MIDDLE EAR ADENOMA PRESENTING AS A PETROUS MASS LESION

Authors

Pearly PK (1), Amit Keshri (2) Ravi Shankar (3), Raj Kumar (4), Rajat Jain (5)
Senior Resident (1), Associate Professor (2), Assistant Professor (3), Professor & Head (4), Senior resident (5),
Neuro otology unit, Department of Neurosurgery, SGPGI, Rai Bareilly Road, Lucknow, UP 226014

Abstract:

Introduction: Inspite of modern radiological work up, surgeons can still be surprised by intraoperative findings or by pathologists report.

Materials and methods: We present the case history of a 51 year old male who came to our institute with complaints of unilateral hearing loss, involuntary facial movements and retroauricular pain for a period of 1 year. He underwent Fisch approach with short segment mobilization of facial nerve.

Results: Though pre-operative CT and MRI findings were suggestive of a petroclival mass he had middle ear adenoma.

Key Words: Middle ear, Middle Ear Adenoma, Middle Ear benign tumor, Adenoma

Introduction:

Pathologies of middle ear can be broadly classified as that of being congenital, infective and inflammatory and neoplastic. Though infective and inflammatory pathologies are more common, tumors can also arise in middle ear. Middle ear primary tumors are rare conditions that can produce a diagnostic challenge. According to literature glomus tympanicum is the most frequent of these followed by middle ear adenoma.^{1,2}

Case Report:

We present the case history of a 51 year old male with complaints of hearing loss, involuntary facial movements and ear ache on right side. Otoscopy revealed normal tympanic membrane. Audiometry showed right CHL and left SNHL. No other neurological deficit. MRI head showed an enhancing lesion in right petrous apex extending into ipsilateral cerebello pontine angle and mastoid (Figure 1). He underwent right trans-mastoid pre- sigmoid approach with tumor removal by Fisch approach. Tumor found involving presigmoid dura and infiltrating sigmoid sinus, jugular bulb and posterior semicircular canal. Short segment mobilization of facial nerve was done. Tumor was reaching hypotympanum medial to facial nerve and extending towards petrous apex with encasement of ICA in region of foramen lacerum. Subtotal tumor removal was achieved. Tumor was firm, pinkish-grey, and adherent to facial nerve sheath, infiltrating mastoid air cells and labyrinthine bone. Intra operative diagnosis of malignant petroclival mass was made. Frozen section findings were consistent with final HPE which revealed fibrous connective tissue and osteoid infiltrated by tumor arranged in trabeculae of polygonal cells displaying round isomorphic nuclei and moderate amount of cytoplasm, with no mitosis or necrosis. Immunohistochemistry showed positivity for creatine kinase, synaptophysin, chromogranin and EMA. It was

negative for vimentin, CD 10 and CK 7. Thus the diagnosis of middle ear adenoma (MEA) was made. Patient had grade IV facial palsy for which tarsorrhaphy was done. He was symptom free for one and half years.

He got re-admitted with complaints of involuntary movement of tongue. MRI showed evidence of an enhancing lesion in right petrous temporal bone, right half of clivus and occipital condyle encasing right ICA with involvement of internal acoustic meatus (IAM) with involvement of intracanalicular 7-8 cranial nerve complex extending into ipsilateral cerebello pontine angle suggestive of recurrence (Figure 2). He underwent re-exploration with right fronto-temporal craniotomy with zygotomy and near total tumor excision via extradural subtemporal approach. The roof of petrous was eroded by the tumor (Figure 3). Anteriorly the tumor was reaching up to the posterior cavernous sinus with petrous ICA being pushed anteriorly and medially. Part of tumor anteriorly encasing the ICA was left behind. (Figure 4) shows the tumor cavity. Histopathology revealed the same findings which were suggestive of MEA recurrence. Since the patient in our study had recurrence within 2 years of follow up, he was planned for adjuvant radiotherapy.

Discussion:

MEA, a benign epithelial tumor was first described by Hyams and Michaels¹ in 1976 and later that year by Derlacki and Barney³. It is a rare disease occurring in < 2% of all ear tumors which is thought to originate in middle ear mucosa. There are two types of secretory cells in middle ear mucosa - goblet cells and intermediate cells. MEAs arise from neuroendocrine cells which in turn are derived from pluripotent endodermal stem cells.

It occurs over a wide age range (mean age 45 years) and has no gender predilection². The clinical, otoscopic and imaging findings are nonspecific^{1,2}. Most common complaints are

conductive hearing loss, ear fullness, tinnitus and dizziness unilaterally. Majority of middle ear adenomas do not invade or erode the temporal bone or infiltrate the facial nerve. But in our case the patient experienced facial palsy and ataxia.

Temporal bone imaging will show a localized tumor in the middle ear without ossicular destruction.⁷ Differential diagnosis of MEA includes carcinoid, schwannoma, teratoma, meningioma, paraganglioma, ceruminous gland adenoma, endolymphatic sac tumor and lipoma. However congenital cholesteatoma, meningioma which extends into middle ear, acquired cholesteatoma and epidermoid should be kept in mind.^{5,6}

MEA can have both neuroendocrine and epithelial differentiation. This led to the misbelief that neuroendocrine (carcinoid) tumors and MEAs were separate tumors, but recent studies have demonstrated that these terms describe the same tumor which can have mixed patterns of differentiation. The classification for NAME proposed by Saliba and Evrard is based on immunohistochemistry findings and metastasis. Currently this is the most used classification of middle ear glandular neoplasms (Table 1)

Type	Immune-histochemistry	Metastasis	Percentage (%)
NEAME	+	-	76
MEA	+	-	20
CTME	+	-	4

Abbreviations: NEAME, neuroendocrine adenoma of middle ear; MEA, middle ear adenoma; CTME, carcinoid tumor of middle ear. Note: Adapted from Saliba and Evrard, 2009.

Complete surgical excision with or without the ossicles is the treatment for MEAs. Local recurrence rates as high as 12.7% are reported.⁴

Conclusion:

MEAs are rare neoplasms and have no specific symptoms or signs. Total exploration and removal

is required for treatment. Microscopic and immune histo-chemical examinations are required for a definitive diagnosis. Adjuvant radiotherapy or chemotherapy as used for pulmonary or gastrointestinal carcinoid is not generally recommended except in aggressive variety.

Figure 1

Figure 2

Figure 3

Figure 4

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