

Extracranial Meningioma Presenting as an External Auditory Canal Polyp**Dr. Sunita Singh^{1,2}, Dr. Yashika Bhatia^{2,3}, Dr. Shivani Dua^{2,4}, Dr. Jyoti Sharma^{2,5}, Dr. Rajeev Sen^{2,6}**¹Senior Professor, ²Department of pathology, PT BDS PGIMS, Rohtak, Haryana, India³Resident, ⁴Assistant Professor, ⁵Senior Resident, ⁶Senior professor and head of department, Department of pathology, PT BDS PGIMS, Rohtak, Haryana, India***Corresponding author**

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Abstract: Meningioma is a well-recognized tumor of the central nervous system with various histomorphological forms that typically arises in proximity to the meninges. Extracranial meningioma of the head and neck region are rare neoplasm making upto 1% of all meningiomas. We present a case of meningioma presenting as an external auditory canal polyp. Prognosis of this tumour is generally excellent. Surgical excision is the treatment of choice, with no need for further treatment.**Keywords:** meningioma, extracranial, external auditory canal.

INTRODUCTION

Meningiomas are usually benign tumors arising from meningotheial cells. They have been found to be more common in the sixth and seventh decade of life with female predominance[1]. These tumors account for 24-30% of all primary intracranial tumors with an incidence of 1.4-4.5/100,000 per year[2-3]. Extra-cranial or ectopic primary meningioma is a rare entity. They most often involve the head and neck region specifically the ear and temporal bone, nose, paranasal sinuses and floor of the mouth. Other rare sites include mediastinum, lung, skin and retroperitoneum where they arise from the arachnoid cells along the peripheral nerves. Ectopic meningioma involving the external auditory canal and the middle ear can be primary or secondary due to direct extension of intracranial tumor[4].

CASE REPORT

A 52-year-old female presented with a 3-month history of discharge and pain in the right ear

along with hearing loss. Examination of the right ear revealed a polypoidal growth filling the external auditory canal whereas the other ear was normal. Tuning fork tests and audiometry showed conductive hearing loss in the affected ear. CT head and neck revealed opacified right mastoid air cells and middle ear cavity with erosion of the middle ear ossicles, bony wall of facial nerve canal and extension of the lesion in the right external ear without any evidence of intracranial origin. The patient underwent right-sided modified radical mastoidectomy. The tissue excised from the middle ear and EAC was sent for histopathological examination. Microscopic examination showed nests and whorls of uniform small cells with moderate cytoplasm, fine nuclear chromatin and inconspicuous nucleoli along with psammoma bodies suggestive of meningotheial meningioma (Fig. 1). On Immunohistochemistry PR was positive in tumor cells whereas CK was negative (Fig. 2 & 3).

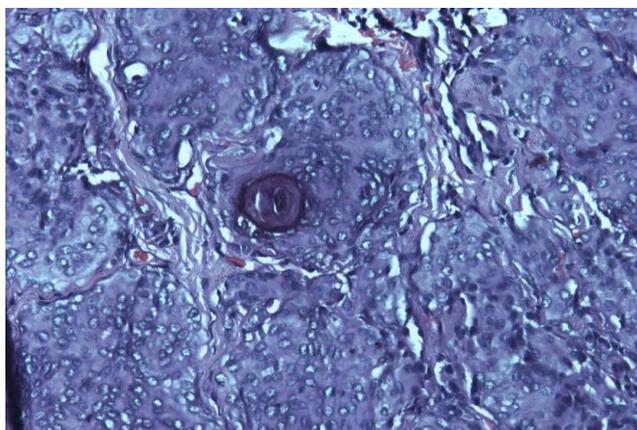


Fig-1: Meningotheial meningioma with whorled syncytial architecture showing cells having round nucleoli and indistinct cytoplasmic borders. Psammoma body is also included in the section. (H & E, 200X)

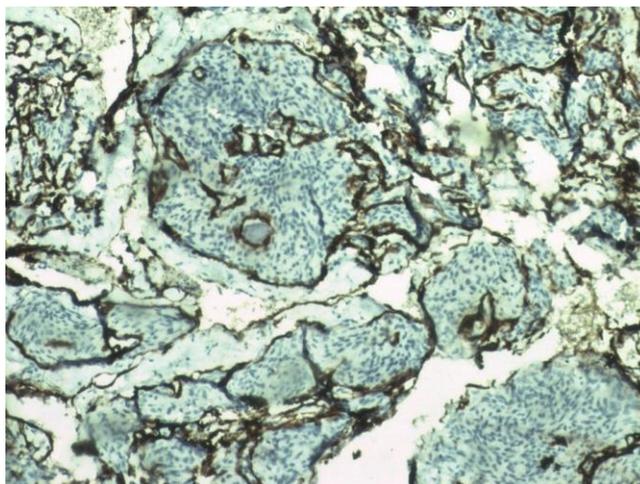


Fig-2: Tumor cells negative for CK immunostaining. (IHC, 100X)

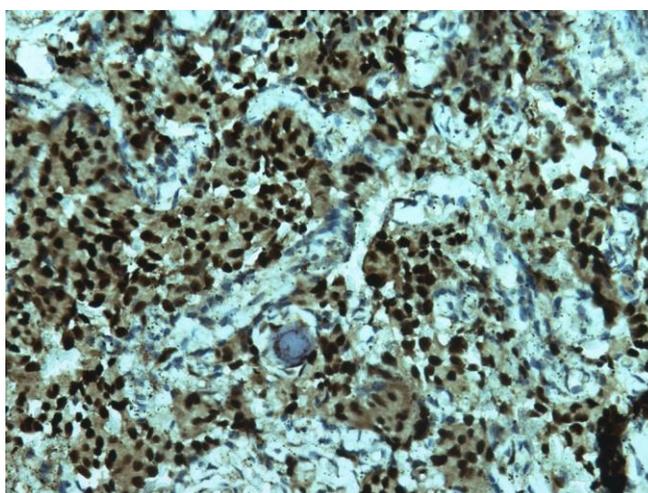


Fig-3: Meningothelial cells showing positivity for PR immunostain. (IHC, 200X)

DISCUSSION

Meningioma is a common tumor of the Central Nervous System that originates from the arachnoid cells. These cells line the inner aspect of the arachnoid membrane and fill the cores of the arachnoid villi that project into the lumens of dural veins and venous sinuses. Extra-cranial and extra-spinal meningiomas are rarely reported which comprise 0.9-2.0% of all meningiomas[5]. There are four different mechanism suggesting occurrence of extracranial meningioma: direct extension of an intracranial tumor, distant metastasis from an intracranial lesion, origination from arachnoid cells within the cranial nerves sheaths and origination from extracranial embryonic nests of arachnoid cells[6]. Upto 20% of intracranial meningiomas show extracranial extension and usually extends into the temporal area and ear by the path of least resistance through the tegmen tympani, the internal auditory canal, the sulci of the greater and lesser superficial petrosal nerves, the jugular foramen or the posterior fossa plate[7]. Primary meningioma of the ear and temporal bone without any evidence of associated intracranial tumor exhibit similar histological features as their intracranial counterparts. Various

histomorphological forms are known namely meningothelial, fibromatous, angiomatous, psammomatous and clear cell meningioma. Meningothelial meningioma is the most common variant characterised by cells having delicate round nucleus, nuclear clearing, indistinct cytoplasmic borders, cellular whorls and psammoma bodies. The differential diagnosis includes a variety of benign and malignant conditions like paraganglioma, schwannoma, squamous cell carcinomas, adnexal tumors, middle ear adenomas, ceruminous gland tumors, primary and metastatic carcinomas, and malignant melanomas. Histopathology and immunohistochemistry can generally differentiate these tumors from meningioma[3]. Clinically patient usually presents with hearing loss. Other symptoms include headache, discharge, vertigo, tinnitus and otalgia. Facial or other cranial nerves may be involved in late cases[8]. Surgery is the treatment of choice but there are number of challenges due to complex anatomy of the ear and invasiveness of tumour. Meticulous surgical extirpation is important to minimize the recurrence rate, without the necessity of adjuvant therapy. Recurrence rate for

meningiomas vary widely (7–84%) but prognosis is good[9].

CONCLUSION

Primary meningioma presenting as a mass in the external auditory canal is rare but needs to be considered by the otolaryngologist in the differential diagnosis of ear pathology. Presenting symptoms are non-specific which may lead to incorrect diagnosis and delayed treatment. Histopathologically extracranial meningiomas are similar to their intracranial counterpart and diagnosis is usually straightforward but a high degree of suspicion is needed when they present in unexpected location. Diagnosis and differentiation from other benign and malignant conditions is very important as surgical excision is curative.

REFERENCES

1. Saat R, Lempinen LJ, Laulajainen-Hongisto A, Markkola A, Jero J. An En Plaque Meningioma of the Temporal Bone, Complicating with a Cholesteatoma, Chronic Otitis Media and an Intracranial Abscess. *Ann OtolaryngolRhinol.* 2015;2:1050.
2. Marosi C, Hassler M, Roessler K, Reni M, Sant M, Mazza E, et al. Meningioma. *Crit Rev Oncol Hematol.* 2008;67:153-71.
3. Possanzini P, Pipolo C, Romangnoli S, Falleni M, Moneghini L, Braidotti P, et al. Primary extracranial meningioma of head and neck: clinical, histopathological and immunohistochemical study of three cases. *Acta Otorhinolaryngol Italica.* 2012;32:336-8.
4. Tomaru U, Hasegawa T, Hasegawa F, Kito M, Hirose T, Shimoda T. Primary extracranial meningioma of the foot: a case report. *Jpn J Clin Oncol.* 2000;30:313-7.
5. Thompson LD, Bouffard JP, Sandberg GD, Mena H. Primary ear and temporal bone meningiomas: a clinicopathologic study of 36 cases with a review of the literature. *Mod Pathol.* 2003;16:236-45.
6. Chang CY, Cheung SW, Jackler RK. Meningiomas presenting in the temporal bone: the pathways of spread from an intracranial site of origin. *Otolaryngol Head Neck Surg.* 1998;119:658–64.
7. Rushing EJ, Bouffard JP, McCall S, Olsen C, Mena H, Sandberg GD, et al. Primary ExtracranialMeningiomas: An Analysis of 146 Cases. *Head and Neck Pathol.* 2009;3:116–30.
8. Hemanth V, Sreenivas V, Chaturvedi J. Meningioma Presenting As A Mass In The External Auditory Canal. *The Internet Journal of Otorhinolaryngology.* 2010;13:1-4.
9. Manjaly JG, Watson GMT, Jones M. Primary meningioma of the middle ear. *J R Soc Med Sh Rep.* 2011;2:92.