

Neural Tumors of Oral and Maxillofacial Region

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ABSTRACT

Neural tumors are rare in the oral and maxillofacial region which occurs as both benign and malignant. They are neuroectodermal in origin. Most of the neural tumors arises from schwann cells and peripheral nerve sheaths. This paper reviews about the histological difference of the neural tumors of the oral region.

Keywords: Neural Tumor, Oral region, Nerve sheaths.

INTRODUCTION

Nerve cells and supporting cells are the 2 main cells in the nervous system. Neuron or nerve cell is the main structural and functional unit of the system. Supporting cells of the CNS are neuroglia cells, peripheral nervous system are schwann cells and satellite cells. Neural tumors are of neuroectodermal in origin. It appears as painless swelling which is of smooth surface in the soft tissues of the mouth. The most common site is being the tongue. In the head and neck region the most of the them are benign tumors

Classification of neural tumors: (WHO 1992)

Benign Tumours:

1. Traumatic Neuroma
2. Perineural Fibrosis
3. Neuromuscular Hamartoma, Fibroblastoma, Neurinoma, Lemmoma
4. Nerve Sheath Ganglion
5. Schwannoma - plexiform, cellular, ancient or degenerated
6. Neurofibroma and Neurofibromatosis or

Vonrecklingausen' Disease Of Skin

7. Diffuse Neurofibroma
8. Pacinian Neurofibroma
9. Epitheloid Neurofibroma
10. Granular Cell Tumor
11. Melanocytic Schwannoma: Pigmented Schwannoma
12. Nerve Sheath Myxoma
13. Ectopic Meningioma.
14. Ganglioneuroma
15. Pigmented Neuroectodermal Tumor of Infancy

Malignant Tumours

1. Malignant peripheral nerve sheath tumors (MPNST).
2. Malignant Schwannoma
3. Neurofibrosarcoma
4. Malignant peripheral nerve sheath tumor with Rhabdomyosarcoma, glandular differentiation and Epitheloid
5. Malignant granular cell tumor
6. cellular cell sarcomas: Malignant melanoma of soft parts
7. Malignant melanocytic Schwannoma.

8. Neuroblastoma
9. Ganglioneuroblastoma
10. Neuropithelioma

Traumatic neuroma

It occurs due to the trauma during extraction of tooth, any physical injuries or at the site of previous injuries¹. Clinically it appears as a swelling or small nodule near the mental foramen, lips or tongue. Traumatic neuroma of the mental foramen was first reported by Cahn². Bone associated with nerve trunk may also occur. It is slow growing and becomes larger in size.

Histopathological appearance of traumatic neuroma of irregular mass with interlacing neurofibrils and Schwann cells. Connective tissue is scanty and it is derived from perineurium. Proliferating nerve fibers may occur diffusely throughout the lesion or small discrete islands.³

Special stains used to identify collagen are the trichrome methods (Masson, Mallory)⁴. In IHC staining methods S100 and EMA appears to be positive for perineural components

Palisaded encapsulated neuroma

It is a benign true neoplasm. It causes primary hyperplasia of nerve fibers, axons and their sheath cells.⁵ Clinically appears as a solitary, cutaneous tumor occurring mainly at mucocutaneous junctions on the face. Oral cavity is the rarest site of origin. Histopathological appearance of neuroma shows an encapsulated mass which may be complete or incomplete. Tumor consists of interlacing fascicles of spindle cells with moderately cellular and some have lobulated appearance. Special stains like Schofield method reveals the presence of numerous axons in the tumor. Differential diagnosis of the lesion is neurofibroma and neurilemoma^{5,6}.

Schwannoma (neurilemoma, perineural fibroblastoma)

It is a benign neural neoplasm of nerve sheath origin. Schwann cells form the inner layer of the peripheral nerve sheath. Head and neck region are most commonly affected. Bilateral or multiple lesions is a feature of neurofibromatosis which is a hereditary condition⁵. Clinically it appears

as solitary, slow growing and pain may be associated in some lesions. In the head and neck region tumors occur on both oral and paraoral region, in which tongue being the most commonly affected site^{3,5}. Intraosseous or central lesion affects both the maxilla and mandible. Pain, bone destruction and expansion of cortical plates may be seen in some of the lesions. The tumor is composed of 2 types of tissues Antoni A and Antoni B. Histopathological appearance of Antoni A shows spindle shaped cells with palisading arrangement and fibers are arranged in whorls or swirls. Verocay bodies are a striking feature of the lesion. Antoni B does not exhibit a palisading arrangement and spindle cells are disorderly arranged. Microcyst formation is seen⁷. One of the histological variants of schwannoma is the ancient schwannoma (hemosiderin and hemorrhagic areas can be seen)⁸. Schwannoma cells are positive for S100 protein and not for EMA

Neurofibroma

It arises from both the Schwann cells and perineural fibroblasts. Two forms of neurofibroma are solitary neurofibroma and neurofibromatosis

Solitary neurofibroma – It is a non-encapsulated tumor of slow growing consisting of Schwann cells and perineural cells. Neurofibroma is a well-demarcated lesion with proliferation of interlacing bundles of spindle cells, wavy nuclei and sometimes melanocytes can be seen. Mast cells are the characteristic feature of the lesion which helps in diagnosis³. Solitary plexiform neurofibroma is a variant of neurofibroma which may be associated with neurofibromatosis and has a risk of malignant transformation³.

Neurofibromatosis (Von Recklinghausen's Disease of Skin) – It is an autosomal dominant condition with multiple neurofibromas, skin pigmentation and bony abnormalities which may transform into neurofibrosarcoma⁵. Two types of neurofibromatosis are defined: NF type 1 – mutations on gene neurofibromin and NF type 2 – mutations on gene schwannomin. Clinical appearance shows café au lait macules, axillary freckling (Crowe sign), iris freckling (Lisch spots) and bony changes are also seen⁹.

Malignant peripheral nerve sheath tumor (neurogenic sarcoma , neurofibrosarcoma)

It arises from peripheral nerve schwann cells and its rare in the oral cavity. Sarcomas are of 2 types 1. Malignant transformation of neurofibromatosis to sarcoma¹⁰ 2. Malignancy in the absence of neurofibromatosis ¹¹. Malignant schwannoma commonly seen in lips, buccal mucosa and palate where as in central tumors mandible is more common. Histopathological appearance shows spindle shaped cells with comma shaped nuclei and cellular , nuclear

pleomorphism , mitotic figures are very high. Schwannoma appears in 3 forms of pattern 1. Epitheloid 2. Rhabdomyoblastic differentiation 3. Glandular or mesenchymal ³.

CONCLUSION

There is a chance of malignant transformation in benign tumors is seen so it is important to distinguish between the benign neoplasms and peripheral nerve neoplasms

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