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ORAL HAMARTOMAS – A SHORT REVIEW

ABSTARCT

The majority of oral diseases constitute growth and masses of various cellular origin. Such masses may include hamartoma, choristoma, teratoma, benign or malignant neoplasms.

1. INTRODUCTION

The term hamartoma is derived from the Greek word “hamartia” referring to a defect or an error⁽¹⁾. It can be defined as a non-neoplastic, unifocal/multifocal, developmental malformation, comprising a mixture of cytologically normal mature cells and tissues which are indigenous to the anatomic location, showing disorganized architectural pattern with predominance of one of its components. In the oral cavity, indigenous tissues that might result in hamartomatous growths include odontogenic and non-odontogenic epithelial derivatives. The pathogenesis of hamartomas still remains speculative⁽¹⁾. They are derived from any one of the embryonic lineages, most commonly the mesoderm. This is almost never in the case of neoplasm, where the neoplastic cells are clonally derived⁽²⁾.

2. HAMARTOMATOUS GROWTH OF ODONTOGENIC APPARATUS

Dens invaginatus

Dens invaginatus is a malformation of teeth probably resulting from an infolding of the dental papilla during tooth development. The teeth most affected are maxillary lateral incisors and bilateral occurrence is not uncommon and occurs in 43% of all cases⁽³⁾. Various syndromes are associated which includes William’s syndrome, Nance-Huran syndrome, cranial suture syndromes.

Dens evaginatus

Dens evaginatus (DE) represents an accessory cusp and is predominantly seen in people of Asian descent. The incidence is 0.5–4.3%. Clinically, it may cause malocclusion resulting in abnormal wear or fracture and is treated accordingly⁽⁴⁾.

Enameloma

The enamel pearl is an ectopic globule of enamel that firmly adheres to the tooth’s root. It occurs more commonly on the roots of maxillary molars, especially the third molar, followed by the roots of mandibular molars⁽⁵⁾. It can be noticed rarely within the dentin then it is called as internal enamel pearl.

Odontoma

Odontomas are the most common developmental anomalies of the jaw which delay the eruption of the associated tooth. They are the most common benign odontogenic tumors of epithelial and mesenchymal origin. They are of two main types- compound and complex⁽⁶⁾. The incidence of compound odontome ranges between 9 and 37% and the complex odontome is between 5 and 30%⁽⁷⁾.

3. HAMARTOMATOUS GROWTH OF EPITHELIAL DERIVATIVES

Oral and labial melanotic macule

A labial melanotic macule is a well-defined, oval, brown to black, flat patch on the central third of the lower lip. A solitary labial melanotic macule is most commonly seen in adult women but it also occurs in males and in young people. The size normally ranges from 1-8mm. It is commonly associated with Peutz–Jeghers syndrome and Addison’s disease.

Oral melanocytic nevi

It represents collection of nevus cells which are derivatives of melanocytes or their precursor neural crest cells. Oral nevi are usually small and show regular symmetrical outline with no change in colour, shade or texture over time. It doesn’t require excision and may be followed up⁽⁸⁾.

4. HAMARTOMATOUS GROWTH OF MESENCHYMAL DERIVATIVES

Congenital and infantile hemangioma

Infantile hemangiomas (IHs) are vascular tumors, usually absent at birth or present as a premonitory mark with rapid post-natal growth followed by slow involution. IHs are the most common tumor of infancy, occurring in 4% of children⁽⁹⁾.

Vascular malformation

A vascular malformation is an abnormal development of blood vessels. They might be found in the large arteries and veins, arterioles, venules and in the lymphatic channels that carry lymphatic fluid and white blood cells outside of the arteries and veins. VM may occur as primary or in association with diffuse syndromes such as Sturge–Weber, proteus syndrome and Osler–Weber–Rendu syndrome⁽¹⁰⁾.

Glomuvenous malformation

Glomuvenous malformation is an unusual form of venous malformation, typically diagnosed at birth or later in infancy or childhood. It is also known as glomangioma or glomangiomas. The lesions tend to get more widespread, thicker, and deeper in colour with age. Clusters of alpha-actin-positive glomus cells (benign tumour cells in the soft tissue) are found lining the venous spaces.

Exostoses

Exostoses are benign protuberances of bone that arises on the cortical surface of the jaws. They usually appear as nodular masses on the buccal alveolar process. Surgical intervention is required only in case of tissue trauma, periodontal or prosthodontics implantation⁽¹¹⁾.

Other mesenchymal derived hamartomatous growth occurs rare in oral cavity and includes Rhabdomyomatous mesenchymal hamartoma, Leiomyomatous hamartoma, Neurofibroma, Fibrolipomatous hamartoma of nerve, oral neurovascular and neuromuscular hamartome

5. SYNDROMIC HAMARTOMAS

Tuberous sclerosis

Tuberous sclerosis (TS) is a rare syndrome characterized by the classic triad of seizures, mental deficiency and angiofibromas. It affects about 1 in 6000 people⁽¹²⁾. Tuberous sclerosis is often detected during infancy or childhood. Depending on the site and size of the benign tumors, they can cause severe or life-threatening complications in people with tuberous sclerosis.

Cowden syndrome/multiple hamartoma syndrome

Cowden syndrome represents the gene-related disorder which occurs 1 in 200,000 people⁽¹²⁾. Its origin is PTEN gene which results in the development of hamartomatous lesion. The oral manifestations include multiple papules involving the gingivae, buccal mucosa and dorsum of tongue.

Orofacial digital syndrome

Oral-facial-digital syndrome is actually a group of related conditions affecting the development of the oral cavity, facial features, and digits. The abnormalities oral-facial-digital syndrome include a split (cleft) in the tongue, a tongue with an unusual lobed shape, cleft lip, a wide nose with a broad, flat nasal bridge; and widely spaced eyes (hypertelorism).

6. CONCLUSION

The oral hamartoma are unique presentation of the head and neck region. Oral hamartomas should also be included in the differential diagnosis of the tumours of the oral cavity. Early detection of the tumour should be considered to avoid aggressive treatment and morbidity.

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