

A review of oral pathology in orthodontics. Part 1: Soft-tissue pathology

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Orthodontists are well placed to detect soft-tissue disease of the oral cavity and associated structures because of the frequent repeat examinations of their patients. This review describes the clinical manifestations, pathologic features, and treatment of the soft-tissue pathology most likely to be encountered by the orthodontist and uncommon soft-tissue disease with significant implications for the patient. The recognition of soft-tissue disease will allow reassurance, referral, and early intervention when required. (*Am J Orthod Dentofacial Orthop* 2024;165:7-17)

Orthodontists are likely to be the dental clinicians who frequently assess the oral cavity of children and young people. Many patients' first full jaw imaging will be requested by an orthodontist, and the soft-tissue examinations carried out throughout treatment can detect asymptomatic lesions. In addition, several of the most common oral soft-tissue entities are caused by trauma, with orthodontic appliances being a potential source of this trauma. In the past, most patients undergoing orthodontic treatment in the United Kingdom were children and young adults. On average, orthodontists in the United Kingdom start treatment in <30 adult patients a year.¹ However, an increasing number of adults are undergoing orthodontic treatment, widening the range of diseases that may be encountered.² This review discusses the common soft-tissue pathology an orthodontist will likely encounter and the uncommon but important disease that presents in children, young adults, and adults (Table). The pathology of the jaw bones and the management of soft- and

Table. Summary of pathologic entities discussed in this review categorized into reactive, inflammatory, or infective, benign neoplasms, and malignant neoplasms

<i>Reactive, inflammatory, and infective</i>	<i>Benign neoplasms</i>	<i>Malignant neoplasms</i>
Mucocele	Squamous papilloma	Salivary gland malignancies
Fibrous hyperplasia and fibroepithelial polyp	Hemangioma	Rhabdomyosarcoma
Pyogenic granuloma	Pleomorphic adenoma	Ewing sarcoma
Frictional keratosis	Lipoma	Squamous cell carcinoma
Recurrent aphthous stomatitis		
Herpes simplex infection		
Geographic tongue		
Gingival overgrowth and gingival hyperplasia		
Oral candidiasis		
Orofacial granulomatosis and Crohn's disease		

hard-tissue pathology by the orthodontist will be discussed within Part 2 of this series of papers.³

COMMON ENTITIES

The most commonly biopsied soft-tissue diseases in children aged <16 years include mucoceles, fibrous hyperplasia, fibroepithelial polyps, pyogenic granulomas, squamous papillomas, peripheral giant cell granulomas, hemangiomas, and frictional keratosis.^{4,5} In addition, though infrequently biopsied, recurrent aphthous stomatitis (RAS), herpes simplex infections, geographic

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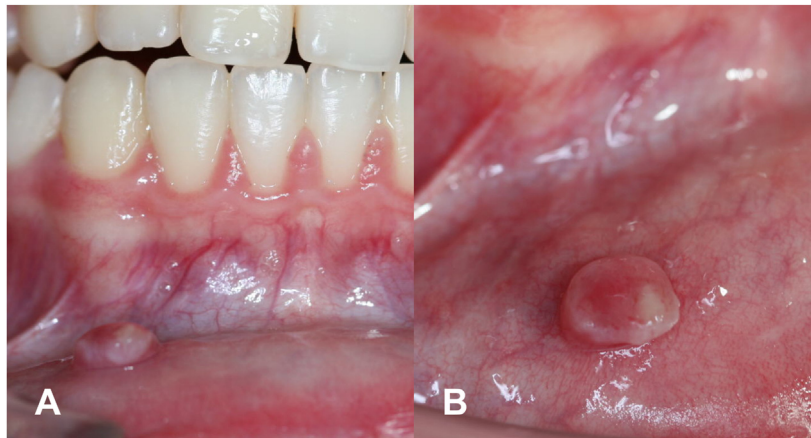


Fig 1. An example of a mucoccele: **A**, Intraoral photograph of a mucoccele on the lower right labial mucosa, identified after removal of fixed appliances; **B**, Intraoral photograph showing the lesion, which is polypoid, mucosa colored, and approximately 4 mm in diameter.

tongue, and gingival overgrowth (gingival hyperplasia) are commonly reported.^{6,7} Moreover, these conditions are all commonly seen in adults.⁸ The above entities will be discussed below, except for giant cell granulomas, which will be discussed in the bone pathology section.

Mucocelles

Of the commonly biopsied soft-tissue pathologies, mucocelles are most likely to be encountered,^{4,5} with mucocelles being more common in children and young adults than in older adults.⁹ They tend to present on the lower lip as a fluctuant blue, purple, gray, or mucosa-colored lump (Fig 1). Most have a history of trauma and are <1 cm in diameter on average.⁹ Less frequently, they are seen at other sites of the oral cavity but rarely at the upper lip,⁹ where neoplasia is a more common cause of swelling. Approximately 90% of mucocelles are mucus extravasation reactions on histologic examination, with the remainder being mucus retention cysts.¹⁰ The histologic features of a mucoccele include a lumen containing mucin, neutrophils, and macrophages, a lining of either compressed granulation tissue (mucoccele) or ductal epithelium (mucus retention cyst), and a fibrous wall often with a chronic inflammatory infiltrate.¹⁰ When a mucus extravasation reaction affects the sublingual gland, a large swelling in the floor of the mouth may form, which is described as a ranula.¹¹ Although some mucocelles will resolve without intervention, the mucoccele and the associated gland most often must be removed to resolve the lesion,¹² regardless of the mucocelles' location in the oral cavity.

Fibrous hyperplasia and fibroepithelial polyp (fibroma)

Fibrous hyperplasia refers to reactive lesions comprising increased and often densely collagenous fibrous connective tissue.¹³ When a distinct polyp is formed, these lesions are described as fibroepithelial polyps (fibromas). These lesions are relatively common in children, comprising 35.8% of biopsied mucosal pathology in children,⁵ although they are more common in adult patients.¹³ Most lesions occur on the tongue or buccal mucosa; both sites are prone to trauma.¹³ Fibroepithelial polyps are well-demarcated sessile or pedunculated polyps that can be soft to firm, mucosa colored, and generally <1 cm (Fig 2).¹³ Histologic examination shows a mass of densely collagenous fibrous connective tissue covered by epithelium that may exhibit hyperkeratosis and acanthosis.¹⁴ Inflammation is not present unless the lesion is ulcerated, and metaplastic bone formation may be seen if the lesion is present on the gingivae.¹⁴ When bone or calcifications are present, these lesions are sometimes referred to as peripheral ossifying fibroma. The term "giant cell fibroma" is sometimes used when large stellate or multinucleated fibroblasts are seen in the connective tissue of a fibrous polyp, although their presence is of unknown clinical significance. Excision of these lesions is curative¹²; however, a new lesion may form when trauma persists.

Pyogenic granuloma

Most (80%) of pyogenic granulomas occur on the gingivae.¹⁵ They appear as either sessile or pedunculate erythematous masses and are often soft and



Fig 2. An example of fibroepithelial polyps: **A** and **B**, Intraoral photographs showing fibroepithelial polyps on the right and left buccal mucosa. Both polyps are pedunculated and whiter than the surrounding mucosa, likely because of hyperkeratosis; **C**, After the removal of the Twin-block appliance (the source of the trauma causing the polyps), the polyps have almost completely resolved.



Fig 3. An example of a pyogenic granuloma: **A**, An intraoral photograph showing a pyogenic granuloma (*white*) on the patient’s palatal gingivae in association with the transpalatal arch; **B**, A further intraoral photograph showing the same pyogenic granuloma with the transpalatal arch removed. The lesion is an erythematous, pedunculated mass; **C**, An intraoral photograph showing almost complete resolution of the pyogenic granuloma 3 months after the removal of the transpalatal arch (the source of the trauma causing the pyogenic granuloma).

compressible (Fig 3). They are rarely >2.5 cm in diameter and may be ulcerated.¹⁶ Generally, they are asymptomatic but can be infrequently associated with bone

loss.^{15,16} Histologically, these lesions are polypoid, comprising granulation tissue in a lobular pattern with overlying ulceration.¹⁶ Pyogenic granulomas develop



Fig 4. An example of a squamous papilloma. This lesion on the patient's right dorsal tongue is a pedunculated polyp <5 mm in diameter; it did not recur after excision.

in response to either chronic local irritation or as a result of hormonal changes.^{15,16} As such, pyogenic granulomas are common in adolescence and may be seen when orthodontic appliances are causing mucosal trauma.¹⁷ If the source of irritation or trauma is removed, recurrence is uncommon after the excision of these lesions.¹⁵

Squamous papilloma

Squamous papilloma is the most common benign epithelial entity in the oral cavity of both adults and children.¹⁸ It is driven by low-risk human papillomavirus infection and is usually present on the tongue or palate.¹⁸ Clinically, it appears as a fronded, pedunculated lesion, <1 cm in diameter and is usually white but may be mucosa colored (Fig 4).¹⁸ The histologic features match the clinical presentation with exophytic fronds of hyperplastic, hyperkeratotic epithelium supported by vascular cores.¹⁸ Excision of these lesions is curative.

Hemangioma

Of all connective tissue tumors, hemangiomas are the most common in the oral cavities of children and young adults.^{4,5} Moreover, they are one of the most common tumors in children.¹⁹ The clinical manifestation of hemangiomas varies with the type of hemangioma. Infantile hemangiomas are the most common, with congenital hemangiomas being rare.¹⁹ They appear as well-demarcated, red, expansile lesions and tend to involute over time; most resolve by 7 years old.¹⁹ Some



Fig 5. An example of a vascular malformation on the upper lip. This extraoral photograph demonstrates a large vascular malformation with a vascular appearance and a somewhat purple color. It would be pulsatile on palpation.

hemangiomas may be large and persist into adulthood (Fig 5). They often blanch with pressure. Many hemangiomas are managed conservatively and are allowed to involute; however, when a lesion is likely to have a significant cosmetic or functional impact, it may be treated with surgery or medications such as propranolol.¹⁹

Frictional keratosis

Frictional keratoses are common, with a prevalence of 0.26%–5.3% in children and young adults,^{20,21} although they are more common in adults.²¹ They are reactive lesions in response to chronic trauma.²¹ Given the reactive etiology, frictional keratoses typically lack sharp demarcation. The clinical appearance of these lesions is variable and depends on the extent of the trauma. They range from faint whitening of the mucosa to extensive thickening and roughening of the surface with an opaque white color.²¹ Moreover, these lesions may be associated with erosions of the mucosa or ulceration in which the trauma has led to loss of the epithelium.²¹ A common presentation is bilateral white lines along the occlusal plane described as *linea alba*.²¹ The histologic appearance comprises hyperkeratosis, epithelial hyperplasia, and possible edema with ballooning of the prickle cell layer keratinocytes. Few inflammatory cells will be observed in the epithelium and lamina propria.²¹ Frictional keratosis will resolve by removing the source of trauma, for example, by smoothing a restoration or trimming the distal ends of orthodontic archwires.

Recurrent aphthous stomatitis

RAS is a common noninfectious ulcerative condition of the oral cavity with a lifetime prevalence of approximately 40%²² that disproportionately affects young adults.^{22,23} RAS causes recurrent painful ulcers that may present as single or multiple lesions across the oral cavity.²⁴ They usually heal over a few weeks.²⁴ There may be a period of months or sometimes years among bouts of ulcers.^{24,25} It is uncommon for these ulcers to appear on the keratinized mucosa,²⁴ whereas ulcers caused by herpes simplex virus (HSV) often occur on the keratinized mucosa of the gingivae or palate. The ulcers are shallow, with a yellow base and an erythematous rim (Fig 6).^{24,26} Although the causes of RAS are poorly understood,²⁵ there are many suggested predisposing factors.²⁶ These include family history, trauma, some drugs, iron or B12 deficiencies, hormonal changes, or stress.²⁶ However, the evidence to support these causative factors is weak.²⁶ RAS is classified into minor (most cases), major (the minority of cases), and herpetiform (rare) types depending on the size, number, and location of the ulcers.²⁴⁻²⁶ RAS is usually diagnosed from a patient's history and a clinical examination of the ulcers present²⁶; histologic examination of tissues is rarely helpful.^{24,25} However, further investigation by an appropriate specialist may be valuable to rule out systemic causes of oral ulcers such as Crohn's disease or celiac disease.^{24,25} Moreover, this specialist will be able to instigate the management of the patient's RAS; the goal of treatment is symptomatic relief as RAS is not curable.²⁵ If possible, topical agents are more appropriate as they have fewer systemic adverse effects.^{25,26} Generally, this comprises topical corticosteroids to reduce the inflammation driving ulcer formation, benzydamine hydrochloride for analgesia, and chlorhexidine or tetracycline mouthwashes, which may reduce the duration of ulcers.^{25,26} If topical therapy fails or a patient's RAS is severe, systemic treatments such as oral corticosteroids including prednisolone or immunomodulatory agents may be provided.^{25,26} However, there is limited evidence supporting any one option²⁷; therefore, the potential adverse effects from systemic therapies must be cautiously weighed against the potential benefits.

Herpes simplex virus infections

HSV infections are very common, with 25%-30% of children estimated to be affected by primary herpetic gingivostomatitis (PHG).²⁸ Approximately 30% of young adults have recurrent herpes labialis (RHL),²³ and although less common than RHL, recurrent herpetic stomatitis (RHS) may also be observed.²⁹ Most infections

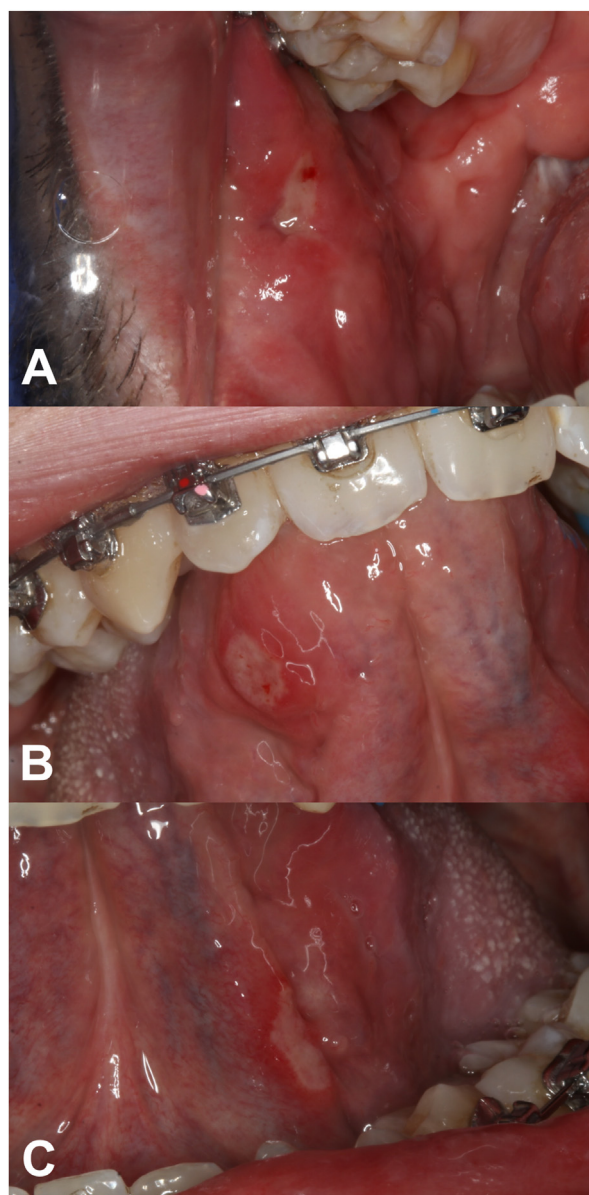


Fig 6. An example of minor recurrent aphthous stomatitis: **A-C**, Intraoral photographs showing aphthous ulcers on the right buccal mucosa and right and left ventral tongue. Each ulcer has a yellow to gray base and erythematous rim.

are caused by HSV-1 followed by HSV2.²⁸ PHG prodromal symptoms include fever, nausea, malaise, and headache,^{28,29} with additional symptoms of salivation, pain, halitosis, and lymphadenitis as the clinical lesions develop.^{28,29} PHG presents as a combination of gingivitis and ulcers.^{28,29} The ulcers begin as multiple small vesicles on the oral mucosa, pharynx, or perioral skin, breaking down and coalescing into larger irregular ulcers.^{28,29}



Fig 7. An example of recurrent herpes labialis. This extraoral photograph shows crusted lesions on the vermilion mucosa and perioral skin. These lesions are actively infective at this stage of the clinical course.

The lesions usually resolve completely within 2 weeks.^{28,29} RHL results from reactivation of HSV, which lies quiescent in sensory neurons, often of the trigeminal nerve. Usually, the perioral skin and vermilion mucosa are affected.^{28,29} RHL may be accompanied by a prodrome of paresthesia, burning, or itching at the site of reactivation.²⁹ The lesions appear as red macules, which become vesicles before breaking down into crusted pustular scabs or ulcers (Fig 7).²⁹ RHS has an entirely intraoral presentation, usually affecting the keratinized mucosa with a limited number of small ulcers,²⁹ in contrast to RAS, which often affects the nonkeratinized mucosa.²⁴ RHS often has a palatal distribution and may occur after local anesthetic injection; the ulcers usually heal within 10 days.²⁹ Generally, the clinical picture is sufficient to diagnose an HSV infection; however, a swab of the lesion for polymerase chain reaction analysis to identify viral DNA can be used when appropriate.²⁹ These conditions are usually self-limiting and are treated with supportive advice and antivirals such as acyclovir.^{29,30} There is limited evidence for the use of topical or systemic antivirals to prevent RHL.³¹ As there is a risk of transmission of HSV to the orthodontist while a lesion is active, it is essential to wear personal protective equipment. Moreover, there may be a benefit in delaying all but urgent treatment until the lesion has resolved to prevent infection risk.^{30,32}

Geographic tongue

Between 0.28% and 14.3% of children have geographic tongue.^{6,33,34} It is a benign condition of unknown etiology with a distinctive clinical appearance.^{33,34} As such, these lesions are infrequently biopsied. Rarely, this condition may affect sites in the



Fig 8. An example of geographic tongue. There are multiple well-defined but irregularly outlined areas of erythema of the dorsal and lateral tongue. This appearance is classical, and these lesions rarely require a biopsy for diagnosis.

oral cavity other than the tongue and is referred to as geographic stomatitis. Geographic tongue comprises varying-sized areas of depapillation and atrophy of the lateral, dorsal, and occasionally ventral tongue, giving an erythematous appearance (Fig 8).³³ There may be one or multiple lesions whose size and shape change over time.^{33,34} When biopsied, histologic examination will reveal a loss of filiform papillae in erythematous areas with variable hyperkeratosis, epithelial hyperplasia, and neutrophil micro-abscesses in the epithelium.^{33,34} As these lesions are usually asymptomatic, no management beyond the reassurance of the patient and his or her family is required.³⁴ However, the avoidance of spicy foods, topical corticosteroids, or analgesic mouthwashes (eg, benzydamine hydrochloride oral rinse) may be needed in a minority of patients.

Gingival overgrowth and gingival hyperplasia

Gingival overgrowth is a clinical term referring to an increase in the size of the attached gingiva, whereas gingival hyperplasia is a histologic diagnosis in which an increase in gingival cellularity causes overgrowth.⁷ Gingival overgrowth can be generalized or localized, and scoring systems have been developed to describe the extent of the overgrowth.^{7,35} The extent of overgrowth ranges from increased dental papillae to the coverage of the crowns of teeth by the gingivae.⁷ Gingival overgrowth may be in reaction to dental plaque; hormonal changes during pregnancy; medications



Fig 9. An example of gingival overgrowth (hyperplasia). This intraoral photograph shows gingival overgrowth caused by a patient receiving cyclosporin. In this example, the maxillary gingivae are greatly affected than the mandibular gingivae. The gingivae have maintained their normal stippled appearance despite the hyperplasia.

such as phenytoin, nifedipine, or cyclosporin (Fig 9); secondary to leukemia or lymphoma; or as part of a genetic disease such as hereditary gingival fibromatosis.⁷ A degree of gingival overgrowth is often seen in orthodontic patients in response to the brackets and gingival accumulation of dental plaque. If an obvious cause is absent, it would be important to investigate (with the help of the patient's general medical practitioner) the patient's drug history or to determine whether there is a need for further investigation such as biopsy with submission of representative lesional tissue for histopathologic examination to rule out other more serious causes of gingival overgrowth such as leukemia. Features that raise clinical concern for leukemia include swollen, bleeding gingivae without poor oral hygiene, skin bruises, or fatigue.

LESS COMMON ENTITIES

Although uncommon, the following diseases may present in children, young adults, and adults receiving orthodontic treatment. Candidiasis rarely occurs in healthy children or young adults but may be identified, especially when an orthodontic appliance is a contributing factor. Pleomorphic adenoma is the most common benign salivary gland tumor, and mucoepidermoid carcinoma, adenoid cystic carcinoma, and acinic cell carcinoma are the most common salivary gland malignancies in children and adults.^{5,8} Orofacial granulomatosis, although uncommon, is often the first manifestation of systemic disease and extremely important to recognize.^{36,37}

Candida infections

Candida species are commonly found in the oral cavity of children and adults.³⁸ They rarely cause infections in healthy subjects, although several factors predispose them to *Candida* spp infections. These include a dry mouth, drugs such as steroids (both topical and systemic) or antibiotics, dentures or orthodontic appliances, immunosuppression, and poor oral hygiene.³⁹⁻⁴¹ The most common causative species is *Candida albicans*.³⁹⁻⁴² Multiple clinical patterns of oral candidiasis³⁹⁻⁴² have been described; however, the most likely to be encountered by orthodontists include acute pseudomembranous, acute atrophic, and chronic atrophic candidiasis. Pseudomembranous candidiasis accounts for the third of *Candida* spp⁴⁰ and appears as white plaques in the oral cavity, which can be brushed off, revealing erythematous mucosa.^{40,42} It is more common in patients with systemic diseases that lead to reduced immune capacity.^{40,42} Acute atrophic candidiasis may be seen in patients receiving broad-spectrum antibiotics or steroids.⁴⁰ It appears as widespread erythematous patches in the mouth, and patients may complain of a burning sensation.^{40,42} Chronic atrophic candidiasis is usually caused by wearing a removable appliance such as a denture or orthodontic appliance^{40,42} and appears as an erythematous lesion often restricted to the footprint of the causative appliance (Fig 10).⁴⁰ Although a clinical history and examination are often sufficient for diagnosis, oral swabs or rinses followed by culture for *Candida* spp can help support the diagnosis.^{40,41} Management is done by correcting causative factors, if possible, and antifungal drugs such as miconazole or nystatin.^{39-41,43,44} For example, advice on appliance hygiene can help manage chronic atrophic candidiasis. When no obvious predisposing factors are identified, referral to the patient's general medical practitioner may be valuable for investigating potential systemic causes.

Pleomorphic adenoma

Although uncommon, pleomorphic adenoma remains the most frequently observed salivary gland tumor in children and adults.^{5,8} Most pleomorphic adenomas arise in the parotid gland, although they are the most common tumors of the intraoral minor glands.⁴⁵ They present as slow-growing, painless lumps within the major glands or the submucosal tissue of the oral cavity.⁴⁵ However, some lesions may expand more rapidly, and intraoral pleomorphic adenomas may present with ulceration.^{46,47} Histologic assessment reveals various architectural patterns composed of epithelial components forming ducts alongside myoepithelial

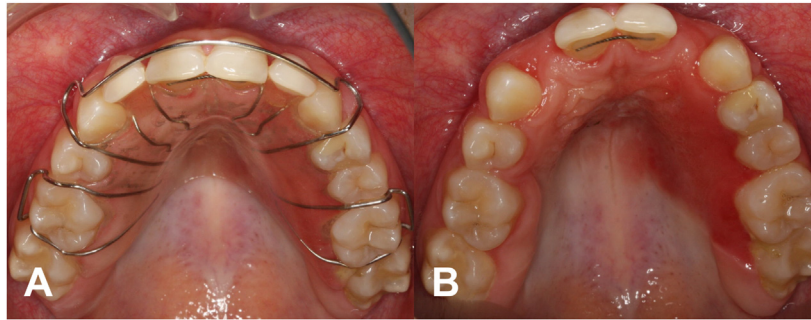


Fig 10. An example of chronic atrophic candidiasis: **A**, An intraoral photograph showing a removable orthodontic appliance in situ; **B**, An intraoral photograph showing an area of erythema corresponding to the appliance's outline.

components in a mucoid, myxoid, or chondroid matrix.⁴⁵ Treatment is through surgical excision,⁴⁸ with a recurrence rate of 1%-5%.⁴⁵

Malignant salivary gland tumors

Malignant salivary gland tumors are rare in children, accounting for <10% of pediatric head and neck cancer,⁴⁹ and are much more likely to occur in adults.⁵⁰ Despite this, a salivary gland tumor in a child is more likely to be malignant than in an adult patient.⁴⁹ The most common salivary gland malignancies in children are mucoepidermoid carcinoma, acinic cell carcinoma, and adenoid cystic carcinoma.⁴⁹ These tumors are most common in the parotid gland but may also present intraorally.^{49,50} The usual presentation is a palpable, painless, slow-growing mass in the salivary glands or submucosa.^{49,50} Moreover, patients may present with pain or evidence of cranial nerve deficits.^{49,50} Microscopic evaluation of mucoepidermoid carcinoma shows a combination of mucous, epidermoid, and intermediate cells in a variably solid or cystic pattern.⁴⁹ Acinic cell carcinoma has a variety of patterns, but all contain epithelial cells with a granular cytoplasm. The cytoplasmic granules are stained with a Periodic Acid-Schiff diastase stain.⁴⁹ Adenoid cystic carcinoma shows a combination of epithelial and myoepithelial cells in either tubular, cribriform, or solid patterns. Immunohistochemistry helps with detecting the 2 cell types.⁴⁹ Treatment for all these tumors is excision with a surgical margin of normal tissue, and in some patients, postoperative chemotherapy or radiotherapy may be used.^{49,50}

Orofacial granulomatosis and Crohn's disease

Orofacial granulomatosis is diagnosed when there is a swelling of the oral soft tissues in the presence of granulomatous inflammation on histopathologic



Fig 11. An example of orofacial granulomatosis. This intraoral photograph shows the gingival enlargement and cobblestone mucosa often observed in orofacial granulomatosis.

examination without a diagnosis of a systemic granulomatous inflammatory disorder such as Crohn's disease.⁵¹ Alternatively, granulomatous oral cavity inflammation may present as part of the spectrum of Crohn's disease in which other areas of the gastrointestinal tract are involved.⁵¹ The most common clinical feature is swelling of the lips, followed by intraoral manifestations, including ulceration, gingival swelling, and cobblestone mucosa (Fig 11).^{37,51} Biopsy of the affected areas will show noncaseating granulomatous inflammation.^{36,37,51} Other causes of granulomatous inflammation must be ruled out, including sarcoidosis and tuberculosis.^{37,51} Excluding the involvement of other sites is important, as up to 68% of children and 32% of adults will have Crohn's disease with oral manifestations rather than orofacial granulomatosis in isolation.^{36,37} Treatment usually includes exclusion diets, steroids, and steroid-sparing drugs,⁵¹ although no randomized controlled trials have been developed.⁵²

Lipoma

Other than hemangiomas, connective tissue pathology is the least common category of soft-tissue disease found in children.⁵ Most are tumors of endothelial origin; however, a small number of lipomas is observed in children.^{5,53,54} Intraoral tumors are most common in the buccal mucosa, followed by the lip and tongue; however, they may also be present in the parotid or submandibular glands.^{53,54} Most present as asymptomatic soft submucosal masses with an average diameter of just >2 cm.^{53,54} Although there are many histologic variants, most are classical lipoma, which comprises lobules of mature adipocytes with a fibrous capsule.^{53,54} Recurrence is rare after conservative excision.^{53,54}

RARE BUT IMPORTANT ENTITIES

Here, we discuss rare but important entities. These diseases are unlikely to be encountered, although they are still some of the more common malignant head and neck diseases in children other than those discussed.^{5,55,56} It is important that these life-threatening diseases are identified and managed early. In addition, squamous cell carcinoma is the most common malignant disease of the soft tissues in adults.⁸

Rhabdomyosarcoma

Rhabdomyosarcoma is the most common pediatric sarcoma of the head and neck⁵⁶ and accounts for approximately 60% of sarcomas in children.⁵⁵ Although rhabdomyosarcoma can present anywhere, the most frequent site is the head and neck.^{55,56} Intraoral presentations are uncommon; however, extraoral examination may identify lesions outside the oral cavity.^{55,56} The clinical presentation is usually a painless submucosal mass; some may present with symptoms such as nasal congestion and ear or facial pain.⁵⁶ Moreover, cranial nerve deficits are reported, including facial palsy.⁵⁶ The most common histologic types are embryonal rhabdomyosarcoma (ERMS), which is the most common in children,^{55,56} and alveolar rhabdomyosarcoma (ARMS), which is more common in older children and young adults.⁵⁶ ERMS comprises round to spindle cells with hyperchromatic nuclei and scant cytoplasm with occasional rhabdomyoblasts.⁵⁷ ARMS comprises clusters of small round cells with little cytoplasm surrounded by fibrous septa.⁵⁷ Immunohistochemistry is usually required to reach a diagnosis for both histologic types, and specific molecular markers can be used to distinguish the 2 entities.⁵⁷ Treatment comprises chemotherapy as well as surgery and radiotherapy for local control.⁵⁶ Patients with ERMS have a 5-year survival rate of 77%, whereas for those with ARMS, it is 54%.⁵⁶ Patients who survive their disease may subsequently have a host of



Fig 12. An example of an oral squamous cell carcinoma. This extraoral photograph demonstrates the clinical features of a squamous cell carcinoma. This lesion presented as an irregular, indurated ulcer on the lateral tongue with raised, rolled margins.

dentofacial developmental abnormalities that require further management.⁵⁸

Ewing sarcoma

Although commonly presenting as primary bone tumors, soft-tissue Ewing sarcomas are also seen in up to 30% of patients with head and neck Ewing sarcoma.⁵⁹⁻⁶¹ Most head and neck Ewing sarcomas occur in children and young adults.⁵⁹⁻⁶¹ They may present with pain, a mass, or weakness, or as incidental findings.⁶¹ The histology of these tumors is sheets and groups of small, round, uniform cells. A diagnosis of Ewing sarcoma is confirmed with molecular studies demonstrating an EWSR1 gene fusion.⁵⁷ Most are treated with a combination of resection and radiotherapy, with others with surgery or radiotherapy alone.⁵⁹⁻⁶¹ Five-year survival for multimodal treatment is up to 87%,^{59,60} and outcomes are better in head and neck disease when compared with in other sites.⁶⁰

Squamous cell carcinoma

Squamous cell carcinoma is the most common malignant tumor of the oral cavity in adults,⁸ and its incidence is rising.⁶² It usually presents as a firm lump, a nodular lesion, or an ulcer with raised margins (Fig 12), although it can also present as leukoplakia or erythroplakia.⁶² It may cause erosion of bone, trismus, mobility of the teeth, or pain depending on the site involved.⁵⁷ Moreover, the initial presentation may include a neck lump in which the tumor has metastasized to the neck lymph nodes. Early referral is essential to allow rapid

management and improve patient survival outcomes. Histologic examination reveals islands of pleomorphic squamous cells that invade the connective tissue, often with dysplasia on the surface epithelium.⁵⁷ Squamous cell carcinomas are treated by excision with a margin of normal tissue, often accompanied by a neck dissection and reconstruction of the deficit caused by tissue removal.⁶² Radiotherapy and chemotherapy may be used as adjuncts to surgical management.⁶²

CONCLUSIONS

Orthodontists are well placed to identify soft-tissue disease in their patients because of the frequency of examination. Although most soft-tissue diseases in children and young adults are reactive and require limited intervention, careful examination will allow the early detection of more aggressive entities. Finally, as the number of adults receiving orthodontic treatment increases, a greater number of malignant diagnoses can be expected.

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AUTHOR CREDIT STATEMENT

Paul Hankinson contributed to conceptualization, methodology, original draft preparation, and manuscript review and editing; Catherine Brierley contributed to conceptualization, methodology, original draft preparation, and manuscript review and editing; Daniel Brierley contributed to conceptualization, methodology, original draft preparation, and manuscript review and editing.

SUPPLEMENTARY DATA

Supplementary data associated with this article can be found, in the online version, at <https://doi.org/10.1016/j.ajodo.2023.09.012>.

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