Cystic Clear Cell Myoepithelioma of the Parotid Gland: Case Report

Parotis Bezinin Kistik Clear Cell Miyoepitelyoması

Zeynep KIZILKAYA, MD,[a]
Hatice EMİR, MD,[a]
Kürşat CEYLAN, MD,[a]
Hakan GÖÇMEN, MD,[a]
Müzeyyen ASTARCI, MD[b]

Departments of [a]E.N.T.,
[b]Pathology,
Ministry of Health Ankara Research and Training Hospital, Ankara

Geliş Tarihi/*Received:* 25.05.2007
Kabul Tarihi/*Accepted:* 28.06.2007

Yazışma Adresi/*Correspondence:*
Hatice EMİR, MD
Ministry of Health Ankara Research and Training Hospital,
Department of E.N.T., Ankara,
TÜRKİYE/TURKEY
emir.hatice@gmail.com

ABSTRACT Myoepitheliomas are rare but well-characterized group of tumors, among which myoepithelioma of the salivary glands is the best known. In this paper, a cystic clear cell variant of the parotid gland was reported in an 80-year-old male patient presenting with a slow growing mass who was treated surgically with superficial parotidectomy. To our knowledge, this is the first case of cystic clear cell myoepithelioma ever reported in the parotid gland. The histopathological features, immunohistochemical profile and clinical behavior were discussed.

Key Words: Myoepithelioma; parotid gland

ÖZET Miyoepitelyomalar, nadir ancak iyi tanımlanmış bir tümör grubudur ve içlerinde, tükrük bezlerinin miyoepitelyomaları en iyi bilinenleridir. Bu yazıda, 80 yaşında bir erkek hastada yavaş büyüyen kitle ile ortaya çıkan ve cerrahi olarak süperfisiyal parotidektomi ile tedavi edilen, parotis bezinin kistik berrak hücre varyantı bildirilmiştir. Bilgilerimiz dahilinde bu parotis bezinde şimdiye kadar bildirilen ilk kistik berrak hücre miyoepitelyoma olgusudur. Histopatolojik özellikleri, immünohistokimyasal profili ve klinik davranışları tartışılmıştır.

Anahtar Kelimeler: Miyoepitelyoma; parotis bezi

Turkiye Klinikleri J Med Sci 2009;29(1):254-7

Myoepithelial cells develop in tissues with secretory functions like the breast, the prostate, lacrimal and sweat glands, as the outer layer of acini and ductus cells of the salivary glands.[1] These cells can differentiate bidirectionally and present epithelial and myoid features. They have also contractility and accessory secretion properties.[1]

Myoepitheliomas account for less than 1% of all salivary gland tumors. This finding is contradictory with the active role of myoepithelial cells in the histogenesis of several types of salivary gland tumors (pleomorphic adenoma, adenoid cystic carcinoma, salivary ductus carcinoma).[1] Only a small number were reported to have pathological features of malignancy.[2] The term myoepithelioma was first used by Sheldon in 1943. Malign myoepithelioma was first described much later by Stromeyer et al in 1975.[3] Benign and malignant myoepitheliomas comprise a rare but well-characterized group of tumors.

Myoepitheliomas develop in different glands; the most common site is the parotid gland (50%), followed by the sublingual gland (33%) and the sub-

mandibular gland (13%).[4] The tumors usually present as a slowly enlarging, asymptomatic mass. The mean age of occurrence is over 50 years and there is no sex predilection.[3] Facial nerve is usually not involved.

In this paper, a parotid myoepithelioma with cystic formation was reported for the first time in the literature.

CASE REPORT

An 80-year-old man presented to our clinic in March 2006, complaining of a slow growing and painless mass in the right parotid gland. The mass had been gradually enlarging until the patient had recognized it for the first time 15 years ago. Physical examination revealed a semi-solid and in some parts, cystic mass measuring 8 x 9 cm in the right parotid gland. The lesion was semi-mobile, nonadherent to the underlying tissues, covered by healthy skin and cystic in formation. Facial functions were intact and there were no enlarged lymph nodes of the neck.

Fine needle aspiration biopsy (FNAB) was performed and the diagnosis of benign cystic formation was made.

A computerized tomography scan of the parotid region revealed a tumor measuring 55 x 50 x 40 mm in the right parotid area; it was a cystic lesion localized to the superficial lobe with thick septas in deep layer (Figure 1).

Superficial parotidectomy was performed under general anesthesia in March 2006. The tumor was excised with surrounding normal parotid tissue after finding the facial nerve truncus. Postoperative facial nerve functions were intact. Macroscopically the tumor was generally well-circumscribed and encapsulated with cystic areas.

Histopathological Findings:

Grossly the surgical specimen measured 9.5 x 8 x 2.5 cm. The cut surface was brown and a cystic, lobulated, hemorrhagic lesion measuring 5.5 x 3.5 cm was detected inside the tumor. The cyst was filled with serous liquid (Figure 2).

In microscopic examination, tumor tissue was separated from the adjacent normal parotid tissue by a fibrous capsule (Figure 3).

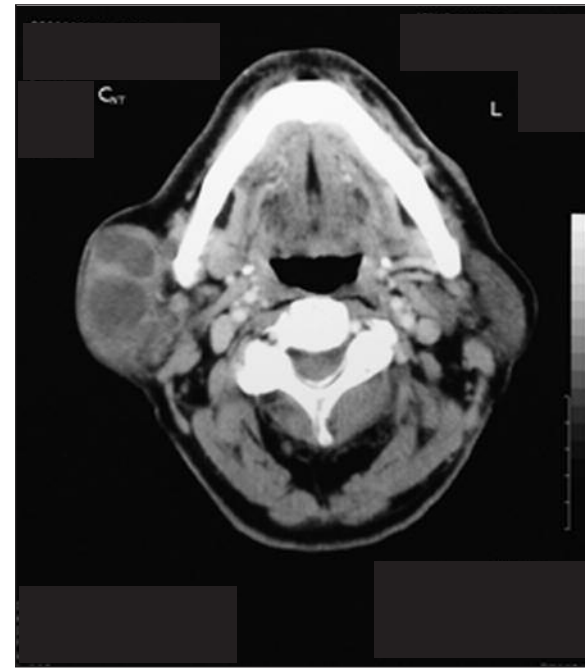

FIGURE 1: On axial computerized tomography section there was a 55 x 50 x 40 mm sized cystic mass lesion localized to the superficial lobe of the right parotid gland. Wide septations separated the cyst into lobules.

The tumor was composed of double-layered cubic epithelium and a myoepithelial cell layer underneath with clear cytoplasms. It contained hyaline-like material (Figure 4).

There was no sign of a destructive and infiltrative growth pattern and cytological atypia.

Immunohistochemical studies were carried out by cytokeratin and epithelial membrane actin EMA as epithelial markers and a positive reaction was obtained. Muscle markers such as smooth muscle actin (SMA) and S-100 protein were positively stained in the cytoplasm of myoepithelial cells. There was a positive staining of hyaline like material with PAS.

DISCUSSION

Myoepithelioma was thought to be a monomorphic variant of pleomorphic adenoma when it was first described.[1] Electron microscopic examination of myoepitheliomas revealed that the tumors were exclusively composed of myoepithelial cells.[3] While stromal variants are very rare, ductal variants occur at a 10% rate. Although the cytomor-

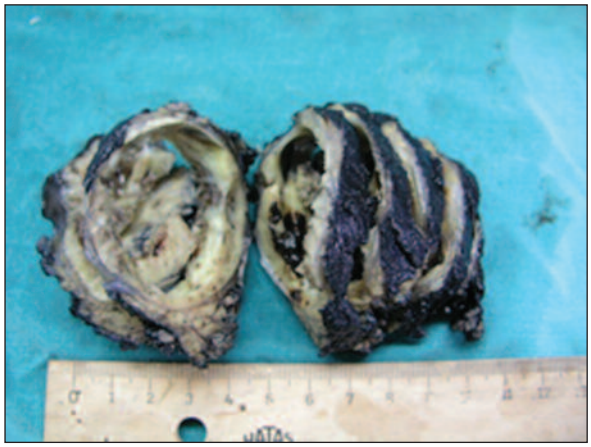

FIGURE 2: A 9.5 x 8 x 2.5 cm sized, brown colored specimen and a 5.5 x 3.5 cm sized cystic, hemorrhagic, partly multilobulated lesion. There was serous liquid in the cystic mass.

phologic features of myoepitheliomas are similar to those of pleomorfic adenomas, they usually do not have ductal and condromyxoid stroma.[3] Most of the researchers prefer to use ductal differentiation degrees to distinguish these two entities.[1] Focal ductal differentiation confirms the diagnosis of pleomorphic adenoma.[4]

The diagnostic criteria for benign myoepithelioma of salivary gland were based on the reviews of Barnes et al.[5] The traditional definition includes only solid tumors containing either spindle or plasmacytoid cells. However, with the recent appreciation of the diverse phenotypic and ultrastructural modifications exhibited by the neoplastic myoepithelial cells of different salivary gland tumors, the morphologic spectrum of myoepithelioma has expanded.[2]

In our case, the tumor was composed of little tubules, which were surrounded by double-layered cuboidal epithelial cells with a myoepithelial cell layer underneath, with clear cytoplasm. There was hyaline like material inside. The most important characteristic feature separating our case from other myoepitheliomas was the cystic formation of the tumor. Our case is the first cystic clear cell myoepithelioma in the literature.

Histologically it is difficult to differentiate myoepithelial tumors as benign or malignant. Histopathological evaluation, immunohistochemical studies and electron microscopic examinations also have limited efficacy in distinguishing benign or malignant features.[1,2] Benign myoepitheliomas are distinguished from myoepithelial carcinomas by the infiltrating and destructive growth pattern of carcinomas, cellular pleomorphism, and mitotic figures.[6] Also the multinodular growth pattern, the presence of necrotic foci, and the lack of encapsulation demonstrate the malignant nature of the tumors.[3,7] In our case, cystic tumor was encapsulated and there was no infiltrative growth pattern and cytological atypia.

Immunohistochemically, the clear cells showed a strongly positive reaction to the S100

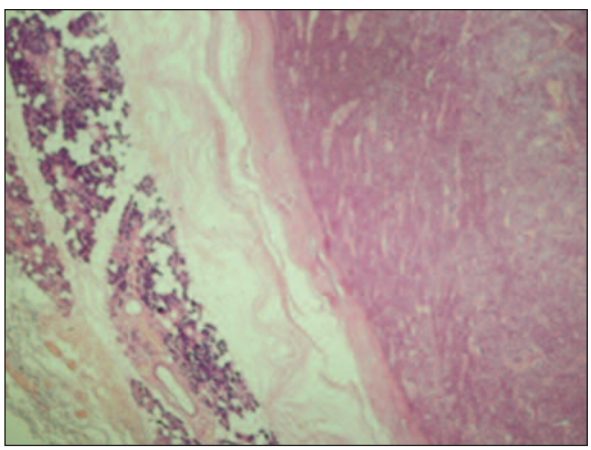

FIGURE 3: Tumor tissue was separated from the adjacent normal parotid tissue by a fibrous capsule (HE, x40).

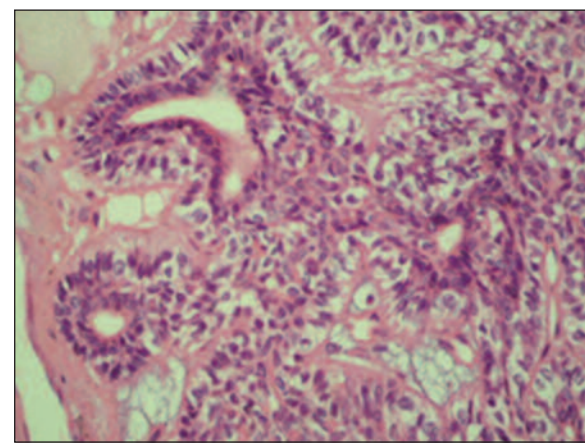

FIGURE 4: Showing double-layered cubic epithelium and a myoepithelial cell layer underneath with clear cytoplasms (HE, x100).

protein and to SMA antibodies and a focally positive reaction to vimentin antibody.[3,8,9] The clear cells present distinct myoepithelial differentiation. Therefore, a cytological diagnosis of a salivary gland tumor with clear cells requires identification of the location and confirmation of myoepithelial differentiation. Immunohistochmically, cells positive to SMA antibody are not always clear cells of the epithelial myoepithelial carcinoma.

Immunohistochemical evaluation plays an important role in the diagnosis of myoepithelioma. The myoepithelial tumor cells are positive for vimentin, S-100 protein, cytokeratin (AE1/AE3), SMA, glial fibrillary acid protein, and calponin.[3,8,9] The presence of vimentin and S-100 protein, which usually are not present in normal myoepithelial cells is a very sensitive marker of a neoplastic myoepithelium.[8,9] Because neoplastic transformation causes the myoepithelium to lose or modify its smooth-muscle phenotype, immunohistochemical studies to demonstrate smooth-muscle differentiation in these cells are inadequate. Savera et al detected smooth-muscle actin and muscle specific action in 50 and 31% of tumors respectively.[9]

In our case, immunohistochemical studies revealed that myoepithelial cells were positive for S100 and SMA (alpha smooth-muscle actin), and keratin and epithelial cells were positive for EMA (epithelial membrane antibodies). In addition, hyaline like material was positive for PAS in histochemical studies. Histologically the primary tumor was mostly composed of clear cells; therefore, the diagnosis was clear cell myoepithelioma.

Although the recurrence rates of myoepithelioma was reported to be less than those of pleomorphic adenoma, total excision of the tumor is the best approach to therapy.[5]

We performed superficial parotidectomy under general anesthesia after finding the main truncus of the facial nerve. We did not perform neck dissection as there was no palpable lymph node and the diagnosis of fine needle aspiration biopsy was benign cystic mass.

Our case of cystic clear cell myoepithelioma of the parotid gland is unique in being the first case ever reported in this localization. Therefore, cystic clear cell myoepithelioma should be considered in the differential diagnosis of the cystic neck masses in the parotid region.

REFERENCES

1. Batsakis JG. Myoepithelioma. Ann Otol Rhinol Laryngol 1985;94(5 Pt 1):523-4.
2. Di Palma S, Guzzo M. Malignant myoepithelioma of salivary glands: clinicopathological features of ten cases. Virchows Arch A Pathol Anat Histopathol 1993;423(5):389-96.
3. Stromeyer FW, Haggitt RC, Nelson JF, Hardman JM. Myoepithelioma of minor salivary gland origin. Light and electron microscopical study. Arch Pathol 1975;99(5):242-5.
4. Isogai R, Kawada A, Ueno K, Aragane Y, Tezuka T. Myoepithelioma possibly originating from the accessory parotid gland. Dermatology 2004;208(1):74-8.
5. Barnes L, Appel BN, Perez H, El-Attar AM. Myoepithelioma of the head and neck: case report and review. J Surg Oncol 1985;28(1):21-8.
6. Dardick I, Cavell S, Boivin M, Hoppe D, Parks WR, Stinson J, et al. Salivary gland myoepithelioma variants. Histological, ultrastructural, and immunocytological features. Virchows Arch A Pathol Anat Histopathol 1989;416(1):25-42.
7. Dean A, Sierra R, Alamillos FJ, Lopez-Beltran A, Morillo A, Arévalo R, et al. Malignant myoepithelioma of the salivary glands: clinicopathological and immunohistochemical features. Br J Oral Maxillofac Surg 1999;37(1):64-6.
8. Hornick JL, Fletcher CD. Myoepithelial tumors of soft tissue: a clinicopathologic and immunohistochemical study of 101 cases with evaluation of prognostic parameters. Am J Surg Pathol 2003;27(9):1183-96.
9. Savera AT, Sloman A, Huvos AG, Klimstra DS. Myoepithelial carcinoma of the salivary glands: a clinicopathologic study of 25 patients. Am J Surg Pathol 2000;24(6):761-74.