Case Report

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Epithelial-Myoepithelial Carcinoma with High Grade Transformation of Nasal Cavity

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Nazal Kavitenin Yüksek Dereceli Transformasyon Gösteren Epitelyal Myoepitelyal Karsinomu

ABSTRACT

High grade transformation in epithelial-myoepithelial carcino¬ma is a recently defined entity with only a few cases reported in the literature. We present a 59 years old female patient with a polypoid lesion in nasal cavity, together with clinical com¬plaints as recurrent epistaxis and stuffiness. Histological and immunohistochemical examination of the surgically excised le¬sion revealed a diagnosis of epithelial-myoepithelial carcino¬ma with high grade transformation. Epithelial-myoepithelial carcinoma originates from salivary glands, mainly parotid gland. Other affected sites are submandibular gland and minor sali¬vary glands. Epithelial-myoepithelial carcinoma is characterized by double layered duct-like structures. Greater cytologi¬cal atypia, higher mitotic rate, presence of necrosis and loss of the biphasic duct-like structures are features of high grade transformation. Local recurrence and cervical lymph node me¬tastases are seen with low incidence in classical epithelial-myoepithelial carcinoma, whereas epithelial-myoepithelial carcinoma with high grade transformation has high tendency for lymph node and distant metastases.

Key words: *Epithelial-myoepithelial carcinoma, high grade transformation, nasal cavity, case report*

ÖZET

Epitelyal-myoepitelyal karsinomda yüksek dereceli transformasyon, son zamanlarda tanımlanmış ve literatürde sadece birkaç vaka olarak bildirilen bir antitedir. Biz bu çalışmada tekrarlayan burun kanamaları ve burun tıkanıklığı olan, nazal kavitede polipoid bir kitlesi bulunan 59 yaşında kadın hastayı sunmaktayız. Cerrahi olarak eksize edilen lezyonun histolojik ve immünhistokimyasal değerlendirmesinde yüksek dereceli transformasyon gösteren epitelyal-myoepitelyal karsinom tanısı kondu. Epitelyal-myoepitelyal karsinom başlıca parotis bezi olmak üzere tükrük bezlerinden kaynaklanır. Etkilenen diğer bölgeler submandibuler bez ve minör tükrük bezleridir. Epitelyal-myoepitelyal karsinom çift sıralı duktus benzeri yapılarla karakterizedir. Daha fazla sitolojik atipi, daha yüksek mitoz oranı, nekroz varlığı ve bifazik duktus benzeri yapıların kaybolması yüksek dereceli transformasyonun özellikleridir. Lokal nüks ve servikal lenf nodu metastaz insidansı klasik epitelyal-myoepitelyal karsinomda düşüktür, buna karşın yüksek dereceli transformasyon gösteren epitelyal-myoepitelyal karsinomun lenf nodu ve uzak metastaz eğilimi yüksektir.

Anahtar kelimeler: *Epitelyal – myoepitelyal karsinom, yüksek dereceli transformasyon, nazal kavite, olgu sunumu*

INTRODUCTION

Epithelial-myoepithelial carcinoma (EMC) was first described by Donath et al in 1972 (1) and included in World Health Organization classification of salivary tumors in 1991 (2-5). Until this time, EMC was reported with different terminologies, such as adenomyoepithelioma, glycogen rich adenoma, glycogen rich carcinoma, clear cell adenoma and clear cell carcinoma (5).

EMC has low rates of local recurrence and cervical lymph-node metastases. Also, distant metastasis is rare (1,5,10,12). Therefore EMC is defined to be a low grade malignancy (2-4,6,8-10). The majority of these tumors

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occur in the salivary glands, predominantly the parotid gland. Submandibular gland and minor salivary glands may also be involved (1,3,5,7,8,12). EMC arising in the nasal cavity is very rare (1,8).

High grade transformation (HGT) is defined as histological progression of low grade malignant neoplasm to high-grade, in which the original line of differentiation is no longer evident (2,6). Tumors with anaplasia have been called as either dedifferentiated epithelial-myoepithelial carcinoma or EMC with HGT (4). EMC with HGT is extremely rare; only a few cases are reported as high grade EMC arising in the nasal cavity (2,4). We report a case of EMC with HGT arising in nasal cavity in order to discuss clinical, radiological, histopatological and immunhistochemical features of this rare type of tumor.

CASE

A 59 years old female was admitted to our center with complaints of intermittent epistaxis and nasal obstruction for two months. In patient's nasal endoscopic examination, left inferior turbinate was hypertrophied and purple in color. Nasal septum and the middle turbinate were normal on endoscopic examination. No associated lymphadenopathy was observed in both sides of the neck. Axial and horizontal sections on tomography scan of the paranasal sinuses revealed a mass which fills the middle and lower nasal cavity, leading to minimal pushing to the nasal septum and medial maxillary wall and invading the inferior turbinate (Figure 1). Surgical excision was planned with a diagnosis of nasal polyp or angiofibroma according to the clinical and radiological findings. Therefore endoscopic view was not saved. Under general anesthesia, the inferior turbinate together with the mass was excised endoscopically. During operation middle turbinate and septal mucosa were normal. There was no complication after the operation. The histopathologic diagnosis was EMC with HGT. Follow-up for 4 months revealed no local recurrence or metastasis.

Pathological findings; Grossly, the tumor was composed of fragmented, grayish polypoid nodule measuring 5x4x1 cm in diameter. On microscopic examination, the tumor was observed to be divided into lobules by fibrous stroma. The tumor was consisted of two components, including classical EMC and high grade EMC (Figure 2). The former component was composed of nests, duct-like and trabecular structures within a hyalinized background. Duct-like formations were lined with double layered epithelium. This epithelium was composed of a luminal layer with eosinophilic cuboidal epithelial cells, and a basal layer with polygonal - elongated cells. The tumor surface was covered with a single row of cuboidal or flattened epithelial cells. Linear PAS positive and diastase resistant material was seen in ductal lumens. The latter component was consisted of solid, cribriform and nested growth patterns with marked necrosis. Capsule of the tumor was invaded and surrounding soft tissues and bones were infiltrated. The number of mitoses was lower at classical EMC areas and higher at high grade areas, approximately 10 mitoses per 10 high power field were found.

Immunohistochemical staining was performed; epithelial cells were demonstrated by epithelial membrane antigen, and myoepithelial cells were stained for vimentin and smooth muscle actin. Myoepithelial cells and some luminal cells are positive for S-100. The tumor cells of classical component were diffusely positive for pancytokeratin. In high-grade component, the tumor cells showed patchy staining with pancytokeratin. Ki67 labeling index of classical EMC component was 5%, and index of high grade component was 25%. P53 was expressed in HGT component higher than classical component.

DISCUSSION

EMC is an uncommon neoplasm, accounting for approxi¬mately 1% of all salivary gland tumors (2-8). It

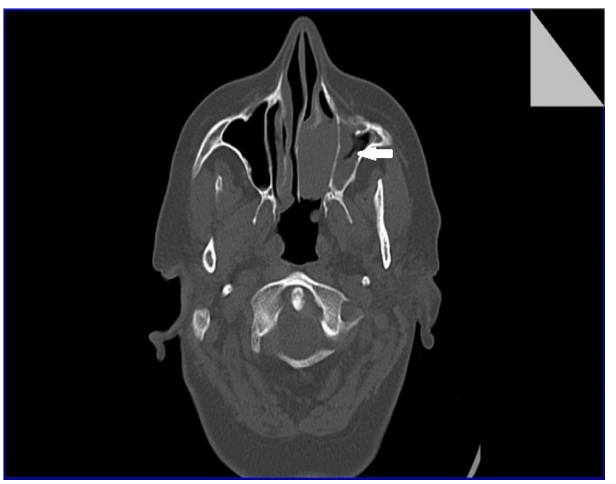

Figure 1. Axial computed tomography (soft tissue window) of the paranasal sinus demonstrating a mass (arrow) that fills the lower nasal cavity

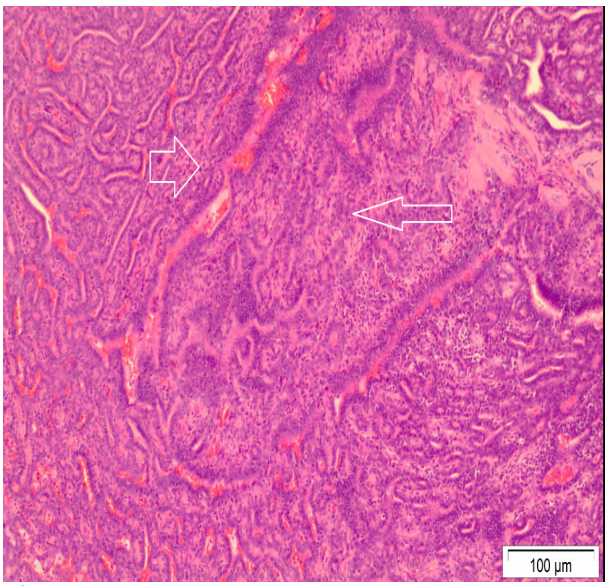

***Figure 2**. Ductal formations in the classical EMC component (thick arrow) and proliferation of solidly arranged cells in the high grade component (thin arrow) (H&E, 100x)*

is mostly seen in fifth to eighth decades and is rare in children (1,4,7). Patients are mostly female (Female/Male=2:1) (2,7,12). It is presumed that EMC origins from interca¬lated ducts of salivary glands (1,2). Tumor affects the parotid gland in 80% of cases (3,8,12). In 10% of cases, submandibular gland is the affected site (3,12). Other affected sites are minor salivary glands of the mouth, maxillary sinus, trachea, larynx, hypopharynx and lacri¬mal glands (1,7). In our case, tumor was originated from minor salivary gland of nasal cavity.

EMC forms a painless, slowly growing mass. Tumors of mi¬nor salivary glands have a tendency of forming ulcerated submucosal nodules with irregular margins (7,11). In four cases with EMC in the nasal cavity in the literature, tu¬mors were polypoid and arose from nasal septum or the inferior nasal turbinate as in our case (8).

EMC is a biphasic tumor (4,6,7,11,13). Histologically, it is characterized by double layered duct-like structures (1,2,6). Recently several histological variants have been reported as double clear EMC, oncocytic adenoma, dedif¬ferentiated EMC and EMC with anaplasia (2). High-grade transformation (HGT), formerly termed as "dedifferen¬tiation", is extremely rare in salivary gland carcinomas, including EMC (2). HGT-EMC mostly affects parotid gland, secondly submandibular gland as being similar with con¬ventional EMC (6). High grade tumors have a tendency to be painful and rapidly growing (7,11). Despite classical EMC, EMC with HGT occurs in older patients with an aver¬age age of 72 years (range=36-103) (6). Our patient was 59 years old.

EMC with HGT differs from classical EMC by greater cyto¬logical atypia, higher mitotic rate, presence of necrosis and loss of the biphasic duct-like structures. Additionally, bizarre, spindle, clear and plasmacytoid tumor cells may be observed in EMC with HGT (6). In our case, dedifferen¬tiated foci were seen adjacent to low grade areas. HGT in EMC can occur in the myoepithelial or ductal component. Yang et al. (2) reported that separating the high grade compo¬nent into ductal versus myoepithelial is subjective, un¬practical and less valuable as suggested by Roy et al. The authors and Roy at al. pro¬posed that these lessions be regarded as EMC with HGT (2). Baker et al. (9) have reported that in their case, p53 was overexpressed in high grade areas similar to case of Kusafuka et al.; and they reported Daa et al. found p53 staining in also typical EMC case in contrast. In our case, p53 was expressed more at high grade areas, but also low grade areas were stained as well. Ki-67 labeling index was higher in the high grade component, like p53 staining.

The differential diagnosis of EMC includes other salivary gland tumors which are composed of two types of cells: pleomorphic adenoma and adenoid cystic carcinoma (8). The pattern of cellular arrangement found in EMC excludes adenoid cystic carcinoma. The distribution of tumor cells in myxoid, hyalinised or condroid stroma is characteristic in pleomorphic adenoma. EMC differs by exhibiting greater epithelial cellularity with less stroma compared to pleomorphic adenoma (8).

EMC with HGT is more aggressive than classical EMC, with a high tendency for lymph node and distant metastases (2,6). A patient was reported which multiple pulmonary metastases 15 months after surgery (2). Baker et al. (9) noted a case with EMC with HGT in the nasal cavity represented recurrence at 15 months, bone metastases at 22 months in a report by Park et al. During follow up periods of 4 months after surgery, there was no local recurrence or distant metastasis in our patient. Although there is no defini¬tive treatment protocol, due to its rarity, some authors propose surgery with a safety margin, neck dissection and, radiotherapy and chemotherapy, because of it has more aggressive behavior and poorer prognosis than clas¬sical EMC (2,6). Although adjuvant radiotherapy may be appropriate, there are no data on optimal manage-

ment because of rarity (9). We reported a case of EMC with HGT, because of its rarity. Presence of only a few reports with involvement of nasal cavity in literature was an ad¬ditional specificity of our case.

In conclusion, we must emphasize the need to recognize this variant of EMC with its aggressive behavior, which may be a reason to redirect surgeons to wide local excision of tumor.

REFERENCES

1. *Lee HM, Kim AR, Lee SH. Epithelial-myoepithelial carcinoma of the nasal cavity. Eur Arch Otorhinolaryngol 2000;257(7):376-8.*
2. *Yang S, Chen X. Epithelial-myoepithelial carcinoma with high grade transformation. Int J Oral Maxillofac Surg 2012;41(7):810-3.*
3. *Lima FJ, Porto DE, Cavalcante JR, Oka SC, Godoy GP. Epithelial-myoepithelial carcinoma of high grade transformation: the case report in the buccal mucosa. Open Dent J 2012;6:111-7.*
4. *Shrivastav A, Patel UV, Gandhi S, Shah HA. A case of epithelial myoepithelial carcinoma – correlation of IHC and histopathological findings. Iranian J Pathology 2014;9(1):66-70.*
5. *Shruthi H, Sumona P. Epithelial myoepithelial carcinoma – a rare case report. Bangladesh J Med Sci 2012;11:63-7.*
6. *Nagao T. "Dedifferentiation" and high-grade transformation in salivary gland carcinomas. Head Neck Pathol 2013;7 Suppl 1:S37-47.*
7. *Thompson LDR, Goldblum JR. Head and Neck Pathology. 2nd edition. Saunders, Philadelphia, 2013: 300-4.*
8. *Yamanegi K, Uwa N, Hirokawa M et al. Epithelial-myoepithelial carcinoma arising in the nasal cavity. Auris Nasus Larynx 2008;35(3):408-13.*
9. *Baker AR, Ohanessian SE, Adil E, Crist HS, Goldenberg D, Mani H. Dedifferentiated epithelial-myoepithelial carcinoma: analysis of a rare entity based on a case report and literature review. Int J Surg Pathol 2013;21(5):514-9.*
10. *Yang S. Myoepithelial carcinoma and epithelial-myoepithelial carcinoma of the paranasal sinuses: two distinct entities. APMIS 2012;120(1):83-4.*
11. *Barnes L, Eveson JW, Reichart P, Sidransky D. World Health Organization classification of tumors. Pathology and Genetics of Head and Neck Tumors. Lyon, IARC Press, 2005: 225-6.*
12. *Mills SE, Carter D, Greenson JK, Oberman HA, Reuter V, Stoler MH. Sternberg's Diagnostic Surgical Pathology. 4th edition, volume 2. Lippincott Williams and Wilkins, Philadelphia, 2004: 950-1.*
13. *Teppo H, Paronen I. Epithelial-myoepithelial carcinoma in minor salivary gland of the hard palate. J Craniofac Surg 2008;19(6):1689-91*