging features narrows the differential diagnosis while providing useful information for management and prognosis. Incisional biopsy must be avoided due to the possibility of tumour spillage and facial nerve damage.

The treatment of salivary gland diseases is categorised into medical and surgical, depending on the nature of the disease condition. The neoplastic lesions usually require surgical intervention, with or without radiation and chemotherapy, whereas the nonneoplastic/inflammatory diseases are managed symptomatically and conservatively. A protracted conservative medical management is strongly advised, however, before surgical ablation is considered in children.

Investigations

Salivary gland enlargement is a diagnostic challenge to the attending surgeon because the glands could be involved in a wide spectrum of diseases.

Table 39.1: Classification of salivary gland diseases in children.

Nonneoplastic tumours
Congenital/developmental
Agenesis/aplasia, hypogenesis/hypoplasia
Aberrant/ectopic salivary gland
Haemangioma
Lympangioma
Inflammatory and infection.
Acute sialadentis
Mumps, cytomegalovirus, Coxasackie A or B or parainfluenza virus)
Human immunodeficiency virus (HIV)-associated salivary glands
Recurrent parotitis in children (RPC)
Autoimmune
Sjogren's syndrome
Cysts
Ranula mucocele (mucous retention cyst)
Salivary gland dysfunction
Xerostomia
Sialorrhea/ptyalism
Neoplastic tumours
Benign
Pleomorphic adenoma
Warthin's tumour
Malignant
Mucoepidermoid carcinoma
Acinic cell carcinoma
Adenoid cystic carcinoma
Mesenchymal tumours
Neural tissue
Neurofibroma
Muscular tissue
Rhabdomyosarcoma

Investigative modalities in the diagnosis of salivary gland disorders in children are listed below. In an African setting, however, the cost of computed tomography (CT) and magnetic resonance imaging (MRI) systems usually limits investigations to ultrasonography and fine needle aspiration (FNA) cytology.

1. Ultrasonography (US) is useful in assessing the size of the gland and the vascularity of the lesion. It differentiates between a focal and diffuse disease, cystic and solid lesions, and is a useful adjunct for the assessment of adjacent vascular structures. US also guides FNA. The normal gland is hyperechogenic, whereas the diseased gland varies in hypogenicity.

2. A *CT scan* defines the nature and exact extent of the disease. It is useful in the diagnosis of acute inflammatory glands, abscess, and solid tumours. This is the imaging modality of choice in salivary gland diseases in children.

3. MRI defines the extent of the lesion similar to the CT scan. It is superior to CT, though, in that MRI also demonstrates the facial nerve within the parotid gland.

4. FMA cytology is a useful diagnostic tool. It can be used to establish whether a lesion is inflammatory benign or malignant. When used in children, however, there may be a need for sedation due to lack of cooperation. FNA cytology accurately diagnoses whether a lesion is benign or malignant in about 84–97% of the cases.

5. Sialography is very useful in the evaluation of autoimmune and chronic inflammatory diseases. The appearance is described as "cherry-blossom" or "branchless fruit laden tree" or '"snow storm".

6. Sialochemistry is useful in inflammatory and nonneoplastic diseases of the salivary glands. In inflammatory diseases, the IgA, IgG, IgM, albumin, transferring, lysozyme, Na, and protein are raised, and the phosphate level is decreased.

7. Sialometry is the determination of salivary flow rate. It is useful in the detection of salivary gland hypofunction.

8. Chest x-rays are used to detect any lung metastasis.

9. Radiographs of the jaws are used for the localisation of ectopic salivary gland tissues.

Congenital and Developmental Diseases

Aplasia (Agenesis) and Hypoplasia

Aplasia is the absence of any or a group of salivary glands; they could be unilaterally or bilaterally absent. Hypoplasia of the gland is reduced glandular tissue associated with hypofunction. Aplasia and hypoplasia are very rare, and only case reports have been documented.

Aplasia is of unknown aetiology; it may be isolated or occur in association with other developmental defects, such as hemifacial micostomia and the mandibulo-facial dysostosis (Treacher Collins syndrome), Down syndrome, and ectodermal dysplasia.

Aplasia presents with the development of xerostomia and its effects; however, other causes of xerostomia should be excluded. The effects of aplasia (agenesis) include dryness of the mouth, difficulty in mastication and swallowing of solid foods, an unusual pattern of dental caries, erosion of teeth, the presence of plaque, periodontal disease, soft tissue infection, chelitis, atrophic mucositis, and the absence of the salivary ducts. CT and MRI indicate the absence of the glands and replacement with fatty tissues; a salivary flow rate lower than 50% of its normal value is diagnostic.

Treatment is usually conservative; artificial saliva is used for frequent lubrication of the oral cavity. Comprehensive dental preventive and restorative therapy is strongly advocated.

Aberrant/Ectopic Salivary Gland

In aberrant/ectopic glandular anomaly, there is the presence of the salivary gland tissue in an abnormal location. This condition is rare in children, probably due to the difficulty in localising the lesion with a periapical radiograph in children and because it is usually asymptomatic. Locations include the tonsils, rectum, and the mandible. Inclusion of the gland in the angle of the mandible is seen frequently and referred to as Stafne's idiopathic bone cyst. Radiographically, this anomaly presents as a round or oval radiolucency between the mandibular canal and the inferior border of the mandible. The usual treatment is exploration of the cavity, whereas others believe it should just be kept under observation.

Haemangiomas

Haemangiomas are congenital malformations of the vascular system. They are the most common benign salivary gland lesion in children and the commonest benign tumour of children and adolescents. They have a female predilection. The majority are capillary in nature, occurring during the first year of life. Haemangiomas are usually soft masses noted shortly after birth, mainly in the parotid region, and rarely in the sublingual gland (Figure 39.1). They grow rapidly during the first year, with slow spontaneous complete regression at adolescence. They could be unilateral or bilateral.

At ultrasound, haemangiomas are hypoechoic relative to the gland, with a variable abnormal flow at Doppler US; contrast-enhanced CT reveals hypervascular mass with variable intensity of enhancement with an occasional demonstration of phleboliths. FNA cytology demonstrates elongated spindle cells arranged in coils and arcades.

Histopathologic analysis shows areas composed of an unencapsulated mass of closely packed, thin-walled capillaries with plump endothelial cells. Immunochemistry reveals vascular spaces lined by CD34 and factor VIII-positive flattened endotheliail cells.

Most capillary haemangiomas regress spontaneously, so surgical treatment should be delayed. Treatment options include injection of sclerosants, such as injection of boiling water normal saline, steroid, alfa 2a or 2b interferone (3 million units/m^2 per day, occlusion of feeder vessels, ligation of feeder vessels, surgical and laser ablation, or a combination of these modalities. Surgery is indicated in rapidly growing haemorrhagic tumours or following failure of the tumour to regress after achieving fibrosis postsclerotherapy.

Complications of haemangiomas include infection, ulceration, and occlusion of the larynx.

Figure 39.1: Haemangioma involving the parotid gland in a 13-year-old girl.

Lymphangiomas

Lymphangiomas are congenital malformations of the lymphatic system usually involving the parotid gland. They are the second most common nonneoplastic salivary gland tumours in children, occurring from birth to about 12 years of age, with a majority at 4 years of age and younger. About 65% are present at birth and 90% are detected during the second year, with a peak during the first decade. Lymphangiomas occur more in girls than boys. They present as a soft, asymptomatic swelling with facial asymmetry (Figure 39.2). Unlike haemangiomas, lymphangiomas rarely undergo spontaneous regression due to the extent of involvement and multispatial character of the lesion. Lymphangiomas are classified on the basis of the size of cystic spaces as simplex, cavernous, and venolymphatic.

Figure 39.2: Lymphangioma of the parotid gland.

Ultrasonography typically reveals thin-walled septations with occasional solid areas, CT shows a multispatial mass with heterogeneous septation and cystic areas often containing fluid levels; however, solid portions of the lesion may show enhancement. MRI demonstrates heterogeneous multiple cystic areas. Contrast-enhanced imaging may show enhancement of the solid portions of the lesion.

Treatment options for lymphangioma include intralesional injection of sclerosing agents; OK432 (picibalin), bleomycin, or surgical excision. Complications include infection and haemorrhage.

Parotid Swelling

Several nonspecific conditions are characterised by unilateral or bilateral enlargement of the parotid gland. Differential diagnosis of bilateral parotid swellings in a child may have the following aetiologies:

- viral (mumps, HIV-associated salivary disease);
- immunological (Sjogren's syndrome); and
- nutritional (obesity, hypervitaminosis A, beri-beri, hypoproteinaemia).

In addition, of unknown aetiology are hypertrophy of the masseter muscle and juvenile recurrent parotitis (JRP, also known as recurrent parotitis in children, or RPC).

Viral

Mumps

Mumps (epidemic parotitis) is now rare in the developed world due to the availability of the measles-mumps-rubella (MMR) vaccines, except for sporadic outbreaks that occur in adolescents and adults. However, in the developing world, it is the most common cause of parotitis in children, primarily affecting children younger than 15 years of age. Mumps frequently occurs as an epidemic between ages 5 to 15 years. It is usually contagious, with an attack conferring a lifelong immunity.

The aetiology of mumps is due to the paramyxovirus group with an incubation period of about 21 days. A similar clinical picture may present in Coxasackie A or B or parainfluenza virus. It is characterised by mild fever, malaise, and pain and sudden distention of the involved gland, usually the parotid gland. Initially, it involves one side, but within 3 to 5 days both glands become involved. The involved gland feels tensed and tender with congested punctum.

Mumps is a self-limiting disease. The treatment is primarily symptomatic: analgesic for pain, antibiotic to prevent secondary infection, and rehydration with adequate bed rest. Its postpuberty complications are orchitis and oophoritis in the male and female, respectively. Prevention is by the administration of the MMR vaccine.

HIV-associated salivary gland lesions

HIV-associated salivary gland lesions have become common in the African setting following the HIV/AIDS pandemic. The pandemic is a leading cause of immunodeficiency in infants and children. Lesions commonly involve the parotid glands; however, parotid involvement in the paediatric group is associated with a better prognosis. Typical lesions are of the benign lympyhoepithelial types and cystic. Other presentations are with xerostomia and sialorrhrea. On ultrasonography, 70% show multiple hypoechoic or anechoic areas, with the remaining 30% being anechoic. CT and MRI demonstrate bilateral parotid enlargement with intraglandular cystic and solid masses. No surgical intervention is needed, as resolution of swellings occurs following the administration of antiretroviral drugs.

Immunological: Sjogren's Syndrome

Sjogren's syndrome is an autoimmune disease characterised by mononuclear infiltration and destruction of the salivary and lacrimal glands. Two types are recognised: primary Sjogren's is a sialolacrimal disease without associated autoimmune disease, and secondary Sjogren's is a sialolacrimal disease with an autoimmune disease, usually rheumatoid arthritis.

Clinical features include those due to xerostomia with dryness of the eye and keratatis. Diagnosis is based on determination of the parotid rate lower than 1–2 ml/min; ultrasonography studies showing a snowstorm, cobblestone appearance; labial gland biopsy; detectable rheumatoid factor; antinuclear and antisalivary duct; and antithyroid antibodies. Histologically, it is characterised by infiltration, replacement, and destruction of the salivary and lacrimal glands by lymphocytes and plasma cells.

Treatment for Sjogren's syndrome is symptomatic, as for xerostomia: treat connective tissue diseases and keratoconjuctivitis with artificial saliva and ophthalmic lubricants.

Unknown Aetiology

Recurrent parotitis in children

RPC is a nonobstructive, nonsuppurative inflammatory disease of the parotid salivary gland of unknown aetiology in children, although congenital autoimmune duct defects are implicated. It is the second most common salivary gland disease in children after mumps. Clinically, it is characterised by a sudden onset of intermittent unilateral or bilateral parotid swellings over a period of years. The child is usually not ill, although there may be a mild rise in temperature; leucocytic count differentiates it from mumps. This lesion should also be differentiated from Sjogren's syndrome and HIV-associated salivary gland diseases. It predominates in male children between 3 months and 16 years of age with remission at puberty. There is usually a widening of the Stenson's duct with mucopurulent discharge.

Diagnosis is usually from parental history and a report of recurrent unilateral or bilateral parotid gland infections. Sialographic studies show a pattern of sialectasis similar to Sjogren's syndrome strictures, dilations, and kinks. Salivary chemistry is usually altered as it also is in adult patients: increased amounts of sodium, and protein, IgA, IgG, IgM, albumin, transferrin, and myeloperoxidase.

Treatment is conservative by lavage, ductal dilatation and hydrocortisone (100 mg) injection via sialoendoscopy, glandular massage salivary stimulation with sugarless sour candy, and antibiotics augmentin (25 mg/kg) or clindamycin (150 mg, 8-hourly, for 7 days). In an African setting, glandular massage of the parotid with antibiotics may be very helpful.

Masseteric hypertrophy

This is an asymptomatic bilateral enlargement of the masseter muscles as a result of hypertrophy. It is associated with bruxism and clenching of the teeth. Treatment usually involves debulking the masseter.

Cysts

Ranula

A ranula is a cyst-like soft swelling in the mouth. It presents as a translucent bluish colour under the tongue (Figure 39.3). The appearance is that of a frog's belly. Its aetiology is as a result of mucous extravasation of the sublingual gland following trauma, and obstruction or infection of the gland ducts, resulting in leakage and escape of secretion into the surrounding tissue. Simple ranula occurs when the extravasation is into the oral aspect of the mylohyoid muscle, whereas involvement of the hernited sublingual gland through the mylohyoid muscle results in a plunging ranula manifesting extraorally in the neck (Figure 39.4). Simple ranulas are noted more commonly in females, and plunging ranulas noted more frequently in males. CT scan demonstrates a cystic mass in the suprahyoid anterior neck.

Marsupialisation of the simple intraoral ranula involves deroofing of the cyst, suturing of its wall to the surrounding mucosa with packing of lumen. Plunging ranula is excised with the involved sublingual gland. Recurrence usually necessitates excision of the involved sublingual gland (see Figure 39.4).

Mucoceles

A mucocele (mucous extravasation cyst) develops mostly in children and young adults and mainly from the minor salivary glands following trauma and leakage of saliva into the surrounding submucosal tissue. The most common sites are the lower lip and inner aspect of the cheek, which are areas susceptible to trauma during oral function. A mucocele has no epithelial lining; rather, it is contained within a wall of fibrous and inflammatory tissue. It typically presents as a slow-growing and superficial soft wall fluctuant fluid containing mass of diameter 1.0–2.0 cm.

The overlying mucosa is usually of a translucent bluish color. A mucocele causes little or no discomfort. Marsupialisation of the cyst with overlying mucosa leads to recurrence of this pseudocyst. Occasionally, it ruptures spontaneously and forms again because of the accumulation of secretions beneath the healed surface. Excision of the cyst with the overlying mucosa lining is the treatment of choice.

Salivary Gland Dysfunctions

Xerostomia

Xerostomia is dryness of the mouth. The major cause in children is dehydration. Other causes are the use of antihistamine-containing drugs or decongestants; autoimmune diseases such as Sjogren's syndrome, sarcoidosis, and HIV; agenesis/aplasia or salivary gland hypofunction; and ectodermal dysplasia.

Symptoms of xerostomia include dryness of the oral cavity, burning sensation, soreness of the lips, difficulty with speech, difficulty with mastication and swallowing of solid foods, and altered taste sensations.

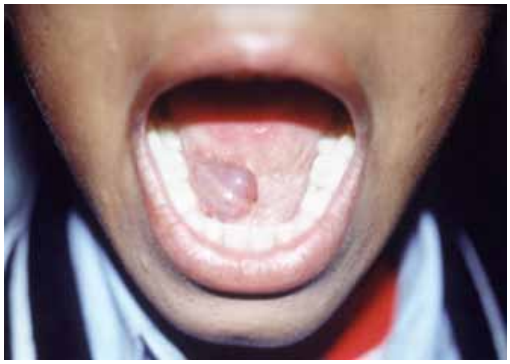

Figure 39.3: Simple ranula in a 6-year-old boy.

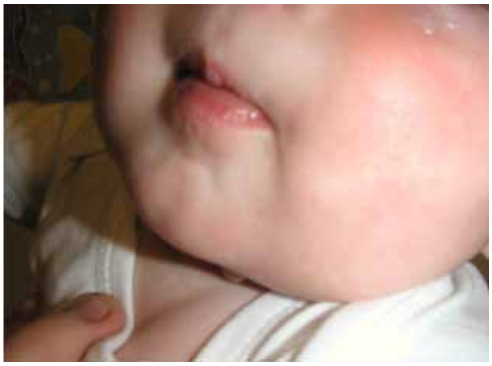

Figure 39.4: Plunging ranula.

Signs of xerostomia are dryness of the oral mucosa, mucositis, angular chelitis, dental plaques, dental smooth surface caries and demineralisation of the enamel, inflamed gingivae with periodontal diseases, and candidiasis. Diagnosis is based on history, clinical examination, sialometry (salivary flow is decreased), sialography, scintigraphy, sialochemistry (sodium and chloride lactofarrin levels), gland biopsy, and whole saliva immunotesting for antinuclear antibodies.

Treatment is conservative. The use of stimulants, sugarless candies and gums, artificial saliva; increased fluid intake; oral lubricants; and nonirritating toothpastes is recommended. Oral rehabilitation may be required to correct rehydration in some cases. Comprehensive dental management is strongly advocated.

Sialorrhea

Sialorrhea (ptyalism) is a persistent increase in salivary flow rate. It differs from drooling. In children, the most common cause is teething. Other causes are childhood epilepsy, HIV parotid enlargement, mental retardation, cerebral palsy, and herpes infection. In children, it requires the constant change of clothing and the use of bibs.

Treatment for sialorrhea is usually conservative, involving the use of anticholinergic agents (atropine) and antidepressants. However, surgical management such as parotid duct rerouting (Wilkie procedure), submandibular duct rerouting, tympanic neurectomy, or excision of the glands may be carried out when conservative management fails.

Neoplastic Epithelial Tumours

The parotid gland is the most common site of tumours (85.1%), followed by the submandibular (11.7%) and the sublingual (3.2%). In the minor salivary gland, the most common site is the palate. Overall, salivary gland tumours occur more in girls than in boys.

The majority of salivary gland neoplasms in children are benign. The pleomorphic adenoma is the most common benign neoplasm, and the most common malignant tumour is mucoepidermoid carcinoma. About 11.5–35% of salivary gland tumours in children are malignant, and 60–90% of these are mucoepidermoid carcinomas; adenoid cystic

and acinic cell carcinomas follow in frequency, each occurring at approximately 5–10%. The mainstay of treatment of salivary gland tumours is surgery.

Pleomorphic Adenoma

Pleomorphic adenoma (mixed tumour) is the most common childhood salivary gland tumour, occurring mostly in the parotid gland. Typical features are as a hard or firm or fluctuant, painless, slow-growing, freely mobile, bossellated mass. Facial nerve paralysis in association with pleomorphic adenoma never occurs, even in large grotesque swellings seen in Africans (Figure 39.5). The most common intraoral site is the palate, followed by the buccal mucosa and the lip (Figure 39.6).

In the minor salivary glands, the features include bossellated ulcerated swelling, causing ill-fitting dentures and difficulty in speech, which may occasionally erode the palatine bone. Ulceration is usually as result of trauma or following topical application of herbal medication. No childhood age is exempt, with a median of 15 years from some studies, occurring predominantly in females.

At ultrasound, the pleomorphic adenoma varies from hypoechoicity to isoechoicity relative to the rest of the gland, with occasional hyperechogenic foci due to some calcifications within the mass. CT and MRI demonstrate varying findings depending on the tumour size. Small lesions are homogenous with well-defined margins, whereas larger lesions are more heterogeneous with less well defined margins. FNA cytology confirms the benignity of this tumour.

Microscopically, the pleomorphic adenoma tumour is composed of varying proportions of glandular-like epithelium and connective tissue stroma. Epithelial cells may show nests, solid sheets, or ductal structures with varying stroma, which may be myxoid, chondroid, fibroid, or osteoid with some areas of squamous metaplasia and foci of keratin.

Treatment is parotidectomy (superficial or total) with facial nerve sparing. In the submandibular gland; treatment is submandibulectomy. In the minor salivary glands, wide local excision with a circumscribed incision of 3–5 mm of apparent normal tissue is made around the tumour. There is a high recurrence, usually due to enucleation of the tumour.

Parotidectomy procedure

1. The external auditory canal is blocked with a pledget of cotton wool to prevent blood from entering into the ear canal.
2. A lazy S incision starts anterior to the ear helix, extends posteriorly over the mastoid bone, and curves anteriorly parallel to the angle of the mandible or running along the submandibular incision line up to about 3 cm.
3. A skin incision is raised, exposing the superficial parotid fascia, and is retracted and sutured down (Figure 39.7).
4. The mastoid, the anterior border of the sternocleidomastoid muscle, and the posterior belly of the digastrics muscles are identified. Finger pressure is applied medial to feel for the styloid process.
5. The posterior belly of the digastrics muscle is retracted, exposing the facial nerve as it exits from the stylomastoid foramen (see Figure 39.7).
6. The nerve is followed into the parotid gland, where it divides into its five terminal branches under the superficial lobe.
7. For superficial parotidectomy, the gland is dissected off the nerve. For total parotidectomy, the superficial lobe is removed and the deep lobe is excised after elevating the nerve.
8. The surgical site is irrigated and a suction drain is inserted.
9. The wound is closed in layers and a compression dressing is applied (Figure 39.8).

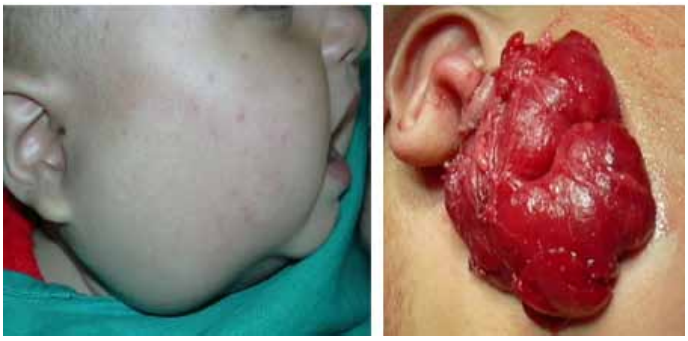

Figure 39.5: Pleomorphic adenoma of the parotid gland

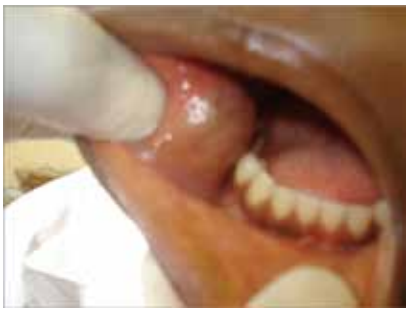

Figure 39.6: Pleomorphic adenoma of the buccal mucosa.

Figure 39.7: Skin incision exposed and sutured down. The posterior belly of digastrics and facial nerve are identified with forceps.

Figure 39.8: Wound closure with an in situ drain.

Complications of parotidectomy

An immediate complication of parotidectomy is bleeding. Delayed complications include haematoma, infection, transient facial nerve paralysis, facial nerve paralysis, Frey's syndrome, salivary fistula, and recurrence.

Warthin's Tumour

The second most common benign salivary gland neoplasm in children is Warthin's tumour (papillary cystadenoma lymphomatosum, or adenolymphoma). This occurs mainly in the parotid gland. It is a slow-growing, painless swelling, usually unilateral with multiple masses in the gland. Developmentally, there is incorporation of the lymphatic element, salivary gland tissue, and the paraparotid lymph nodes.

CT and MRI demonstrate a well-circumscribed, homogenous cystic or solid lesion in the parotid gland.

Grossly, the tumour appears as multiple cysts of varying diameters containing viscous fluid with solid areas of lymphoid follicles. Microscopically, the tumour comprises tall eosinophilic columnar cells with papillary projections into cystic spaces in a background of lymphoid stroma.

Treatment is surgical excision (parotidectomy) of the tumour with preservation of the facial nerve.

Carcinomas

Acinic Cell Carcinoma

Acinic cell carcinoma (also known as acinic cell adenocarcinoma or acinous cell carcinoma) is a malignant epithelial neoplasm of salivary glands demonstrating serous acinar cell differentiation. It is second in occurrence to mucoepidermoid carcinoma in the paediatric population. It is of varying malignancy, accounting for 6–37% of the total malignancies in children. It is more common in girls than boys, and 4% of the patients are younger than 20 years of age.

The majority occur in the parotid gland. It typically presents as a painless, slow-growing, enlarging solitary mass in the parotid, occasionally multinodular and fixed to the skin and underlying tissues. It is occasionally painful, with associated facial nerve paralysis.

The treatment modality is parotidectomy (superficial or deep) with preservation of the facial nerve. Nodal and distance metastasis with recurrence may occur.

Mucoepidermoid Carcinoma

Most studies agree that mucoepidermoid carcinoma (mixed epidermoid and mucus-secreting carcinoma) is the most common salivary gland malignancy in children. The majority are in the major salivary glands, with about 45% in the parotid. The most frequent intraoral sites are the palate and the buccal mucosa. It has a female predilection, and occurs in the 5- to 15-year-old age group, peaking between 12 and 14 years of age.

The clinical features depend on the grade of the tumour. They are classified as low, intermediate, or high grade. In children, the majority are of the low-grade variety with a very good prognosis. The low-grade presentation may simulate a benign tumour. Otherwise, it typically presents as a rapidly growing swelling with perineural and soft tissue invasion. High-grade tumours are usually found in children younger than 5 years of age.

Diagnosis is based on clinical, radiological, and pathological findings. The low-grade type shows smooth "benign appearing" margins at US, CT, and MRI, containing cystic low attenuation and occasional calcified foci at CT. The high-grade type is more solid and homogenous at both CT and MRI.

The cut surface may contain cystic or solid areas, or both. Cystic areas contain viscous or mucoid materials. Microscopically, this tumour contains two types of cells—mucous and epidermoid cells; the proportion determines the grade of the tumour. The low-grade tumour is characterised by prominent cystic structures and mature cellular elements; the intermediate-grade tumour has fewer and smaller cystic structures and occasional solid islands of epidermoid cells; and the high-grade tumour is a hypercellular solid tumour with cellular atypia and high mitotic figures.

The treatment of mucoepidermoid carcinoma is surgery, depending on the grade, location, and tumour extent. Wide local excision is adequate for low-grade tumours. Block composite excision with radical neck dissection is appropriate for the intermediate- and high-grade varieties. This may involve excision of the gland and facial nerve; petrosectomy; mastoidectomy; or resection of the ramus of the mandible, zygomatic arch, and the overlying skin, with subsequent nerve grafting and facial reconstruction. In the palate, palatoalveolectomy, maxillectomy with or without radiotherapy, and chemotherapy are used postoperatively. Radiation should be used with caution in children due to the long-term effects. Tumour grading and subtype have a tremendous predictive effect on the prognosis.

Rhabdomyosarcoma

Rhabdomyosarcoma is the most common childhood soft tissue sarcoma, usually of the embryonic subtype. About 40% of all rhabdomyosarcomas arise in the head and neck region, and the parotid and the adjacent structures are usually involved by direct invasion. It is common between 1 and 13 years of age, with most patients dying a few months after initial diagnosis. Prognosis is usually poor because of late presentation (Figure 39.9).

CT demonstrates heterogeneous attenuation of the tumour, and MRI shows a poorly defined, heterogeneous mass. The treatment is wide surgical excision of the tumour and adjacent structures with reconstruction of the lost tissues. For advanced inoperable tumours, as is commonly seen in the African population, palliative chemotherapy is recommended.

Neurofibroma

Neurofibroma is a benign nerve sheath tumour that may arise from the facial nerve within the parotid gland. It is rarely in the submandibular gland. It may be solitary with gross facial disfigurement (Figure 39.10) or have multiple lesions, as seen in von Recklinghausen's disease. Clinically, it feels like a bag of worms. CT demonstrates it as well-demarcated, homogenous, and isoattenuated relative to muscle. It shows moderate enhancement following injection of contrast media. Occasionally, it may push the larynx or the trachea, causing respiratory difficulty (Figure 39.11). MRI may show low to intermediate signal intensity.

Histologically, neurofibroma consists of loose collagen fibre with long, thin processes of nerve fibre and darkly stained elongated nuclei. The treatment is surgical excision. Recurrence occurs because of inadequate excision.

Evidence-Based Research

Table 39.2 presents a study of surgical procedures on tumours of the salivary glands in children and adolescents.

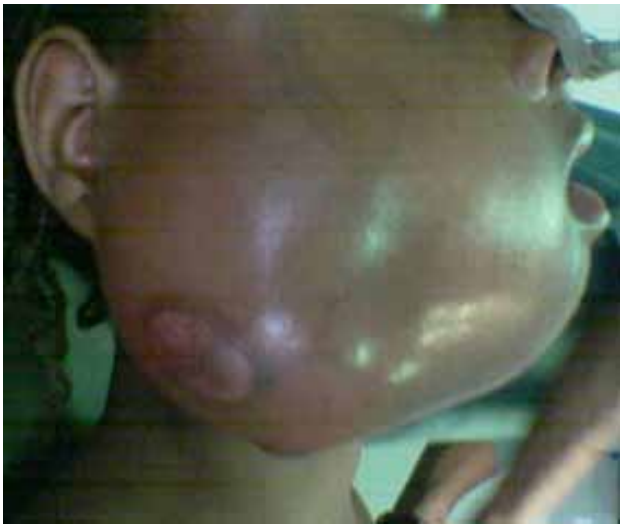

Figure 39.9: Rhabomyosarcoma in a 15-year-old girl.

Figure 39.10: Neurofibroma involving the parotid gland in a 16-year-old girl.

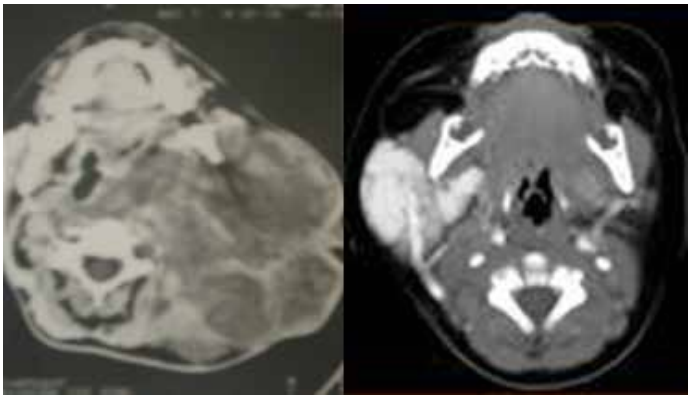

Figure 39.11: CT scan of neurofibroma pushing the airway in a 16-year-old girl.

Table 39.2: Evidence-based research.

Title	Tumours of the salivary glands in children and adolescents
Authors	Ellis M, Schaffranietz F, Arglebe C, Lawskawi R
Institution	Department of Otorhinolaryngology, Head and Neck Surgery, Universitats-HNO-Klinic, Germany
Reference	Int J Oral Maxillofac Surg 2006; 64:1048–1058
Problem	To examine the outcome and side effects of therapy and recurrence of 52 salivary gland tumours in juveniles and adolescents.
Intervention	Surgical procedures.
Outcome/ effect	Forty benign and 12 malignant tumours underwent various surgical procedures. For the benign tumours, 20 cases (50%) were parotidectomies; 2 (5%) of these with facial nerve graft, 14 (35%) had lateral parotidectomies, 2 (5%) had subtotal parotidectomies, 3 (7.5%) had submandibulectomies, and 1 (2.5%) had enucleation. For the malignant tumours, 11 (91.75%) had total or radical parotidectomies; 4 (33.3%) had total parotidectomies with excision of facial nerve and facial nerve grafting with the greater auricular nerve; 5 (41.7%) had radical parotidectomy with facial nerve and adjacent structures (ascending ramus, temporomandibular joint, zygomatic bone, and mastoid); 2 (22.2%) had parotidectomies with neck dissection; and 1 (11.1%) had superficial parotidectomy.
Historical significance/ comments	This report of 52 cases of salivary gland tumours demonstrates the various types of salivary gland tumours in the parotid region, provides a useful practice guide, and attests to the rarity of these tumours in the paediatric population. Irrespective of the type of tumour, surgery is the preferred mode of treatment. However, the postoperative complications of the parotidectomy procedure should be noted. The authors recommend a multidisciplinary approach involving the paediatrician, the surgeon, the radiotherapist, and the oncologist. The findings of this report have important implications for the African setting in that primary surgery is most effective and the surgeons and paediatricians are readily available, whereas the radiotherapists and oncologists are not. Furthermore, in the African setting, microscopic surgery for nerve grafting is still elusive.

Key Summary Points

1. Salivary gland tumours in children are rare.
2. Most salivary gland lesions in children are either inflammatory or vascular in origin.
3. Classification of salivary gland diseases is complex but important for management planning.
4. Major glands are more readily affected.
5. In Africa, investigation may be limited to ultrasound and fine needle aspiration.
6. Most lesions are self-limiting or require medical management.
7. Surgical management is reserved for tumours and nonresolving lesions.
8. Facial nerve damage is the major morbidity in salivary gland surgery and should be avoided.

Suggested Reading

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