LEIOMYOMA OF ORAL CAVITY – A REVIEW

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Abstract

Leiomyoma, the commonest benign tumor of the uterus is a rare occurrence in the oral cavity and when present mimics various other soft tissue tumors of the oral cavity and lead to a diagnostic dilemma to the clinician. Leiomyoma is a muscle origin spindle cell tumour and its FNAC, histopathology and immune-histochemistry are required to reach at a definitive diagnosis. In the present review we will be discussing pathogenesis, clinical presentation, classification, FNAC, histopathology. Immunohistochemistry and differential diagnosis of leiomyoma of oral cavity.

Key words – FNAC, Spindle Cell Tumour, Leiomyoma.

Introduction

The leiomyoma is a benign tumor derived from smooth muscle and is found in a variety of anatomic sites, including the skin, subcutaneous tissues and the oral cavity. Most leiomyomas occur in the uterus (fibroids).¹

Blanc in 1884, reported the first case of a leiomyoma in the oral cavity in a 33 year-old man with a large tumor at the base of the tongue.^{2,3}

Leiomyomas of the head and neck region fall into two general types,

- Solid leiomyoma (sometimes referred to as the leiomyoma of deep soft tissue) and
- Vascular leiomyoma (angiomyoma, angioleiomyoma). The vascular leiomyoma accounts for almost three fourths of all oral cases, and rare examples of a third type,
- Epithelioid leiomyoma (leiomyoblastoma) have been reported.

Pathogenesis

The origin of leiomyomas of oral cavity is restricted to three areas with smooth muscle in their histological analysis: Tunica media of the blood vessels as suggested by scouts, ductus lingualis and circumvallate papilla as proposed by Glass. ²

Duhig and Ayer suggested that vascular leiomyoma represents only a stage within a continuous process of smooth muscle maturation. The maturation sequence would be as follows: hemangioma, angioma, vascular leiomyoma, leiomyoma and solid leiomyoma.^{3,4}

According to Damm and Neville, solid leiomyoma is histologically very different from angioleiomyoma, and the two entities therefore should be regarded as separate tumors.⁵

Demographics

Age: - Oral leiomyomas can appear at any age, but, majority of them occur in adults in the middle decades of life, over 65 per cent being found in patients older than 30 years of age. Peak age incidence is 40-49 years old.¹⁻⁵

Sex: - Regarding gender distribution, some authors consider both males and females to be affected in equal proportion, though considerable controversy exists on this point, since other investigators have reported a 2:1 female predilection, ^{2,5} while others consider males outnumber females (2:1). ^{3,4}

Site: - The common sites are posterior portion of the tongue, followed by palate, cheeks, gingiva, lips and salivary glands which together account for 80% of cases. Other less frequent locations are the floor of the mouth and the gingiva. 1-5

Clinical Presentation

Solid leiomyoma typically presents as a slowly enlarging, asymptomatic, firm submucosal mass or nodule. Colour of the lesion is similar to the adjacent mucosa or may show a greyish tone. Surface of the lesion is smooth and it rarely ulcerates.

Vascular leiomyomas often exhibit a blue or red discoloration. Usually, they are tender or painful, especially the vascular leiomyomas. On palpation, the tumors are generally well delimited, with free displacement within the tissues of the lip and oral mucosa. The size of the lesion ranges from a few millimetres to 3 cms. 5

Leiomyomas can be classified as

- Cutaneous Leiomyoma: Cutaneous leiomyomas commonly arise from the pillar erector muscle of hair follicles and are significantly painful. They are usually firm, subcutaneous nodules that are thin and expanded, but with an intact overlying skin.
- Vascular Leiomyoma: These lesions are almost identical to the cutaneous leiomyomas, except that their vessel walls are thick, whereas those of the cutaneous type are thin and indistinct. They have a predilection for women, except in the oral cavity, where they have a predilection for men. This type of leiomyoma is less common than the cutaneous type but tends to occur more frequently in the oral mucosa and head and neck area. These probably arise from smooth muscle around arteriovenous connections. They do not pose an enhanced bleeding potential. This type is also noted for its pain.
- Leiomyoma of Deep Soft Tissue: Leiomyoma of deep soft tissue is the rarest form of leiomyoma, the lesions probably arise from vascular smooth muscle and attain the largest size. They are not painful and are usually discovered only as an incidental finding or when their size produces secondary symptoms. Degenerative changes, including calcifications that may be identified radiographically, are found.

Fine Needle Aspiration Cytology

Cells are arranged in singles or in clusters, stripped nuclei are found among the single cells. Typical tumor cell nuclei are cigar shaped, sometimes truncated and sometimes contain vacuoles. Chromatin is finely granular and the nucleoli are inconspicuous. Many nuclei are fibroblast-like.⁷

Histopathological Features

Leiomyomas are located in the submucosa, separated from a typically intact mucosa. They are composed of spindle cells arranged in orderly fascicles, whorls and intersecting bundles. The cells have elongated, vesicular to stippled nuclei with blunt ends (cigar shaped), surrounded by fibrillar eosinophilic cytoplasm. They are highly differentiated, with little or no atypia, although rare cells may exhibit nuclear pleomorphism. Necrosis and invasion are absent, and mitotic activity is scarce. Mucinous degeneration, hyalinization or fibrosis, and adipocytes can be seen, but these features are usually focal and more likely seen in larger lesions.

The World Health Organization (WHO) classifies leiomyomas into three histological types: solid leiomyoma (25 %), vascular leiomyoma or angiomyoma (74 %) and leioblastoma or ephitheloid leiomyoma (1 %).³

Solid leiomyoma is a well delimited tumor not associated with the vascular smooth muscle. It is composed of interlacing bundles of smooth muscle fibers interspersed by varying amounts of fibrous connective tissue. The bundles of fibers appear to form whorls because of their fascicular arrangement in varying planes. The nuclei are typically spindle-shaped with blunt ends (cigar shaped) and quite vesicular. Mitotic figures are uncommon. Intra-cytoplasmic myofibrils are present and can be demonstrated by phosphotungstic acid- hematoxylin special stain.⁵

Angiomyomas are well circumscribed lesions that demonstrate multiple torturous blood vessels with thickened walls caused by hyperplasia of their smooth muscle coat. Vascular leiomyoma (angiomyoma) contains capillary, cavernous or venous vascular spaces. Intertwining bundles of smooth muscle may be found between the vessels, sometimes intermixed with adipose tissue.⁶

Epitheloid leiomyoma or leiomyoblastoma is composed of round or polygonal cells with clear areas surrounding the nucleus, and an acidophilic cytoplasm. Smooth muscle fibers are rarely found. ^{1,2}

Ultrastructure

Electron microscopically, the tumor cells contain elongated nuclei with rounded ends. Characteristic filaments for smooth muscle cells, marginal densities, well-developed mitochondria, and rough endoplasmic reticulum are seen in the cytoplasm of the cells. Dense patches and basal laminae are also observed along the plasma membrane.⁴

Immunohistochemistry

Desmin, actin, H-caldesmon and vimentin are diffusely and strongly positive in the neoplastic cells. Most tumor cells are also positive for smooth muscle actin and Myoglobin. The Ki-67 index is usually <5%.

Differential diagnosis

Myofibroma: - Myofibroma can be easily distinguished by the characteristic zonal phenomenon.

Hemangiopericytoma: - Hemangiopericytoma does not present with lesional cells arranged in long bundles or fascicles and is negative for smooth muscle-specific antibodies.

Schwannoma: - Neural tumors usually positively stain for S-100 protein and neuron-specific enlace but not for desmin and smooth muscle actin.

Leiomyosaroma: - The presence of mitoses is a hallmark of malignancy (at least 1 mitotic figure per 10 high power fields), lesions with 5 or more mitotic figures per 10 high-power fields should definitely be considered malignant.⁸

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