Epithelial-Myoepithelial Carcinoma of Parotid - A Rare Case Report

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Article ID: WMC002566
Article Type: Case Report
Article URL: http://www.webmedcentral.com/article_view/2566
Subject Categories: PATHOLOGY
Keywords: Epithelial-Myoepithelial; Parotid

How to cite the article: Mahapatra S. Epithelial-Myoepithelial Carcinoma of Parotid - A Rare Case Report. WebmedCentral PATHOLOGY 2011;2(12):WMC002566

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Source(s) of Funding:
None

Competing Interests:
Nil

Additional Files:
manuscript
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Abstract

Epithelial-Myoepithelial Carcinoma (EMC) is a rare tumor of salivary gland which presented in a 65 year old female as a swelling behind the right ear. Fine needle aspiration cytology (FNAC) of the swelling was suggestive of pleomorphic adenoma. Grossly, the tumor was well circumscribed. Histologically, the tumor was composed of ducts with double cell lining surrounded by a basement membrane in a sclerotic stroma. The inner cells were epithelial and the outer cells were myoepithelial, with clear cytoplasm. The immunohistochemistry was done to highlight the biphasic nature of the tumor whose cytoplasm was positive for actin. The patient diagnosed as Epithelial-Myoepithelial Carcinoma was treated with wide surgical excision. Reports of EMC are still relatively few with cytology and histology correlation. But, it is recommended that this histologically distinct neoplasm deserves wider recognition.

Introduction

Epithelial-Myoepithelial Carcinoma (EMC) is a rare type of malignant tumor of salivary gland accounting for less than 1% of all salivary neoplasms. It was initially described as a glycogen-rich or clear cell adenoma because of the clear cell component. Donath et al in 1972 introduced the term epithelial-myoepithelial carcinoma by noting the myoepithelial component as an integral part of the tumor. In 1991, the WHO recognized EMC as a distinct entity and subtype of salivary gland adenocarcinoma which became a part of the new classification system. The most common site is the parotid gland, but it has been also described in submandibular gland, in the minor salivary glands and extra oral areas. It is more commonly seen in females, with a peak incidence in seventh decade. We report this unusual entity in a 65 year old female who presented as a swelling of right parotid for duration of 6 months with cytological and histological correlation.

Case Report

A 65 year old female presented with a swelling behind right ear. The swelling rapidly increased in size in duration of 6 months. There was no lymphadenopathy, no history of fever. All other general examinations were within normal limit. It was clinically diagnosed as pleomorphic adenoma. Ultrasonography (USG) Neck with high resolution probe revealed irregular mass echogenicity (solid) mass behind right angle of mandible. The impression given was a space occupying lesion (SOL) of right parotid. Computed tomographic (CT) scan revealed well defined non homogenously enhancing mass. A magnetic resonance (MRI) image showed T1 isodense and T2 nonhomogenously hyperintense mass. The tumor’s interface with the surrounding parotid tissues was well defined. But, there were no well defined fat planes. There was no abnormal surrounding lymphadenopathy. All hematological parameters were within normal limits. Fine needle aspiration (FNA) was done from the swelling with 23G needle. Cytology revealed epithelial cells in clusters, singles, few spindloid cells in a scanty myxoid stromal background (Fig 1). From, the cytoscans the diagnosis given was suggestive of Pleomorphic adenoma of right parotid. En mass removal of the parotid mass was done by giving an S shaped incision in front of the ear below the ear lobule behind the parotid to the front again. By raising the skin and subcutaneous tissue flap parotid fascia was identified and removed. Facial nerve was identified followed by superficial parotidectomy. The mass was 4cms in size with slight adhesion to the surrounding tissues. No lymph node structure was identified. Deep parotidectomy was done without removal of the mandible. Parotid was removed enmass for adequate excision with negative tissue margin. The formalin fixed tumor was a well circumscribed grayish brown mass of 3cms in diameter. The cut section of the gross was solid with few areas of hemorrhage (Fig 2). The histopathology sections prepared from the tissue showed a normal parotid structure and tumor nest being separated by a capsule (Fig 3). The tumor was well circumscribed and was arranged in lobular pattern. Each lobule comprised of inner ductal cells with eosinophilic cytoplasm and outer layers of clear myoepithelial cells. These cells were further enveloped on outside by a
well defined basement membrane material (Fig 4). There was variable degree of intervening sclerotic stroma. At places the tubules were surrounded by thicker mantles of clear cells which coalesced to form larger islands of cellular islands. There was no evidence of nuclear atypia or mitotic figures. Few ductules contained mucin in the lumen. Periodic Acid Schiff test was positive for the intraluminal mucin and basement membrane material (Fig 5). Hence, the diagnosis of Epithelial- myoepithelial carcinoma was given. In immunohistochemistry, the myoepithelial cells were 4+ positive for actin (Fig 6). Thus, the final diagnosis of Epithelial- myoepithelial carcinoma of the parotid was made.

Over the first postoperative year after the surgical excision, the patient has done well, without evidence of either local recurrence or metastasis.

Discussion

Epithelial-Myoepithelial Carcinoma (EMC) also known as adenomyoepithelioma is a rare malignant salivary gland neoplasm accounting for less than 1% of all salivary gland neoplasms 1,2 . This unusual salivary tumor, so descriptively named by Donath et al in 1972 3 , was included in the World Health Organization classification of salivary gland tumors in 1991 4 . This tumor occurs in older persons (sixth decade and beyond), and has a female predominance 6. Parotid gland is the most commonly involved site (75%), followed by submandibular gland (12%) and palate (7%) 5. The Histogenesis of EMC is uncertain and it seems to arise in two different clinical settings: either de novo or in a recurrent pleomorphic adenoma9 suggesting that there bidirectional differentiation from a stem cell to form myoepithelial and intercalated ductal epithelial cells 10. De novo EMCs arise in normal salivary gland, tend to be more aggressive with a shorter clinical history. Recurrences may not develop or may occur as a single event within a short time interval, and generally metastases develop in the lungs. Common clinical features are sudden and rapid tumor growth, superficial ulceration, bony destruction and nerve infiltration. In our case, patient presented with de novo right parotid swelling since 6 months without any evidence of metastasis. Clinically the patient was diagnosed having pleomorphic adenoma.

In our case, the cytoaspirates revealed only epithelial cells in clusters and singles in a scanty myxoid stromal background. Few spindled shaped cells were present. "Thus, it was interpreted as Pleomorphic adenoma (PA). The aspirates of epithelial- myoepithelial carcinomas have been frequently misread as pleomorphic adenoma 11. A dual cell population representing duct epithelial and myoepithelial cells with stromal substance is a feature of both EMC and PA. The latter being more common neoplasm, its diagnosis is favoured. Although the presence of double-layered tubules consisting of duct epithelial cells surrounded by myoepithelial cells is diagnostic of EMC; this pattern is not consistently observed in aspirates of EMC. A fibrillar and chondromyxoid matrix and plasmacytoid myoepithelial cells lead to the diagnosis of PA.

Most EMCs show a characteristic nodular or multinodular growth pattern and classic biphasic tubular histology of inner ductal cells with cuboidal epithelium and outer clear myoepithelial cell layers which are enveloped by basement membrane as seen in our case. The inner cells of these ductules constitute the epithelial component of EMCs. These are mild to moderately pleomorphic cells. Mitotic figures are not common. The outer cell layer is the clear cell myoepithelial component of EMCs. The clear areas are glycogen- positive and stain for periodic acid Schiff (PAS) and sensitive to diastase 12. In our case intracellular mucin and basement membrane were positive for PAS stain. The other histological variants are verocay like change, sebaceous differentiation, dedifferentiated EMC, oncocytic EMC, EMC ex pleomorphic adenoma, double-clear EMC and EMC with myoepithelial anaplasia 9. In routine histological sections, the morphologic variants of myoepithelial cells are clear, sindle, stellate, polygonal, angular, epithelioid and plasmacytoid. Immunohistochemically, epithelial component is selectively well highlighted by pan-cytokeratin (CK) and epithelial membrane antigen (EMA). Myoepithelial component is demonstrated by S100, smooth muscle actin (SMA), p63 and vimentin 13, 14, 15. The newer markers like calponin (CALP), caldesmon (CALD) and smooth muscle myosin heavy chain may be useful tools for identifying myoepithelial cells when myoepithelial cell differentiation is not easily identified on routinely stained sections 16. In our case, immunohistochemistry was done to highlight the biphasic nature of the tumor. The cytoplasm of myoepithelial cells was 4+ positive for actin. The pathological differential diagnosis includes myoepithelial carcinoma, clear cell carcinoma, and pleomorphic adenoma. In tumors with spindled cells predominance, myoepithelial carcinoma can be suggested. Myoepithelial carcinoma is not well circumscribed. Finding few ductal element rule out myoepithelial carcinoma 12. Clear cell carcinoma is also poorly circumscribed and the cells are usually more epithelioid, than those of EMC and are set in a
hyalinized stroma. The cells of clear cells lack S 100 positive myoepithelial differentiation 10. In a well circumscribed tumor, the tubular structures enveloped by myoepithelial mantles submerging in a chondromyxoid stroma suggest pleomorphic adenoma. Myoepithelial mantle melt into sea of stroma which stain positive for alcian blue 17.

Clinically, EMC usually appears as bulky, slow growing mass within parotid gland, ranging in size from 2 to 12 cms 10. The CT and MR finding of the EMC of parotid gland are non specific and EMC can not be differentiatated from more common parotid neoplasm on the basis of its imaging characteristics. Local spread to lymph nodes has been reported 6. The usual treatment is wide surgical resection, including adjacent lymph nodes. Radiation therapy may be used for tumors in which resection involves a significant cosmetic or functional deficit. Patients with EMC showing marked cellular pleomorphism, tumor necrosis; angiolymphatic invasion and perineural invasion have a poor prognosis 9. EMC is a rare low-grade malignancy with distinct cytological and histological appearance. It carries low potential for lymph node or distant metastasis but relatively high tendency for local recurrences. The biologic behavior also varies, depending on the site of involvement. But, early diagnosis is helpful for proper patient management.

References

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Illustrations

Illustration 1

Fig 1 Cytosmear showing round to oval epithelial cell clusters, few spindeloid cells (Papanicolaou, x400)

Illustration 2

Fig 2 Well circumscribed grayish brown tissue of 3cms diameter with c/s showing few areas of hemorrhage
Illustration 3

Fig 3 Normal parotid structure being separated from tumor tissue by a capsule (H&E, x100)

Illustration 4

Fig 4 Tumor lobule with inner ductal cell surrounded by outer clear myoepithelial cells which are enveloped by outer basement membrane (H&E, x100)
Illustration 5

Fig 5 PAS stain positivity of basement membrane (Periodic acid Schiff, x400)

Illustration 6

Fig 6 Myoepithelial cells showing actin (4+) positivity (IHC, x100)
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