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Primary mucoepidermoid carcinoma of middle ear: A rare tumour

[1] Dr. Avni Gupta, [2] Dr. Hoogar MB, [3] Dr. Reeta Dhar, [4] Dr. Atul Jain, [5] Dr. Vaidehee Naik

[1] Senior post-graduate student, Department of Pathology, M.G.M's Medical College, Kamothe, Navi Mumbai, Maharashtra, India
[2] Associate Professor, Department of Pathology, M.G.M's Medical College, Kamothe, Navi Mumbai, Maharashtra, India
[3] Professor, Department of Pathology, M.G.M's Medical College, Kamothe, Navi Mumbai, Maharashtra, India
[4] Assistant Professor, Department of Pathology, M.G.M's Medical College, Kamothe, Navi Mumbai, Maharashtra, India
[5] Junior post-graduate student, Department of Pathology, M.G.M's Medical College, Kamothe, Navi Mumbai, Maharashtra, India

Abstract
Malignant tumours of middle ear are highly uncommon, though squamous cell carcinoma is the most common among malignant tumours. Mucoepidermoid carcinoma is a malignant tumour of varying indolent to aggressive biological behaviour, which is histologically composed of an admixture of mucous secreting cells, epidermoid cells and intermediate cells, which is commonly found in salivary glands, accessory salivary glands of lips, tongue and other sites such as larynx and pharynx. Its occurrence in middle ear is very rare and as such its origin in the middle ear which does not show the histologic presence of any glands or mucous cells, makes its histogenesis a bewildering enigma. The case being presented here is one such rare case of a mucoepidermoid carcinoma of the middle ear in a middle-aged woman which disguisedly manifested as chronic suppurative inflammation of left ear.

Keywords: middle ear, low grade, mucoepidermoid carcinoma

Introduction
Malignant tumours of middle ear are highly uncommon and some of the rarely occurring tumours in the ear are squamous cell carcinoma, basal carcinoma, adenocarcinoma, acinic cell carcinoma, adenoid cystic carcinoma, melanoma, osteosarcoma, chondrosarcoma, rhabdomyosarcoma, paragangliomas and lymphoma [2]. Mucoepidermoid carcinoma, described first in 1945 by Stuart *et al*, is a very rarely occurring tumour in middle ear, which is characterized by the presence of squamous, mucus-producing cells and cells of intermediate type along with clear cells, Mucoepidermoid carcinoma is commonly described in salivary glands, lips, tongue, palate, larynx and pharynx [3]. It is unusual to find mucoepidermoid carcinoma in middle ear in as much as the histological conformation of middle ear renders ambiguity to its histogenesis, and this possibly could explain rarity of its occurrence in that location that only four published cases of primary mucoepidermoid carcinoma of middle ear are found in English literature. Being presented here is one of the rare cases of epidermoid carcinoma of middle ear in a middle-aged woman which masqueraded as chronic supporative inflammation in left ear.

Case Report
A 44-year-old female presented to a tertiary care centre with persistent left sided otalgia of six months duration, associated with a mucopurulent otorrhoea and mild hearing loss. Patient had history of cortical mastoidectomy two years back. She had no history of vertigo, headache, fever, skin rashes or head trauma or other neurological disease. She had no other debilitating systemic illnesses such as diabetes mellitus or tuberculosis or hypertension. On examination there was a fleshy, proliferative soft tissue mass occupying the left ear canal. HRCT showed a destructive lesion involving the middle ear cavity, mastoid and external auditory canal causing destruction of wall of middle ear, scutum, external canal and middle ear oscicles. A clinical curettage was done from the left ear canal and the tissue specimen was sent to the histopathology department of the central laboratory.

Histopathology
Histopathological examination revealed multiple fragments of squamous epithelium with occasional sheets of round to polygonal cells with mild to moderate nuclear pleomorphism and abundant pale eosinophilic cytoplasm with focal areas showing poorly formed keratin pearls (Figure 1 & 2). Also noted were multiple cords and nests of round to cuboidal cells forming glandular structures at places. At places, sheets of round cells with bland and mildly enlarged nuclei displaying inconspicuous nucleolus were seen forming cribriform pattern with cribrifom or microcystic areas containing pools of mucin (Figure 3 & 4).
Sections stained with special stains for mucin such as Periodic Acid Schiff (PAS) and Alcian Blue (Figure 5 & 6) showed glandular structures and cribriform areas displaying abundant mucinous material with overall histomorphological features being suggestive of Low Grade Mucoepidermoid Carcinoma.

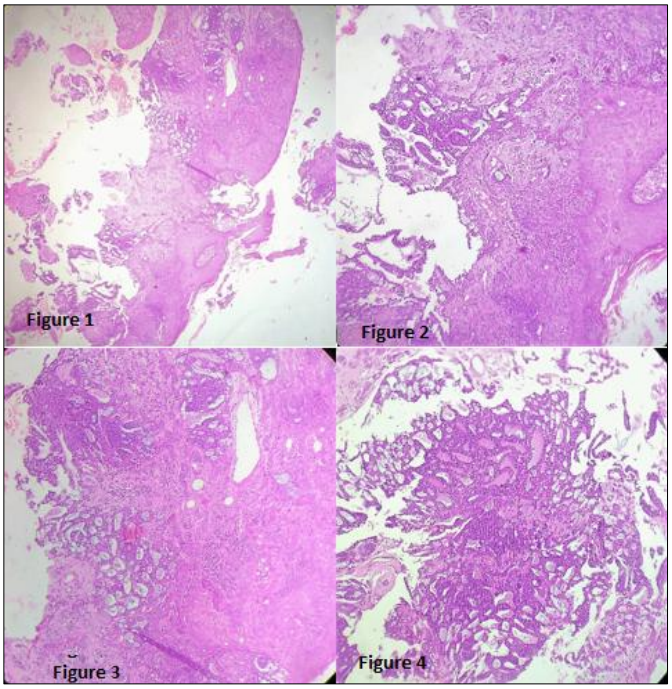

Fig 1: (H & E Stain ,40x) ; Figure 2 ,3& 4 (H & E Stain ,100x) : show fragments of squamous epithelium and occasional sheets of round to polygonal cells displaying mild to moderate nuclear pleomorphism, along with few glandular elements, cribiriform architecture and mucinous areas at places.

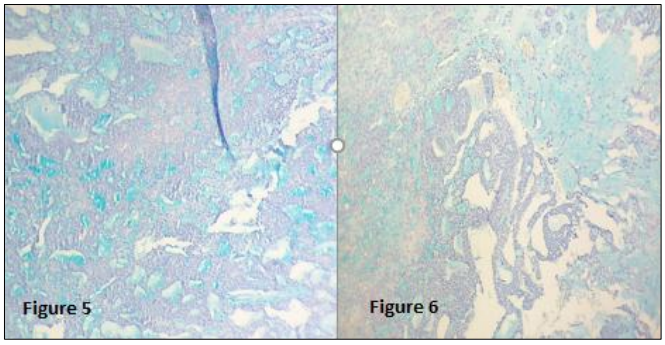

Fig 5-6: (Alcian Blue 100x) show positive staining in mucinous areas.

Discussion

Mucoepidermoid carcinoma is a malignant tumour which is often found to be arising in Salivary glands, lips, buccal mucosa, tongue, larynx, pharyx and also relatively uncommonly seen in other parts of aero-digestive tract with 70 percent of the tumours occurring in major or minor salivary glands [1]. Mucoepidermoid carcinoma is a tumour of generally indolent biological behaviour which rarely may transform into high-grade tumour with marked aggressiveness. Mucoepidermoid carcinoma is a unique tumour in that it is composed of an admixture of different type of cells such as epidermoid, mucous, intermediate and clear cells. Ultrastructural studies indicate that both squamous cells and mucous cells may differentiate from intermediate cells and that these cells are also a feature of these tumors, and myoepithelial cells are not found in mucoepidermoid carcinomas [4]. Histologically, mucoepidermoid carcinomas are commonly divided into three grades depending upon the predominance of one type or other of the two dominant cells: mucous cells and epidermoid cells. The presence of cystic component of less than 20 percent, neural invasion, necrosis mitoses (more than 4 per 10 high power fields) and anaplasia. Auclair *et al* proposed a grading system in which each of these parameters were given a point value of 2 each for cystic component and neural invasion, 3 for necrosis and mitoses and a point value of 4 for the presence of anaplasia. A total point score of all the existing parameters of 0-4 in a tumour was considered low grade, while point score of 7 and above was considered as High-grade. A score between 5 and 6 was assigned to intermediate grade tumours [5]. Most low-grade mucoepidermoid carcinomas consist of predominance of mucous cells and multiple well-developed cystic or microcystic structures lined by mucus producing, intermediate, or epidermoid cells. As mucoepidermoid carcinoma grows less differentiated become higher grade, the nests of tumor cells become larger, more irregular, and more solid with fewer cystic spaces containing mucous secretion. The high-grade carcinomas characteristically form solid sheets or nests or cords composed of intermediate and epidermoid cells, with paucity of mucin-producing cells. Perineural invasion and lymph node metastases are frequently associated with high-grade mucoepidermoid carcinomas.

The common malignant tumours arising in the external and middle and temporal bone are the squamous cell carcinoma, basal cell carcinoma, adenocarcinoma, acinic cell carcinoma, adenoid cystic carcinoma, melanoma, osteosarcoma, chondrosarcoma, rhabdomyosarcoma, lymphoma, malignant neuroma, malignant paraganglioma [2]. Among all the malignant tumours arising in the ear canal squamous cell carcinoma is most common accounting for approximately 90 percent of all cases [4]. Mucoepidermoid carcinomas involving the middle ear are rarely reported as such since most of these possibly get misdiagnosed as Squamous cell carcinomas. Young Ho Kim *et al* in 2003 reported an incidental finding of a mucoepidermoid carcinoma involving the middle ear in a patient undergoing surgery for chronic otitis media whom was treated with combined radiotherapy and chemotherapy [6]. In the same year, 2003, Rancic *et al* reported a case of high grade mucoepidermoid tumour of the middle ear that was treated surgically [7]. The overall outlook for the patients and the prognosis of mucoepidermoid carcinoma depends on tumour grade, clinical stage, location of the tumour and the clinical efficacy of adequate surgical and other modes and efficacy clinical mangagement. One of the significant parameter of prognosis is grading system proposed by Auclair *et al* mentioned above the mortality rate for low grade was merely 3.3 percent while the mortality rate for high-grade mucoepidermoid carcinoma being 46.3 percent and mortality rate for intermediate grade tumour estimated to be at 9.7 percent. Early diagnosis and efficient clinical management including complete surgical excision with normal tissue margins is considered ideal way of treating mucoepidermoid carcinoma of middle ear. Radiotherapy is indicated as adjuvant method of treating tumours showing predilection for perineural or angiovascular invasion which are not amenable for complete excision [4].

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