International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Review Article

Meningiomas of Oral Cavity - A Review

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Received: 10/06/2016 *Revised: 22/06/2016* *Accepted: 24/06/2016*

ABSTRACT

Meningiomas are very common tumours of the central nervous system with multiple histological presentations. Meningiomas arise from meningothelial cells. Meningothelial cells are thought to be of dual origin; both from neuroectoderm and mesoderm. The histologic diversity and the biologic spectrum of meningiomas are due to this dual origin. Extra cranial meningiomas are very rare. This paper reviews the biological behaviour of meningiomas with a focus on primary extra cranial meningiomas of oral cavity published so far.

Keywords: oral meningioma, extra cranial, epithelial membrane antigen, meningioma review.

INTRODUCTION

Meningiomas are defined by World Health Organization (WHO) as meningothelial (arachnoid) cell neoplasms, typically attached to the inner surface of the dura mater. [1] It is a very common tumor of the central nervous system with multiple histological presentations. Meningothelial cells are thought to be of dual origin; both from neuroectoderm and mesoderm. [2] These cells are seen in the arachnoid membrane and cap the arachnoidal villi associated with intra dural venous sinuses and their tributaries. The histologic diversity and the biologic spectrum of meningiomas are attributed to this dual origin. Extra cranial meningiomas are very rare (less than 2% of all meningiomas) [3] Common extracranial sites reported being the orbit, the outer table of the skull and scalp, the paranasal sinuses, the nasal cavity, the parotid and para pharyngeal region. [4]

Origin

Many hypotheses have been proposed for the occurrence of meningiomas outside the cranium. Earlier it was believed to be a direct extension from intracranial mass where the connecting link would have got destroyed or eroded in due course of time. [5] The reason for presence of arachnoid cells at odd positions has been debated over for many years. The arachnoid cells can be seen in nerve sheath or even in blood vessel sheath where they emerge through the skull foramina. Arachnoid granulations may get detached or entrapped at extracranial location during embryologic development. A traumatic event like tooth extraction or cerebral hypertension that displaces arachnoid islets. An origin from undifferentiated or multipotential mesenchymal cells, such as fibroblasts, Schwann cells, or a combination of these, perhaps explaining the diverse pathologic spectrum found in meningiomas. [3] Distant metastasis from intracranial meningiomas can also be a possibility. This could be true in case of an aggressive meningioma.

Clinical Characteristics

Meningiomas are slow growing tumors and are usually asymptomatic unless, large in size. Meningiomas are broadly categorised into extra cranial and intra cranial. [6] Intra cranial meningiomas are classic and originate from the arachnoid cells innate to the region. Extra cranial meningiomas can be primary or secondary. Primary extra cranial meningiomas are ectopic meningiomas arising in uncommon sites. Secondary extracranial meningiomas are direct extension of the intracranial meningiomas to any possible site in the tumour vicinity. [6] Primary extracranial meningiomas are extremely rare due to the uncommon presence of ectopic arachnoid cells. Extra cranial meningiomas often present as painless swellings and in the head and neck region, may present as swellings causing facial asymmetry. Extra cranially, meningiomas have been reported in many areas including head and neck soft tissue, [7] lungs, [8] mediastinum, [9] mandible, [6, 10-13] peripheral nerve plexus and ganglion [14] and others.

Till date, fourteen cases of extra cranial meningiomas have been reported in the oral cavity, all of which were benign in nature (Table 1). [5-6,10-13,15-21] Among the cases reported in oral cavity, most cases were reported in mandible.

Table1: Primary extracranial meningiomas reported in oral cavity till 2015

Sr. No.	Site of Primary Meningioma	Age	Sex	Author: Year
1	Maxillary Gingiva	**Details not available**		Brown et al:1976
2	Left Maxilla	63	F	Simpson and Sneddon: 1987
3	Left Maxilla	43	F	Landini and Kitano
4	Anterior Maxilla	26	F	Reddi et al: 1999
5	Right Mandible	41	F	Jones and Freedman: 2001
6	Right Mandible	74	F	Jones and Freedman: 2001
7	Left mandible	10	M	Kubotaa et al: 2005
8	Left mandible	62	M	Mussak et al :2007
9	Right mandible	40	F	Lell et al: 2007
10	Right mandible	53	F	Mosqueda-Taylor et al: 2009
11	Mandible	**Details not available**		Rushing et al: 2009
12	Maxilla	59	F	Pinting et al: 2013
13	Right Buccal Mucosa	66	F	Maeng et al; 2013
14	Right Buccal Mucosa	60	F	Nair et al; 2015

Exact aetiology of meningioma is unclear. Sporadic cases have been reported in the past years. Meningiomas are seen to be more common in females with a reported ratio of 2:1. [22] In cases of extra cranial meningiomas of oral cavity, all but 2 cases were reported in females. Hormonal aetiology has also been suggested [23] and also incidence of meningioma is noted to be more in females with history of breast carcinoma. [22]

Pathology: Meningiomas present in diverse histological patterns though many of them does not have any prognostic implications. The various histologic types are enumerated in Table 2.

Table 2: Histologic variants of Meningioma [1]

World Health Organization (WHO) classification of meningiomas
Meningiomas with low risk of recurrence or aggressive growth
Meningothelial Fibrous (fibroblastic) Transitional (mixed) Psammomatous Angiomatous Microcystic Secretory Lymphoplasmacyte-rich Metaplastic
Meningiomas with greater likelihood of recurrence and/or aggressive behaviour
AtypicalI Clear cell (intracranial) Chordoid
Meningiomas of any subtype or grade with high proliferative index and/or brain invasion
Rhabdoid Papillary Anaplastic (malignant)

WHO has graded meningioma into three categories (Table 3). [24] 80% of all meningiomas fall into Grade I and are usually benign in nature. Grade II, atypical meningiomas are more aggressive and shows a higher recurrence rate. Histological features like increased mitosis, brain invasion and spontaneous necrosis can be noticed. 15-20% of all the meningiomas belong to grade II with a 5 year mortality rate of 20%. Anaplastic meningioma (Grade III) is the rarest of all with the highest chances of recurrence (80%). Aggressiveness reflects in the histologic features as well.

Table 3: WHO Grading of meningiomas [1]

WHO Grade I- Benign Meningioma	Histologic variant other than clear cell, chordoid, papillary and rhabdoid Lacks criteria of atypical and anaplastic meningioma
WHO Grade II	Any of the following 3 criteria: Mitotic index >= 4/10 HPF* Loss of whirling/fascicles Small cell formation Macronucleoli Hypercellularity Spontaneous necrosis Brain invasion
WHO Grade III	Either of the following 2 criteria: Mitotic index>=20/HPF Frank anaplasia

*High Power Field - 40X magnification of 0.452 mm in field diameter

Though meningiomas have many histological patterns, the diagnoses of intracranial meningioma are often straightforward. However, the diagnosis can be a dilemma if the lesion is extra cranial or the biopsy sample is too small or the site is not representative.

Fine Needle Aspiration Cytology is a proven useful technique for diagnosis of extracranial meningiomas. [25] But, in oral meningiomas, FNAC has yet been performed, so definitive diagnosis has been achieved by histopathology and immunohistochemistry.

Immunohistochemistry and electron microscopic studies have been employed for distinguishing meningiomas from other lesions. Immunohistochemical analysis of extra cranial meningiomas show cytoplasmic positivity for epithelial membrane antigen (EMA) and vimentin, like their intracranial counterparts. Ki 67 has been implicated as a prognostic marker. S-100 protein reactivity is in a diffuse form and sometimes negative. [24]

All the reported cases of extra cranial meningiomas have been treated by surgical excision. No recurrences of meningioma associated with oral cavity have been reported, though in one case, the patient was later diagnosed with basal cell nevus syndrome with multiple odontogenic keratocysts. [13] Mutations of the patched (PTCH) gene were reported to be associated with odontogenic keratocysts and meningiomas in the syndrome. [26] Mutations in NF2 gene are also shown to have an association in development of meningiomas. [27] Though most cases of meningiomas behave in a non aggressive manner, a long term evaluation and follow-up is mandatory.

CONCLUSION

Since pathology is a science of exception an acquaintance with lesions uncommon in oral cavity is unforeseeable to any oral pathologist. A typical histological pattern, a clinical sign or an immunohistochemical reaction can be a clue to the final diagnosis. Thus knowledge on lesions with otherwise typical extra oral manifestations becomes sine qua non to an oral pathologist.

ACKNOWLEDGEMENTS

Dr Binisree S.S., Junior Resident, Dept of Oral Pathology and Microbiology, Govt. Dental College, Trivandrum and Dr Kartik Kapoor, Senior Resident, Dept of Prosthodontics, Govt. Dental College, Trivandrum.

REFERENCES

1. Perry A, Louis DN, Scheithauer BW, Budka H, von Deiming A. Louis DN, Ohgaki Hiroko, Wiestler OD, and Cavenee WK. *Meningiomas in WHO Classification of Tumours of the Central Nervous System*. Lyon, France: International Agency for Research on Cancer; 2007:164-172.
2. Perry A. McLendon RE, Rosenblum MK, Bigner DD. *Meningiomas in Russell and Rubinstein's Pathology of Tumors of the*

Nervous System. 7th Edition. Hodder Arnold; 2006:427-474.
3. Rushing EJ, Bouffard JP, McCall S, Olsen C, Mena H, Sandberg GD, Thompson LD. Primary extracranial meningiomas: an analysis of 146 cases. Head Neck Pathol. 2009; 3(2):116-30.
4. Iaconetta G, Santella A, Friscia M, Abbate V, Califano L. Extracranial primary and secondary meningiomas. Int. J. Oral Maxillofac. Surg. 2012; 41: 211-217.
5. Geoffray A, Lee YY, Jing BS, Wallace S. Extracranial meningiomas of the head and neck. Am J Neuroradiol. 1984; 5(5):599-604.
6. Mosqueda-Taylor A, Domínguez-Malagon H, Cano-Valdez A, Montiel-Hernandez A. Primary extracranial meningioma of the mandible. Med Oral Patol Oral Cir Bucal. 2009 1; 14 (4):E167-70.
7. Milker-Zabel S, Huber P, Schlegel W, Debus J, Zabel-du Bois A. Fractionated stereotactic radiation therapy in the management of primary optic nerve sheath meningiomas. *J Neurooncol*. 2009; 94(3):419-24
8. Haddad GF, Al-Mefty O. The road less traveled: transtemporal access to the CPA. *Clin Neurosurg*. 1994; 41:150-167.
9. Jaffrain-Rea ML, Minniti G, Santoro A, et al. Visual improvement during octreotide therapy in a case of episellar meningioma. *Clin Neurol Neurosurg*. 1998; 100(1):40-3.
10. Landini G, Kitano M. Meningioma of the mandible. Cancer. 1992; 69:2917-20.
11. Jones AC, Freedman PD. Primary extracranial meningioma of the mandible: A report of 2 cases and a review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 2001; 91:338-41.
12. Mussak EN, Holodny AI, Karimi SK, Carlson DL. Meningioma of the mandible: imaging with CT. AJNR Am J Neuroradiol. 2007; 28:1157-9.
13. Kubota Y, Yamashiro T, Kobayashi I, Kawazu T, Shirasuna K. Primary meningioma of the mandible. Oral Oncol Extra. 200; 41(2):18-21.
14. Kleihues P, Cavanee W. *World Health Organization Classification of Tumours: Pathology and Genetics: Tumours of the Nervous System*. Lyon, France: IARC; 2000.
15. Brown AM, Fordham KC, Lally ET. Meningioma presenting as an intraoral mass. Oral Surg Oral Med Oral Pathol.1976; 41(6):771-6.
16. Simpson MT, Sneddon KJ. Extracranial meningioma of the oral cavity. Br J Oral Maxillofac Surg. 1987; 25:520-5.
17. Reddi SP, Strauss SI, Strauss JE, Blanchaert RH Jr. Anterior maxillary lesion. J Oral Maxillofac Surg. 1999; 57:1234-8.
18. Lell M, Tudor C, Aigner T, Kessler P. Primary intraosseous meningioma of the mandible: CT and MR imaging features. Am J Neuroradiol. 2007; 28(1):129-31.
19. Maeng JW, Kim YH, Seo J, Kim SW. Primary Extracranial Meningioma Presenting as a Cheek Mass. Cli Exp Otorhinolaryngol. 2013; 6(4):266-268
20. Pinting L, Xiaofeng H, Zhiyong W, Wei H. Extracranial meningioma in the maxillary region. J Craniofac Surg. 2013; 24(2):e142-4.
21. Nair KSR, Suresh Babu P, Manoj S, Moinuddin HK. The meningioma that got cheeky. J Maxillofac Oral Surg. 2015:1-3.
22. Longstreth WT, Jr., Dennis LK, McGuire VM, Drangsholt MT, Koepsell TD. Epidemiology of intracranial meningioma. Cancer 1993, 72: 639-648.
23. Saitoh Y, Oku Y, Izumoto S, Go J. Rapid growth of a meningioma during pregnancy: relationship with estrogen and progesterone receptors--case report. *Neurol Med Chir (Tokyo)*. 1989;29(5):440-3
24. Rosenblum MK, Bilbao JM, Cee-Ang L. Neuromuscular system: Central Nervous System Primary Tumours: Rosai J. Rosai and Ackerman's Surgical Pathology 9th ed. China: Mosby 2004. 2564-2572.
25. Tan LH. Meningioma presenting as a parapharyngeal tumor: report of a case with fine needle aspiration cytology. Acta Cytol.2001; 45:1053-9.
26. Tate G, Li M, Suzuki T, Mitsuya T. A new germline mutation of the PTCH gene in a Japanese patient with nevoid basal cell carcinoma syndrome associated with meningioma. Jap J Clin Oncol 2003; 33:47-50.
27. Louis DN, Ramesh V, Gusella JF. Neuropathology and molecular genetics of neurofibromatosis 2 and related tumors. *Brain Pathol*. 1995; 5(2):163-72.

How to cite this article: Aloka D, Padmakumar SK, Sivakumar R et al. Meningiomas of oral cavity - a review. Int J Health Sci Res. 2016; 6(7):337-340.

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