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Dent Clin N Am 49 (2005) 241–258

THE DENTAL
CLINICS
OF NORTH AMERICA

Pediatric soft tissue lesions

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Acute lesions

Bacterial infections

Most infections in children are caused by bacteria [1]. Toddlers, as part of their cognitive and psychologic development, transition through an oral phase during which their perception of the surrounding environment is primarily mediated through the mouth. As a result, oral homeostasis during the early years of life goes through constant changes and frequent imbalance, occasionally leading to infection [2].

Acute necrotizing ulcerative gingivitis

Acute necrotizing ulcerative gingivitis (ANUG) is an acute periodontal emergency that has been described in children [3]. The condition has been linked to elevated levels of spirochetes and *Prevotella intermedia*. Among the pre-existent conditions that can affect the development of the disease, malnutrition and immune deficiency are particularly relevant [3]. The clinical characteristics of ANUG include painful ulceration involving the entire gingiva, focal necrosis of the interdental papillae, and hemorrhage. Oral hygiene is deficient because of the intensity of the discomfort, and halitosis is present with the necrotic debris. Certain children might develop a concomitant viral infection, such as herpetic gingivostomatitis, which complicates the diagnosis because of overlapping signs and symptoms from both conditions.

Management of ANUG relies on initial débridement and antibiotic therapy to decrease the severity of symptoms. Effective periodontal debridement is difficult to achieve in a symptomatic child. Initial therapy

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doi:10.1016/j.cden.2004.07.008 dental.theclinics.com

should include topical anesthetic solutions to avoid interruption of nutritional intake [3]. Parents most often seek professional care when the child is dehydrated and showing signs of confusion. Antibiotic therapy (penicillin V 25–50 mg/kg/d) is effective in resolving the acute symptoms. Periodontal débridement and scaling should follow the initial treatment.

Bacterial sialadenitis

The most common salivary gland complication is acute bacterial sialadenitis [4]. Sialadenitis in children occurs as a result of dehydration [5–7] or obstruction of a major salivary gland. Commonly affected glands include the submandibular and sublingual glands because of the tortuous anatomy of their ducts and anatomic position [8,9]. The parotid glands are also affected in cases of sialolithiasis (salivary stone) secondary to severe dehydration or systemic disease. Extraoral swelling and painful lymphadenopathy can be present as well.

Management of acute sialadenitis underscores the importance of an accurate evaluation and diagnosis. Major salivary glands should be palpated to verify proper flow. A pathognomonic sign of acute infection is the expression of purulent exudate from one of the ducts [10]. Pain on palpation is common, indicating an inflammatory process. Management of sialadenitis involves proper hydration and antibiotic therapy to cover Staphylococcus and Streptococcus spp. Imaging such as an occlusal radiograph or ultrasound often can rule out a sialolith. The clinician is cautioned regarding radiographic evaluations, because many calcified sialoliths are not detected by conventional radiography until they are 60% to 70% calcified. If an obstruction is suspected, CT and ultrasound are valuable diagnostic tools [10]. Severe obstruction of the gland is evidenced by exquisite tenderness and no absence of saliva on palpation. Extraoral and intraoral swelling is a common sign (Fig. 1). Management of severe obstruction usually involves surgical intervention, especially when the obstruction is close to the gland.

Mucocele

Two salivary gland soft tissue abnormalities are prevalent in children. Mucous extravasation phenomenon or mucocele is a common finding in young individuals who traumatize their oral mucosa, causing the obstruction of minor salivary glands. The lesions are commonly seen on the lower labial mucosa (80%) and in less degree on the buccal mucosa [1]. Superficial lesions might develop in the soft palate and retromolar areas. Most mucoceles are fluctuant and have a white to bluish hue depending on the extension of the lesion into surrounding tissues and time of onset [11]. Chronic lesions can appear fibromatous because of repetitive injury. Size can range from a few millimeters to up to 1 cm. A common presenting sign is a change in size associated with meals.

Another acute salivary gland lesion is the ranula, which is a form of mucous retention or extravasation phenomenon located in the floor of the

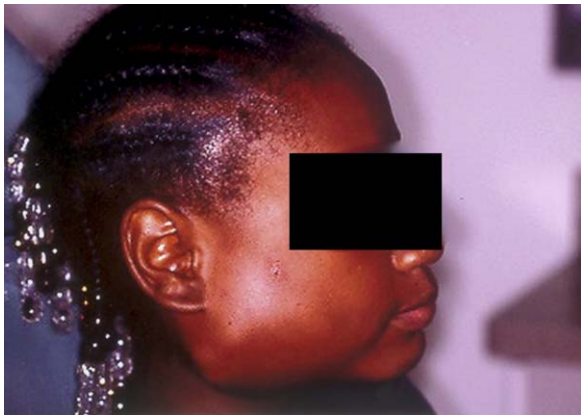

Fig. 1. Parotid enlargement in an 11-year-old child.

mouth, commonly lateral to the midline. It is caused by leakage of mucin as the result of trauma to a salivary gland. The sublingual gland is commonly affected, followed by the submandibular and minor salivary glands [3,11]. Clinically, ranulae present as a smooth domelike swellings, pink to bluish in color and ranging in size from 1 to several centimeters. Occasionally, ranulae can become infected by oral flora. Treatment for ranulae is surgical, with removal or marsupialization of the lesion.

Viral infections

Primary herpetic gingivostomatitis

Primary herpetic gingivostomatitis is a common condition observed in young children and infants. It is caused by herpes simplex virus 1 in the majority of cases, although herpes simplex virus 2 also can present similar characteristics. The incidence of primary herpetic infection increases after 6 months of age, reaching a peak between 24 and 48 months (Fig. 2) [12]. The disease is characterized by an abrupt onset of malaise, irritability, and fever, followed by the appearance of generalized oral lesions. The prodromic phase usually lasts 1 to 3 days before the outbreak of the oral ulcers. The clinical presentation includes generalized gingival ulceration and hemorrhage with cervical lymphadenopathy. Some cases may present with perioral lesions.

Management of primary herpetic gingivostomatitis is symptomatic because the infection is self limiting. Dental care providers can prescribe local anesthetics and antibacterial rinses during the acute phase of the infection. Mucosal-coating agents such as milk of magnesia or kaopectate temporarily relieve symptoms and can be used as often as needed. Therapy is focused toward maintaining adequate hydration and oral intake during the acute phase of the infection. Immune-compromised children are treated with antiviral medications, such as acyclovir [12].

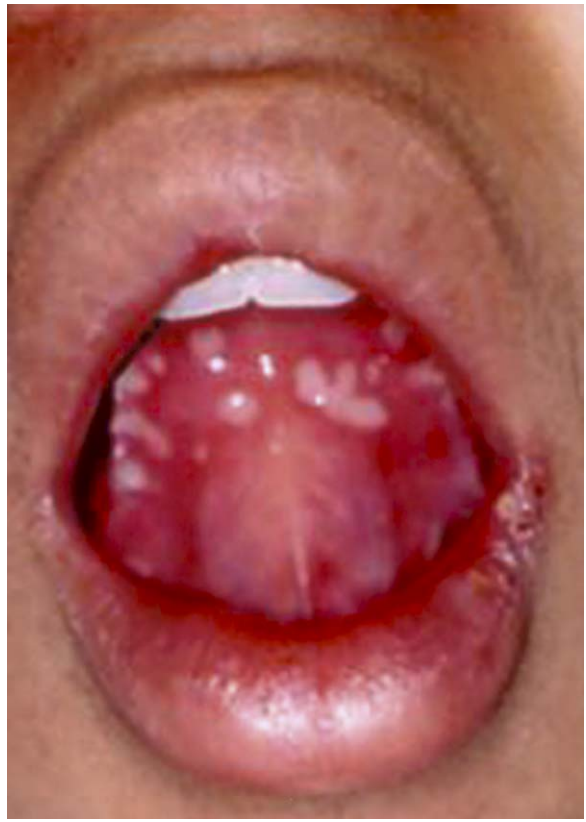

Fig. 2. Primary herpetic gingivostomatitis in a young child. (Courtesy of Dr. Thomas P. Sollecito, Philadelphia, PA.)

Herpangina

Herpangina is an acute viral disease characterized by the appearance of oral ulceration limited almost entirely to the soft palate, uvula, tonsils, and fauces. Members of the coxsackie family (coxsackie subtypes A) have been implicated in herpangina, specifically subtype A_6. Incidence of the disease peaks during the initial months of summer and fall [13]. Clinical presentation includes sudden fever, sore throat, headache, dysphagia, and malaise followed in 23 to 48 hours by the onset of erythema and vesicular eruption. The small vesicles rupture, leaving shallow, painful ulcerations that heal in 7 to 10 days (Fig. 3). Management of herpangina is palliative.

Acute lymphonodular pharyngitis

Acute lymphonodular pharyngitis is an acute febrile disease with oral manifestations. Coxsackie virus subtype A_{10} has been identified as a common cause [13]. The oral lesions are limited to the posterior pharynx,

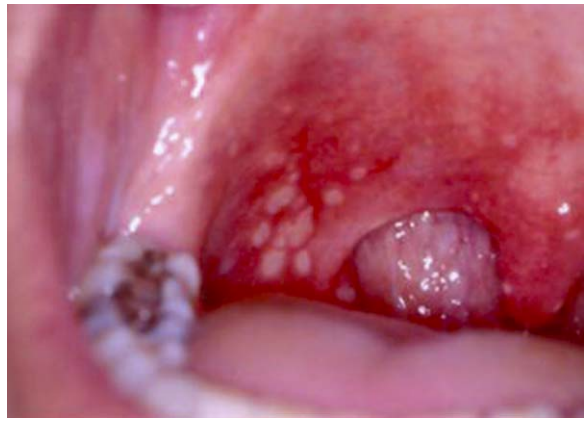

Fig. 3. Herpangina. Multiple shallow ulcers in oropharynx. (Courtesy of Dr. Thomas P. Sollecito, Philadelphia, PA.)

soft palate, uvula, and fauces and usually have a more diffuse presentation than herpangina, with more widely spread nodular characteristics. Clinical presentation involves prodromic symptoms including malaise, fever, and headache, followed by the onset of papular lesions surrounded by an erythematous halo. Treatment of this condition also is palliative, with concomitant use of topical anesthetics and coating agents.

Hand-foot-and-mouth disease

Hand-foot-and-mouth disease is a viral disease frequently seen in epidemic outbreaks in day care or school-aged children [3,13]. Coxsackie virus A_{16} has been associated with this infection. The incubation period ranges from 3 days to 1 week before onset of prodromal symptoms. Presenting symptoms are mild headache and malaise, followed by onset of skin and oral lesions. Skin lesions are predominantly on the hands and feet, affecting lateral and dorsal areas of fingers and toes, palms, and soles. As in many other viral diseases, skin lesions are vesicular in nature surrounded by an erythematous halo. Oral involvement, described as vesicular eruptions with shallow ulcers that may coalesce into major palatal and pharyngeal lesions, occurs in almost all cases. The dentist should include this entity among other differential diagnoses when evaluating a child with acute lesions in the soft palate or pharyngeal lesions. The presence of limb lesions is pathognomonic of this disease. Management of oral complications involves palliation and adequate hydration.

Varicella

Varicella or chickenpox is a benign, eruptive infection almost entirely limited to children [14]. This highly contagious disease as caused by the

varicella zoster virus, a member of the herpesvirus family. Varicella is characterized by a prodromic period of fever and headaches, followed by the onset of dermatologic and oral lesions. The preferred sites for occurrence are the head, face, and trunk. Skin lesions are maculopapular, with formation of vesicles and pustules that rupture, leaving hard, brown-crusted lesions. The crusts eventually exfoliate leaving mild to moderate scarring. The hard and soft palate and labial and buccal mucosa are common sites for appearance of oral lesions, similar to infections by other herpesviruses. Oral lesions of chickenpox are small vesicles that rupture leaving shallow round ulcerations surrounded by an erythematous halo. Management of varicella is symptomatic. Antiviral agents such as acyclovir can be used in the early stages of the disease. A vaccine against the varicella zoster virus is readily available and should be given to infants between 12 and 18 months of age.

Measles

Measles or rubeola is another acute, highly contagious viral disease observed in children. It is caused by a paramyxovirus and is transmitted by droplets through the respiratory tract. Measles has an incubation period of 8 to 12 days [15]. There is an initial period of 3 to 4 days followed by the onset of a prodrome consisting of fever, chills, cough, myalgia, and ocular manifestations. The average age of infection is 4 years, with variations depending on geographic and demographic characteristics of the community. The dermatologic lesions appear as a maculopapular eruption initially on the face, spreading to the neck, trunk, and limbs. The classic oral lesions, Koplik's spots, appear commonly on the buccal mucosa opposing molar teeth and are described as white linear lesions on an erythematous background [15,16]. Other oral presentations include petechiae, erythema, and multiple round, shallow ulcerations. Management of the oral ulcers is symptomatic during the progression of the infection. Oral manifestations of measles are rarely a cause of severe dysfunction. A vaccine exists for measles, mumps, and rubella (MMR), given at or after 12 months of age with a second dose given at 4 to 6 years of age.

Mumps (acute viral sialadenitis)

Mumps is an acute viral infection that involves the parotid glands and, less commonly, the submandibular and sublingual glands. It is caused by a paramyxovirus that is incubated for 16 to 18 days. Systemic symptoms appear parallel to the infection, including low-grade fever, chills, headache, malaise and sore throat. One or both parotid glands become tender and swollen for approximately 1 week. Pain elicited on chewing and beneath the angle of the jaw is a common finding. Frequent complications include orchitis and epidydimitis (painful testicular inflammation). Some cases have reported pancreatitis as a complication [17]. Treatment is mainly symptomatic, including adequate hydration. The MMR vaccine forms part of the routine vaccination schedule for young infants.

Heck's disease

Many viral dermatoses have oral manifestations. Focal epithelial hyperplasia or Heck's disease is a benign, viral-induced hyperplastic lesion of the oral mucosa [18]. It is caused by human papilloma viruses 13 and 32. Lesions are described as multiple, painless, sessile, slightly raised papules and nodules ranging from 1 to 10 mm in diameter. The lesions are whitish or slightly pink. If confined solely to the oral mucosa, the lesions might disappear when the tissue is stretched. Regression of the lesions occurs after a long period of time, usually taking months to years, with no specific documented pattern. Treatment for this disease consists of surgical removal of the lesions when cosmetics are of concern.

Trauma

Trauma is by far the most common cause of oral lesions in children and is probably the single most important complaint in emergency dental visits. Multiple factors play a role in traumatic injuries in the young patient.

Trauma to the oral soft tissue is usually caused by the teeth, either by direct biting of the oral mucosa or secondary to an accidental event during play or exercise [19]. Traumatic ulcers can have multiple presentations and are usually easily identified as opposing a sharp cusp or at the level of the occlusion. Occasionally labial ulcerations appear secondary to unintentional lip biting after local anesthetic [20]. Teeth, sharp dental restorations, orthodontic braces, and foreign bodies can induce trauma to the oral soft tissues. The ulceration presents as a single, ill-defined painful area with a smooth surface and erythematous or whitish borders. Chronic ulcers may have signs of scarring and present with fibrinous pseudomembranes. Bite injuries are described as stellate, irregular, unilateral or bilateral areas opposing the culprit teeth. Children with various genetic disorders (ie, Lesch-Nyhan disease) can engage in self-injurious behavior, which poses a serious challenge to the treating physician and dentist [21,22].

Chemical or electrical injuries to the mouth (caused by biting or exploration of an electrical cord) are common in children under the age of 6 years [23]. The lesions affect mainly the lips and the perioral skin and are described as painless, white-gray coagulated areas with no clinical hemorrhage surrounded by a narrow erythematous halo. The central area progressively necroses, leaving a deep ulcerated base. Sequelae of this injury range from local scarring to dental necrosis and microstomia with commissure involvement [24].

Treatment for traumatic soft tissue injuries is symptomatic. Local anesthetic solutions and coating agents can be prescribed to relieve pain. It is important to avoid prescribing oral rinses to children below the age of 5 years, because their ability to "rinse and spit" is limited. Acute lesions can be treated with topical application of medium- to high-potency steroids if the objective is to hasten the resolution of the ulcer. High-potency steroids should

not be used on the external vermillion of the lips or facial skin. Most traumatic ulcers heal within 7 to 10 days if the offending agent is resolved. Children with behavioral or genetic disease might need further intervention with intraoral appliances and psychologic or psychiatric therapy as appropriate.

Chronic and recurrent lesions

Chronic lesions that involve the oral mucosa may be arbitrarily classified as genetic or dermatologic disease. This section describes important chronic mucocutaneous disease that can be observed in the pediatric population. Certain diseases not mentioned in this section are reviewed in depth in other articles in this issue.

Juvenile bullous pemphigoid

Juvenile bullous pemphigoid is a chronic autoimmune disease of children [25–28]. The precise cause of the condition in children is not known, but it is assumed to be similar to the adult version of the disease, with subepithelial bullae formation. This condition is rare, and it usually presents in children older than 5 years. Although the skin is affected in the majority of cases, the oral mucosa can also be affected. The oral lesions are described as bullae that rupture leaving painful erosions. Oral lesions are not as profound as the dermatologic lesions.

The disease runs a 3- to 4-year natural history with progressive improvement [27,28]. Management of the oral lesions includes topical high-potency steroids, systemic steroids for concomitant dermatologic lesions, and immune modulating medications in severe cases. The use of dapsone has been reported in case studies, but safety concerns in the pediatric population still limit its use to recalcitrant patients.

Childhood linear IgA disease

Childhood linear IgA disease is an autoimmune vesiculobullous condition observed in children characterized by deposition of IgA along the dermoepidermal junction [28]. Onset of linear IgA disease is usually before the age of 5 years, and the skin lesions are symmetrical in distribution in the perioral, genital, or limb area. Dermatologic lesions are characterized by vesicular eruption accompanied by annular erythema. Oral lesions are observed in up to 50% of patients, following the onset of the dermatologic lesions. Oral lesions have been described as vesicular eruptions followed by ulceration [24,28]. The disease usually disappears by the second decade of life.

Treatment for linear IgA is systemic steroid therapy, dapsone, and sulfapyridine. The oral lesions can be managed with topical application of high-potency steroids for brief periods of time. In most cases, systemic administration of steroids controls the oral lesions. The use of

immune-suppressing steroid-sparing therapies is still controversial because it can predispose young patients to malignancy.

Hereditary epidermolysis bullosa

Epidermolysis bullosa (EB) refers to a group of inherited disorders characterized by blister formation as a result of mechanical trauma [29,30]. Hereditary EB is classified in three major types according to the ultrastructural characteristics of skin damage (epidermolytic, junctional, and dermolytic). The suspected cause involves defective collagen metabolism [30]. The severity of EB can range from mild to severe, localized or generalized. Cutaneous lesions include blisters, crusted erosions, milia, cicatricial alopecia, nail dystrophy, and mitten deformity of the hands or feet. Oral lesions have been identified in up to 92% of patients. Many oral changes occur as sequelae to the chronic blistering and scarring. Palatal atrophy and microstomia are the most prevalent sequelae, followed by ankyloglossia and lingual atrophy [31–33]. Among the many somatic expressions of the disease, reports of osteopenia and endocrine dysfunction raise a periodontal concern. Oral leukoplakia and malignancy (squamous cell carcinoma) have also been reported in EB patients [33].

Treatment of EB is based on minimizing cutaneous trauma, supportive care, effective wound management, prevention and treatment of infections, and nutritional support [29]. Skin blisters are aseptically punctured and drained to avoid extension of the lesion with expanded ulceration. Recent advances in dermatology allow the in vitro fabrication of skin grafts for these patients [30]. Recent research focuses on genetic repair of defects observed in specific variants of EB. The oral lesions are managed with intralesional steroid injections for severe ulceration and topical application of high-potency formulations for localized disease. Short bursts of systemic corticosteroids are useful to control acute symptoms. Successful use of azathioprine, intravenous immunoglobulin, and plasmapheresis has been reported. Progressive scarring of the oral mucosa with concomitant stricture complicates the provision of dental care.

Juvenile dermatitis herpetiform

Juvenile dermatitis herpetiform, also known as Duhring-Brocq disease, is a chronic recurrent vesicular dermatologic disease. There is a strong genetic association with multiple HLA subtypes (B8, DR3, DW3, DQW2). Autoimmunity is believed to play a major role, and gluten hypersensitivity has been closely linked to the appearance of the disease in children [34]. The onset of the disease is after the age of 5 years, characterized by symmetric erythematous pruritic papules or plaques, followed by burning papular and vesicular eruptions in a herpetic distribution. Affected areas include the extensor areas of the elbows and knees, shoulders, buttocks, posterior neck,

and scalp. Oral lesions are described as maculopapular or vesicular, which rupture, leaving painful ulcerated erosions [35].

Treatment of this condition consists of elimination of gluten in gluten-associated cases, sulfones, tetracycline, and nicotinamide. These last two medications have been used successfully in the treatment of other oral mucosal ulcerations, although their mechanism of action is still unclear.

Kawasaki disease

Also known as acute infantile febrile mucocutaneous lymph node syndrome, Kawasaki disease was initially described in 1967 [36]. The disease is a multisystem vasculitis of small and medium-sized arteries, predominantly affecting children under the age of 5 years, with a peak incidence between 1 and 2 years of age. There is an increased interest in the apparently infectious origin of the syndrome, because of the characteristics and natural history of the disease. Identification of a specific antigen has been elusive, however, and researchers are still attempting to identify such agent. Genetic factors seem to play a major role in the susceptibility of certain groups of patients to develop the disease.

Kawasaki disease is diagnosed by the appearance of five of the following six symptoms [37]:

Fever of 5 or more days' duration that is refractory to antibiotics and acetaminophen
Bilateral nonexudative conjunctival injection
Oral cavity involvement
Polymorphous rash
Peripheral extremity involvement (edema or desquamation)
Acute cervical adenopathy greater than 1.5 cm in diameter

Coronary artery involvement is a serious complication of this disease. Other otolaryngologic manifestations may occur, including facial palsy, otitis media, and cervical abscess. Oral changes include lip fissuring, strawberry tongue, and mucosal hyperemia, and are observed in almost all patients affected with Kawasaki disease (Fig. 4).

Management of the disease involves administration of intravenous immunoglobulin and aspirin. Most cases respond to initial therapy, although several treatments might be needed in recalcitrant cases. Management of the oral complications is mainly symptomatic [36]. Topical labial moisturizers and antibiotics prevent infections of eroded areas. Severe oral ulceration is uncommon in these patients. Topical anesthetics, coating agents, or steroid formulations are used for management of glossitis [36].

Vitiligo

Vitiligo is an acquired, progressive disease characterized by depigmentation of the epidermis (leukoderma) or, infrequently, by partial loss of

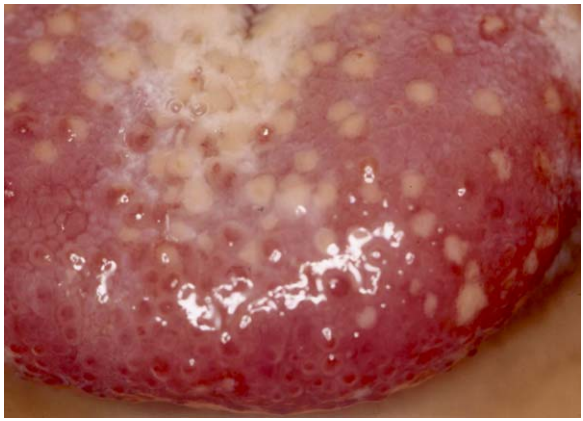

Fig. 4. Strawberry tongue in Kawasaki disease. (Courtesy of Dr. Thomas P. Sollecito, Philadelphia, PA.)

melanin (hypopigmentation). Vitiligo occurs as the result of the destruction of pigment cells in postnatal life by mechanisms that are still unknown [38]. The disease presents as depigmented patches present anywhere in the body, most often seen in the face, backs of hands and wrists, axilla, umbilicus, nipples, genitalia, and around body orifices (eyes, nostrils, mouth, genitalia) [38]. Vitiligo in children presents in the face and neck in up to 50% of patients. Perioral vitiligo is particularly relevant to the oral health provider, because it can be confused with cheilitis or traumatic scarring.

Although not all cases of vitiligo need to be treated, esthetic and self-esteem issues in children play a role in determining the need for therapy. Treatment of vitiligo consists of topical high- and ultra–high-potency steroids and psoralin-UVA. Dentists should be involved in the identification of the condition when it affects the perioral area.

Familial acanthosis nigricans

Cutaneous acanthosis nigricans is a hyperkeratotic skin disorder characterized by verrucous hyperplasia and hypertrophy of the skin with hyperpigmentation and accentuation of skin markings in the flexural side of joints. The onset of the disease may occur during childhood or puberty. The hyperpigmented plaques commonly appear on the axilla and neck. Hyperkeratoses of palms and soles and papillomatous growths on eyelids, lips, and oral mucosa have been reported (oral mucosal involvement is reported in up to 25% of cases). Oral lesions are usually not pigmented, unlike the skin lesions [39]. The disease is inherited as an irregular autosomal dominant trait.

Treatment for oral lesions is limited. Surgical options are available for disfiguring labial lesions, and keratolytic therapy might be useful.

Common pediatric soft tissue tumors

Congenital hemangiomas

Congenital hemangiomas most often arise in the neonatal period. Congenital hemangiomas occur in approximately 10% to 12% of infants by the age of 1 year [40]. The natural history of these hemangiomas can be divided in three phases: the initial or proliferative phase, the involuting phase, and the involuted phase [40,41]. Hemangiomas can appear on the buccal mucosa, alveolar mucosa, and gingiva. Two forms of hemangiomas are described intraorally: capillary (consisting of multiple capillary vascular spaces) and cavernous (multiple dilated sinuses filled with blood). Capillary hemangiomas are the most common. They appear clinically as a bright erythematous area that may be become macular, resolving spontaneously in 2 to 3 years. The cavernous hemangioma presents as an elevated dark-red lesion. This hemangioma rarely regresses spontaneously [41]. The most common complication of hemangiomas is hemorrhage.

Treatment for these soft tissue tumors is surgical if they are persistent. Removal can be partial, depending on the anatomic structures that are involved [41]. Alternative approaches include the use of sclerosing agents or embolization of involved vessels. Capillary hemangiomas, if asymptomatic and present in areas of low trauma, are observed for regression.

Pyogenic granuloma

Pyogenic granuloma presents as a reaction (granulation tissue) to chronic irritation observed in the gingival and periodontal area [42]. Although uncommon, it may present on the tongue, lips, and buccal mucosa. The pyogenic granuloma appears as a nodular mass that can be pedunculated. The lesion is asymptomatic unless it becomes infected. It may present with a lobulated or ulcerated appearance. Treatment of this common lesion is surgical excision with removal of the suspected cause [42].

Congenital epulis

Congenital epulis, also known as granular cell tumor, is a rare tumor of the newborn. Congenital epulis seem to originate from mesenchymal tissue and present as protruding masses from the alveolar ridge [43–45]. The tumor has a marked female prevalence of 8:1. The preferred site for development of the lesion is the maxillary alveolar ridge, with a wide variation in size from a few millimeters to almost 8 cm [44]. Some tumors regress spontaneously, but most are removed surgically. Recurrence is uncommon.

Lymphangioma

Lymphangiomas are not considered true tumors and are classified as developmental malformations [46]. Approximately 50% of congenital

lymphangiomas are present are birth, and almost 90% develop during the first 3 years of life. The most common location in the mouth is the dorsum of the tongue, followed by the lips, buccal mucosa, soft palate, and floor of the mouth. Neck lesions are also common [47]. The lesions consist of dilated lymphatic vessels that are lined by thin endothelium located below the epithelial surface. Lymphangiomas are classified in three forms based on tumor size: capillary, cavernous, and cystic hygroma. Cystic hygroma is a large facial or cervical swelling caused by a lymphangioma. The clinical appearance of lymphangiomas depends on the extension of the lesion. Superficial lesions consist of elevated nodules with pink or yellowish color. Deeper lesions are described as soft, diffuse masses with normal color. Treatment for lymphangioma is surgical, but recurrence is common.

Riga-Fede disease

Riga-Fede disease or Riga-Fede granuloma is a benign, ulcerative, granulomatous process that occurs as a result of repetitive trauma to the oral mucosa by teeth [48,49]. Lesions begin in early infancy and may be an initial sign of a developmental dysfunction or underlying neurologic disorder. Common sites for the granuloma are the ventral surface of the tongue or the lingual frenulum along the midline, corresponding to the offending teeth. Clinically, the lesion is an exophytic, ulcerated, granulomatous mass covered by a yellowish pseudomembrane. Appearance of the lesion coincides with the eruption of primary teeth, usually the lower incisors. Riga-Fede disease has been linked to familial dysautonomia, Lesch-Nyhan disease, and Gaucher's disease. The designation of this condition is specific to infants and children younger than 2 years of age [48].

Treatment of this disorder is aimed toward minimizing trauma to the affected area. Interventions include dental extraction, occlusal adjustment of sharp incisal edges, behavior modification, surgical excision of the lesion, and the use of intraoral appliances to cover the offending teeth [49].

Melanotic neuroectodermal tumor of infancy

Melanotic neuroectodermal tumor of infancy is a rare neoplasm occurring almost exclusively in infants during the first year of life. It is thought to derive from neural crest cells and is associated to elevated levels of vanillyl-mandelic acid [50]. The maxilla is the most commonly affected site, followed by the calvarium and mandible [51]. The tumor is considered malignant in only 4% of cases but is locally aggressive, destroying bone and displacing primary teeth.

The treatment of melanotic neuroectodermal tumor if infancy is surgical bloc excision conserving vital structures. The recurrence rate is as high as 25%. Alternate therapies such as radiation or chemotherapy have limited applications.

Pediatric oral cysts

Gingival cysts of the newborn

Gingival cysts of the newborn present as whitish nodules on the alveolar ridge of neonates [42]. These lesions are small (1–3 mm), keratin-filled cysts that regress within weeks of appearance. Other lesions that present in the hard palate have similar appearance (Epstein's pearls). There is no treatment for these cysts, because they regress spontaneously.

Eruption cyst

Eruption cyst is associated with an erupting deciduous or permanent tooth. The cause for this condition is the separation of the dental follicle from around the crown of the tooth [3]. The most commonly involved teeth are molars and canines. The lesion presents as a soft, well-demarcated swelling overlying the crown of an erupting tooth. Variations in color may exist depending on the amount of blood contained within the cyst. Treatment for an eruption cyst often is not necessary because it ruptures, allowing the tooth crown to continue its eruption.

Developmental cysts

Thyroglossal duct cyst

The thyroglossal duct cyst is rare, and 30% of cases occur in patients under the age of 10 years. At 4 weeks of gestational development, the thyroid tissue is formed and migrates to its final position in the neck. Remnants of this process are responsible for the cyst formation. The thyroglossal duct cyst commonly presents as a soft, fluctuating mass in the midline of the neck area, in the space between the suprasternal notch and the foramen cecum [24]. These cysts might be attached to the hyoid bone or tongue, interfering with deglutition. Intraoral occurrence is rare, and it localizes on the posterior dorsum of the tongue. Treatment of the cyst consists of surgical removal.

Dermoid cyst

The dermoid cyst is an uncommon developmental lesion rising from embryonic epithelial remnants [52]. It is seen along the midline of the floor of the mouth, but other oral and perioral areas can be affected [52]. Dermoid cyst is described as a slow-growing, painless swelling, with a soft doughy consistency on palpation. It may cause superior displacement of the tongue. Treatment of dermoid cyst is surgical excision.

Incisive papilla cyst

The incisive papilla cyst is a soft tissue lesion derived from epithelial rests of the nasopalatine duct [3,42]. The cyst causes soft tissue swelling in the

incisive papilla area. Color ranges from normal to bluish-red, and it is usually asymptomatic. The incisive papilla cyst is easily differentiated from hard tissue lesions and dental pathology by a radiographic examination in which no bony abnormality is noted. Treatment consists of surgical removal.

Soft tissue manifestations of systemic disease

Oral manifestations of systemic disease vary according to the organ system affected by the disease. Some relevant systemic conditions that can present with oral lesions are described here.

Hematologic disease

Congenital neutropenia

Congenital neutropenia is a disorder characterized by a constant decrease of neutrophils in the peripheral blood [53]. Oral lesions vary with neutrophil count. Severe ulceration that is secondarily infected is common in neutropenic children. The susceptible host can harbor viral and bacterial disease, with oral sequelae such as periodontal disease, fungal infection, and virally associated lesions.

Idiopathic thrombocytopenic purpura

Thrombocytopenia involves decreased platelet number. Oral manifestations are described as petechiae, ecchymoses, and hematomas appearing early during the disease. The oral mucosa is particularly susceptible to bleeding secondary to minor trauma in these patients.

Autoimmune disorders

Many autoimmune diseases have oral manifestations. Some examples include lupus erythematosus, Crohn's disease, and HIV infection [54]. The appearance of an unusual lesion in a young child should prompt a thorough work-up because oral lesions can be the initial signs in some cases of autoimmune disease.

Oral lesions in syndromic disease

In some syndromes, such as Behçet's syndrome, oral lesions are presenting and diagnostic signs. Other disease complexes in which oral lesions are present are the metabolic deficiencies (ie, lipid storage deficiency) and syndromes such as the periodic fever, aphthous stomatitis, pharyngitis, and cervical adenitis (PFAPA) syndrome [55–57]. Patients with PFAPA syndrome presumably have aphthouslike ulceration.

Another syndrome, the mouth and genital ulcers with inflamed cartilage (MAGIC) syndrome, has oral lesions as a presenting and diagnostic finding [58]. Description of the lesions observed in these patients is vague.

Summary

A variety of soft tissue lesions can be found in the young population. Because of the importance of an adequate nutrition, many of these diseases or lesions can create a serious concern for the infant and young child. The dental provider should be familiar with the common soft tissue lesions seen in children to provide appropriate management and referral when necessary.

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