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Hamartomas, choristomas and teratomas of the oral cavity: a contemporary narrative review

Vinesh Raj.S[1], Nur Syahirah Mohd Nazar[2]*

[1]*Centre of Oral & Maxillofacial Diagnostics & Medicine Studies, Faculty of Dentistry, Universiti Technology MARA, Sungai Buloh, Selangor, Malaysia.*
[2]*Department of Oral and Maxillofacial Surgery, Medicine and Pathology, Faculty of Dentistry, Universiti Sains Islam Malaysia, Kuala Lumpur, Malaysia.*

Abstract

Hamartomas, choristomas and teratomas represent an important group of developmental lesions that may occur within the oral and maxillofacial region. Although each entity is benign in nature, their clinical and histopathological resemblance to odontogenic tumours, soft-tissue neoplasms and other intraoral proliferations frequently results in diagnostic uncertainty. The overlap in clinical appearance among these developmental lesions demands a clear understanding of their biological basis and diagnostic criteria. This review provides an updated narrative synthesis of the current perspectives on hamartomas, choristomas and teratomas of the oral cavity, including their etiopathogenesis, clinical presentations, diagnostic considerations and management.

Keywords: *choristoma, hamartoma, oral cavity, review, teratoma*

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***Corresponding author**
Address:
Department of Oral and Maxillofacial Surgery, Medicine and Pathology, Faculty of Dentistry, Universiti Sains Islam Malaysia, Level 15, Tower B, Persiaran MPAJ, Jalan Pandan Utama, 55100 Kuala Lumpur, Malaysia

Telephone:
+60183949278

Email address:
syera_nazar@usim.edu.my

Introduction

Hamartomas, choristomas and teratomas are distinct developmental lesions that occasionally present within the oral and maxillofacial complex. Their unusual tissue composition and developmental origin, combined with their potential to mimic benign neoplasms, have contributed to long-standing debates regarding their classification (de Souto Medeiros *et al.*, 2024). The term "hamartoma" derived from the Greek *hamartía* meaning "error" was introduced by Albrecht in 1904 and refers to a mass composed of disorganised but mature tissues native to its anatomical location (de Souto Medeiros *et al.*, 2024; Patil *et al.*, 2015). In contrast, the term "choristoma", originating from the Greek *chōristos* meaning "separated", designates the presence of histologically normal tissue in an anatomically inappropriate site (de Souto Medeiros *et al.*, 2024). These definitions,

though established for more than a century, continue to evolve as new insights regarding the developmental and molecular basis of these entities emerge.

Teratomas represent another category of developmental lesion relevant to the oral cavity, although significantly rarer. These lesions are composed of tissues arising from two or more germ cell layers and may present within the head and neck region as congenital masses (de Souto Medeiros *et al.*, 2024). Oral teratomas, including epignathus, can be life-threatening at birth due to airway obstruction (Kolekar *et al.*, 2016; WHO, 2022). Their inclusion in discussions of developmental pathology is essential, as their biological origin and clinical implications differ from both hamartomas and choristomas (Kolekar *et al.*, 2016).

This narrative review synthesises the current understanding of hamartomas, choristomas and teratomas affecting the oral cavity, incorporating developmental mechanisms, clinicopathological characteristics, diagnostic criteria and contemporary considerations in classification. The review aims to clarify distinctions between these lesions and benign tumours, thereby supporting accurate diagnosis and management.

Definitions and biological distinctions

Hamartomas are benign malformations composed of an abnormal arrangement of mature tissue elements normally present in the region of occurrence (Neville *et al.*, 2023). Despite their tumour-like appearance, their growth parallels that of the surrounding tissues, and they lack the capacity for metastasis or invasive behaviour. Their architecture may be disordered or excessive, but the cellular components remain cytologically benign (Leiter Herrán *et al.*, 2017). They often present in childhood but may first be detected in adulthood, particularly when asymptomatic.

Choristomas exhibit similar clinical behaviour with hamartomas, but differ fundamentally in tissue origin (Neville *et al.*, 2023). They consist of normal, mature tissue located at an anatomically inappropriate site (Fu *et al.*, 2024). The tissue retains its expected morphology and function but is found in a location not typically associated with that tissue type, such as bone on the dorsal tongue or salivary gland tissue within the mandibular bone (Fu *et al.*, 2024).

Teratomas are true developmental growths composed of tissues from more than one germ layer, ectoderm, mesoderm and endoderm (Neville *et al.*, 2023). Although most commonly reported in the sacrococcygeal region, teratomas may occur within the head and neck, including the oral cavity (WHO, 2022). They may be classified as mature or immature based on the degree of cellular differentiation. Oral teratomas, particularly epignathus, may include complex tissues such as cartilage, hair follicles, respiratory epithelium or gastrointestinal structures (Huang & Pan, 2017). Although oral cavity teratomas are rare and generally benign in newborns, they carry a risk of malignant transformation, particularly when incompletely resected or diagnosed in older children (Ueno *et al.*, 2009).

Clear diagnostic differentiation is essential for clinical management. The narrative review proposes refined criteria for the classification of hamartomas and choristomas, stressing developmental origin, mass formation, benign growth patterns, and tissue nativity or ectopic distribution. These criteria help ensure appropriate use of terminology and avoid misinterpretation of developmental defects, reactive lesions or true neoplasms.

Etiopathogenesis

Hamartomas arise from aberrations in embryonic development involving disordered proliferation, differentiation or organisation of tissues that are native to the site. Genetic contributions have been well documented, particularly in syndromic presentations. Mutations involving PTEN, SMAD4, STK1 and BMPR1A contribute to

syndromes such as PTEN hamartoma tumour syndrome, including Cowden syndrome, which frequently presents with multiple mucocutaneous hamartomas and increased neoplastic risk (Pilarski, 2019; WHO, 2022). Gardner syndrome is an autosomal dominant condition resulting from APC gene mutations and is associated with multiple odontomas, osteomas, epidermoid cysts, and supernumerary teeth affecting the head and neck region (Black *et al.*, 2024).

Choristomas result from embryological misplacement of normal tissue into abnormal locations, leading to ectopic tissues such as thyroid, salivary, or respiratory epithelium within the oral region (Walling, 1987). Oral heterotopic gastrointestinal cysts represent a rare form of choristoma, most commonly affecting the tongue and floor of the mouth, and may remain asymptomatic or present with site-dependent clinical manifestations (Maldonado *et al.*, 2025). Their pathogenesis, while developmental, follows a distinct trajectory from hamartomas, as the tissues involved are normal but misplaced.

Teratomas originate from pluripotent germ cells capable of differentiating into multiple tissue types (McDonald *et al.*, 2020). When such cells become sequestered in the craniofacial region during embryogenesis, they may form complex masses containing tissues from multiple germ layers. The presence of tissues foreign to the oral cavity, such as skin appendages, neural tissue or gastrointestinal epithelium, is characteristic (McDonald *et al.*, 2020; WHO, 2022). Although rare, craniofacial teratomas demonstrate the potential for significant morbidity due to airway obstruction, feeding difficulties and craniofacial distortion.

Clinical presentations

Hamartomas of the oral cavity

Hamartomas in the oral cavity encompass a range of odontogenic and non-odontogenic developmental proliferations (Patil *et al.*, 2015; Woo, 2017). Among odontogenic lesions, odontomas remain the most widely recognised example. Although classified as benign mixed odontogenic tumours, these lesions are generally considered hamartomatous, as they consist of fully differentiated dental tissues arranged either in an organized pattern, as seen in compound odontomas, or as an amorphous mass in the complex variant (Neville *et al.*, 2023). Their limited growth potential, spontaneous maturation and virtual absence of recurrence after conservative removal support their interpretation as the terminal stage of odontogenic tissue development (Patil *et al.*, 2015). Several authors have proposed a developmental continuum in which ameloblastic fibroma progresses to ameloblastic fibro-odontoma and ultimately to odontoma. Age (13.5 years) and lesion size (2.1 cm) have been suggested as cut-off points to distinguish a developing odontoma from a true neoplasm; however, the universality of this maturation sequence remains debated (Soluk-Tekkesin & Vered, 2021).

In addition to odontoma, a number of odontogenic lesions exhibit biological behaviour more compatible with hamartomas than true neoplasms (Soluk-Tekkesin & Wright, 2022). Adenomatoid odontogenic tumour is officially classified by the World Health Organization (WHO) as a benign odontogenic tumour; however, its clinical and histopathological characteristics, including its slow growth, encapsulation, negligible recurrence rate and consistently low proliferative indices on immunohistochemistry, have led many investigators to propose that it is essentially hamartomatous in nature (Soluk-Tekkesin & Wright, 2022; WHO, 2022; Woo, 2017). Its behaviour sharply contrasts with that of invasive odontogenic neoplasms and aligns more closely with self-limiting developmental proliferations.

Peripheral odontogenic lesions further illustrate this hamartomatous spectrum. These extraosseous odontogenic proliferations arise from remnants of the dental lamina or from the basal layers of the oral epithelium, and include peripheral odontogenic fibroma, the soft-tissue variants

of calcifying epithelial odontogenic tumour, and the peripheral form of adenomatoid odontogenic tumour (Ide *et al.*, 2005). Although these lesions can clinically mimic neoplasms, they are typically indolent, grow slowly, and rarely recur after simple excision. Their origin from residual odontogenic epithelium and non-invasive behaviour have led many to consider peripheral odontogenic tumours as hamartomas rather than true neoplasms, though the classification of certain lesions remains controversial due to their biological behaviours and limited growth potential.

Non-odontogenic hamartomas, including lymphangiomas, vascular or neurovascular hamartoma, exostoses and leiomyomatous hamartoma are also encountered in the oral cavity (Neville *et al.*, 2023). Lymphangiomas are benign, hamartomatous tumor-like growths of lymphatic vessels. Like other vascular malformations, it is doubtful that they are true neoplasms; instead, they most likely represent developmental anomalies that arise from sequestrations of lymphatic tissue that do not communicate normally with the rest of the lymphatic system, with the dorsal surface of the tongue being the most commonly affected site (Neville *et al.*, 2023). A vascular or neurovascular hamartoma of the oral cavity is a rare, benign developmental lesion characterized by a disorganized proliferation of mature nerve bundles and blood vessels. Clinically, it typically presents as a painless, smooth, pinkish-red nodule and is most commonly located on the dorsal surface of the tongue (Gabriel Levi *et al.*, 2025).

Exostoses are common, self-limited bony outgrowths of the oral cavity with an unclear etiology, though their development may sometimes follow an autosomal dominant pattern (Neville *et al.*, 2023). They are more frequently observed in females between 30 and 50 years of age, possibly reflecting a combination of genetic factors and the slow, gradual growth of the lesion from puberty into adulthood (de Souto Medeiros *et al.*, 2024).

Leiomyomatous hamartoma is a rare, benign developmental lesion characterized by a disorganized overgrowth of mature smooth muscle bundles intermixed with other indigenous tissues (Neville *et al.*, 2023). It most commonly occurs on the midline dorsal tongue and presents as a slow-growing, painless, sessile or pedunculated mass, often detected at birth or during early childhood, although adult cases have also been reported. Histopathologically, it is composed of mature smooth muscle arranged in a haphazard pattern without cytologic atypia, supporting its hamartomatous rather than neoplastic nature (Nava-Villalba *et al.*, 2008).

Choristomas of the oral cavity

Choristomas of the oral cavity represent foci of histologically normal tissue occurring at sites where they are not ordinarily found. These lesions are developmental in origin and reflect heterotopic tissue deposition rather than neoplastic transformation (Neville *et al.*, 2023). Although uncommon, choristomas encompass a diverse range of tissues, illustrating the complexity of craniofacial embryogenesis.

Osseous choristomas are among the best-documented oral choristomas and most frequently arise on the posterior dorsum of the tongue and these lesions present as firm, sessile or pedunculated nodules and contain mature lamellar bone within the soft tissue (Goswamy *et al.*, 2012). Cartilaginous choristomas, although less frequent, similarly occur on the tongue or buccal mucosa and consist of normal hyaline cartilage situated in a non-cartilaginous site (Sakrikar *et al.*, 2022). Both lesions are thought to result from displaced embryonic rests or metaplastic transformation induced by local developmental influences (Gorini *et al.*, 2014).

Salivary gland choristomas constitute another important category and involve ectopic salivary tissue located outside the usual anatomical distribution of the major or minor salivary glands (Torres Medina *et al.*, 2021). The Stafne bone cavity is a developmental lingual concavity of the mandible, most commonly located in the posterior region below the mandibular

canal. It is typically asymptomatic and discovered incidentally on radiographs. Histologically, the cavity may contain normal salivary gland tissue, fat, or connective tissue, leading some authors to describe it as a salivary gland choristoma within bone. As a choristoma, it represents normal tissue in an ectopic location, rather than a true neoplasm. Its non-expansile, benign nature means that intervention is rarely required (Lee *et al.*, 2016).

A range of rarer choristomas has been reported. Lingual thyroid choristoma arises from aberrant thyroid tissue retained along the thyroglossal tract, most commonly at the foramen caecum (Constantin *et al.*, 2023). Sebaceous gland choristomas present as ectopic sebaceous lobules in sites devoid of Fordyce granules, such as the buccal mucosa (Marques *et al.*, 2022). Respiratory epithelial choristomas and gastric mucosal choristomas, although extremely rare, highlight the possibility of misplaced endodermal derivatives within the oral cavity (Ombres *et al.*, 2022). Glial choristomas, consisting of heterotopic neuroglial tissue, are distinctly uncommon but well documented and likely result from embryonic sequestration of neuroectoderm (Glavis-Bloom *et al.*, 2019).

Collectively, the diversity of oral choristomas underscores their developmental nature. Their composition reflects tissues that are entirely normal in structure but rendered abnormal by their ectopic location. Although the classification of oral choristomas as developmental heterotopias is generally well established, the etiopathogenesis of certain subtypes, particularly osseous choristomas, remains incompletely understood. Proposed mechanisms include embryonic rests, branchial arch remnants, metaplastic changes induced by chronic trauma or inflammation, and ossification of ectopic mesenchymal tissue; however, no single theory fully explains all reported cases (Gorini *et al.*, 2014). Recognition of these entities as heterotopias is essential, as their clinical presentation can mimic benign tumours, cysts, or reactive proliferations. Surgical excision is generally curative, and recurrence is exceedingly rare.

Teratomas of the oral cavity

Teratomas of the oral cavity and head and neck are rare germ cell tumours composed of mature or immature tissues derived from all three embryonic germ layers (Neville *et al.*, 2023). Their most dramatic intraoral manifestation is epignathus, a congenital teratoma arising from the oropharynx, palate or adjacent craniofacial structures. These lesions often contain diverse tissue elements such as bone, cartilage, glandular epithelium, skin adnexa and neural tissue. Newborns may present with respiratory obstruction, feeding difficulties and significant craniofacial distortion, making prompt multidisciplinary intervention essential (de Souto Medeiros *et al.*, 2024).

Head and neck teratomas exist along a spectrum ranging from mature to immature and malignant germ cell tumours. Mature teratomas are composed predominantly of well-differentiated tissues and generally behave in a benign manner, whereas immature teratomas contain embryonic neuroectodermal elements associated with more aggressive clinical behaviour (Chakravarti *et al.*, 2011). Malignant transformation, although uncommon, has been documented, and malignant germ cell tumours such as yolk sac tumour or embryonal carcinoma may rarely arise in this region (Zhang *et al.*, 2013).

Histopathologically, the diagnosis is established by identifying derivatives of the three germ layers, with immature neuroectoderm representing the hallmark of immature teratoma (WHO, 2022). Although teratomas of the oral cavity are primarily congenital, malignant germ cell tumours may also occur in older children and adults, albeit infrequently (Lohana *et al.*, 2020). Prognosis depends on tumour maturity, anatomical extent and the need for airway management at birth. Mature teratomas have an excellent prognosis following complete excision, whereas malignant germ cell tumours require multimodal oncological management and carry a significantly poorer outcome.

Figure 1 shows schematic classification of hamartomas, choristomas, and teratomas of the oral cavity with representative examples according to their biological origin and tissue composition.

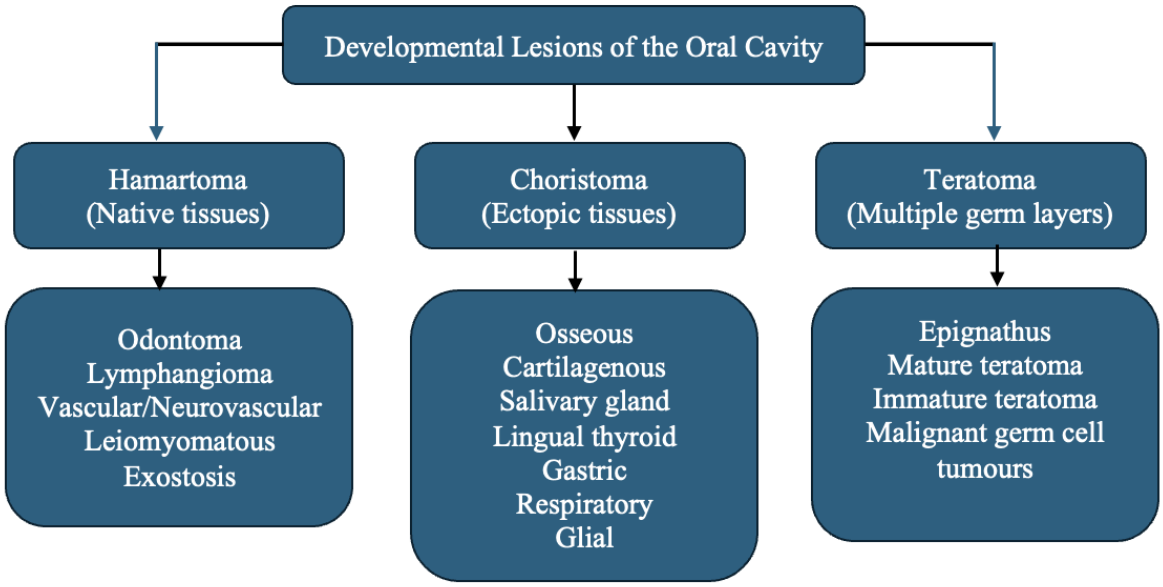

Figure 1. Schematic classification of hamartomas, choristomas, and teratomas of the oral cavity. Representative examples of each developmental lesion are shown according to their biological origin and tissue composition.

Diagnostic considerations and management

The diagnosis of hamartomas, choristomas and teratomas of the oral cavity requires careful integration of clinical, radiographic and histopathological findings, as these lesions may mimic a variety of benign neoplasms (de Souto Medeiros *et al.*, 2024). The anatomical location and tissue composition are the most reliable distinguishing features. Hamartomas are identified by the presence of mature tissues native to the site arranged in a disorganised or excessive fashion, whereas choristomas consist of normal tissue types occurring at anatomically inappropriate locations (de Jesus *et al.*, 2025). Teratomas differ from both by exhibiting elements derived from multiple germ layers and by displaying structural complexity not seen in other developmental lesions. Radiographic evaluation is particularly useful in characterising intraosseous hamartomas such as odontomas or osseous choristomas, which demonstrate well-circumscribed radiopaque components within the jawbones (de Jesus *et al.*, 2025). Teratomas, in contrast, frequently present as heterogeneous masses containing mixed cystic, solid and calcified elements, a pattern that may raise suspicion even before definitive histological assessment.

Management strategies reflect the biological behaviour of these developmental lesions. Hamartomas are generally treated with simple surgical excision or enucleation, and recurrence is exceedingly rare (de Souto Medeiros *et al.*, 2024). Choristomas also respond well to conservative surgical removal, and long-term outcomes are excellent (de Souto Medeiros *et al.*, 2024). Teratomas, however, may require more urgent and extensive intervention because of their size, anatomical relationships, and potential to compromise the airway, particularly in neonates with oropharyngeal involvement. Complete surgical excision is curative for mature teratomas, whereas immature or malignant variants necessitate

ongoing multidisciplinary follow-up, with additional oncological treatment determined by tumour type and behaviour (de Souto Medeiros *et al.*, 2024).. Prompt recognition and appropriate surgical planning remain essential to preventing morbidity and ensuring optimal outcomes.

Comparative summary of hamartomas, choristomas and teratomas of the oral cavity, highlighting their developmental origin, tissue composition, clinical behaviour, diagnostic hallmarks and management considerations to facilitate accurate differentiation from true neoplastic lesions is presented in Table 1.

Table 1. Comparative overview of hamartomas, choristomas and teratomas of the oral cavity.

Aspect	Hamartomas	Choristomas	Teratomas
Biological nature	Developmental malformations composed of disorganised but mature tissues indigenous to the site	Developmental heterotopias consisting of histologically normal tissue located in an anatomically aberrant site	Germ cell–derived developmental tumours containing tissues from two or more embryonic germ layers
Clinical behaviour	Non-invasive, self-limited growth paralleling surrounding tissues	Indolent, non-aggressive lesions with minimal growth potential	Variable behaviour ranging from benign (mature) to aggressive or malignant (immature or transformed lesions)
Age at presentation	Typically childhood or early adulthood; may be incidental findings	Broad age range, often detected incidentally	Predominantly congenital or neonatal; rare cases in older children and adults
Pathogenesis	Aberrant tissue organisation and proliferation during embryogenesis; may be sporadic or syndromic (e.g., PTEN-related disorders)	Embryological misplacement or sequestration of normal tissue during craniofacial development	Abnormal persistence and differentiation of pluripotent germ cells within the craniofacial region
Tissue composition	Mature tissues native to the affected oral site, arranged excessively or irregularly	Fully differentiated, normal tissues foreign to the anatomical location	Complex admixture of ectodermal, mesodermal and/or endodermal derivatives
Common oral examples	Odontoma, haemangioma, lymphangioma, exostoses, oral melanocytic nevi	Osseous and cartilaginous choristomas, salivary gland choristoma (e.g., Stafne bone cavity), lingual thyroid, gastric, glial and respiratory epithelial choristomas	Epignathus and other congenital oropharyngeal teratomas
Diagnostic hallmark	Histopathological identification of an unencapsulated proliferation of mature indigenous tissues with preserved cytological features and no evidence of cytological atypia	Histopathological confirmation of mature, histologically normal tissue in an ectopic anatomical location, supported by correlation with the clinical site	Histopathological demonstration of tissues derived from two or more embryonic germ layers, often corroborated by imaging showing a heterogeneous lesion with calcified, cystic, or solid components
Management and prognosis	Conservative surgical excision; recurrence is rare	Simple excision is curative; excellent prognosis	Complete surgical resection essential; prognosis depends on tumour maturity and extent

Conclusions

Hamartomas, choristomas and teratomas of the oral cavity represent biologically distinct developmental lesions that may closely mimic benign neoplasms in their clinical presentation. A thorough understanding of their embryological origins, tissue composition and characteristic growth patterns is essential to achieving accurate diagnosis and delivering appropriate management.

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