

Epithelial Myoepithelial Carcinoma of Parotid Gland: A Rare Entity

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Abstract

Background: Epithelial myoepithelial carcinoma is a rare tumor that can often be misdiagnosed as benign tumors due to its deceptive nature on radiology. Therefore, histopathological examination is gold standard for diagnosis.

Introduction: Epithelial-myoepithelial carcinoma is a rare tumor of the salivary glands that was described by Donath et al in 1972.[1]

It has a slight female predilection (M:F – 1:1.6) and a wide age range from the 6th to 7th decade [2] (Mean age 64 years)

The most common site is the parotid gland, but other sites have also been described.[3]

Clinical presentation: Usually presents as a unilateral mass which is slow growing and painless.

Case Report: We present a case of Epithelial myoepithelial carcinoma.

A 36-year-old female came with painless right cheek swelling that was gradually increasing in size.

Conclusion: Epithelial-myoepithelial carcinoma is a rare carcinoma. However, it has a low metastatic rate and good survival rate post - surgical resection. Therefore,

correct diagnosis on histopathological examination is very important.

Keywords: Salivary gland tumor, Parotid gland, Biphasic tumor

Case Report

A 36-year-old woman presented with a painless right cheek swelling which was gradually increasing in size for 2 years. However, she gave no history of pain, difficulty in chewing, loss of weight or loss of appetite.

On examination, the pre auricular mass was a 6 x 4 cm firm, mobile and fixed to skin.

Ultrasound and MRI was done and both investigations were suggestive of benign neoplastic lesion in right superficial parotid gland, most likely pleomorphic adenoma.

Fine needle aspiration was done multiple times. However, it was inconclusive, showing very few clusters of epithelial cells against background of haemorrhage.

Operation

Due to cosmetic reasons patient requested to have excision done of the mass.

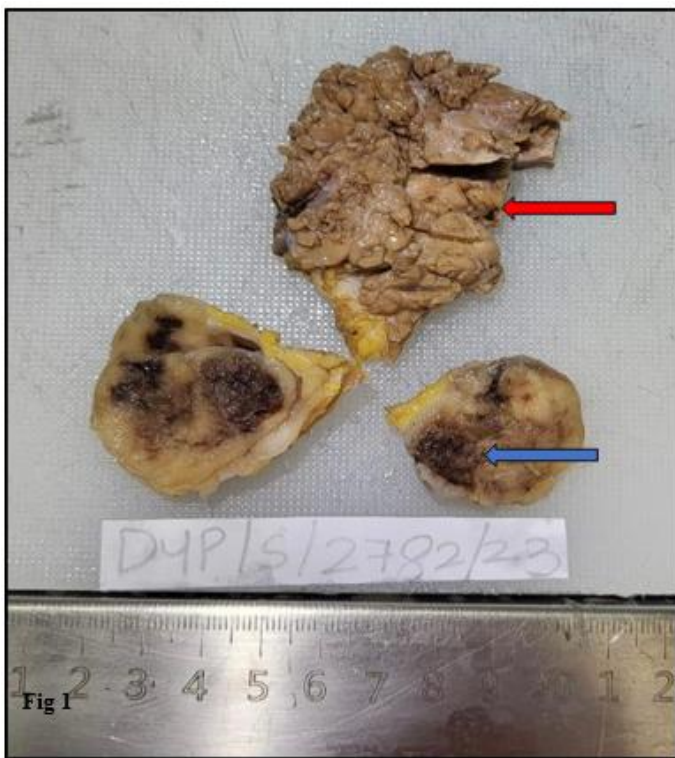
A superficial right parotidectomy was performed on the patient. The specimen was sent for histopathology examination.

Gross Examination (Figure 1): The specimen received consisted of part of salivary gland with attached fibrofatty tissue measuring 10 x 3.6 x 2.5 cm. The tumor along with salivary gland together measures 3.5 x 3.5 x 2.5 cm.

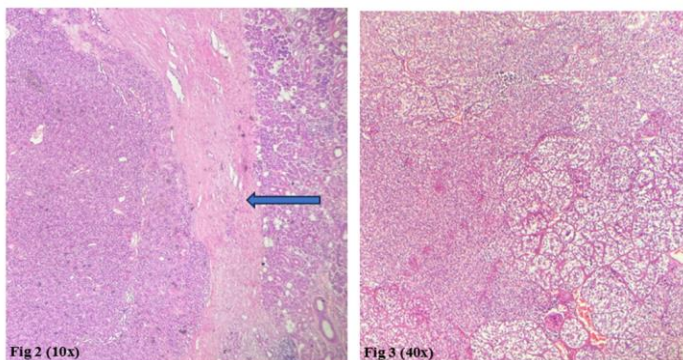
The tumor itself measured 3.5 x 3.2 x 2.5 cm. External surface of the tumor was grey – white in colour and well encapsulated. The cut surface shows a soft to firm, grey-brown, solid tumor mass with few areas of hemorrhage.



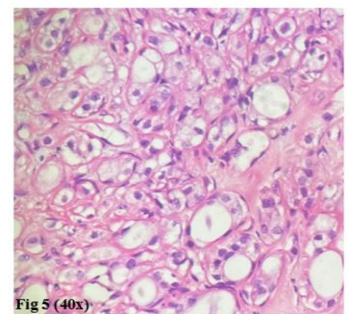
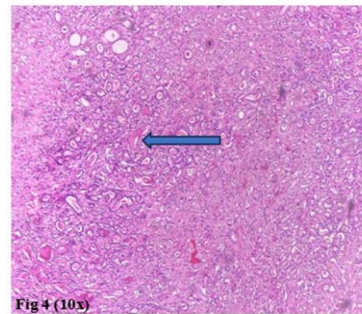
The attached muscle (red arrow) measures 5 x 4 x 1.2 cm



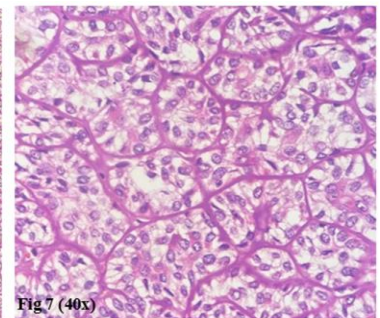
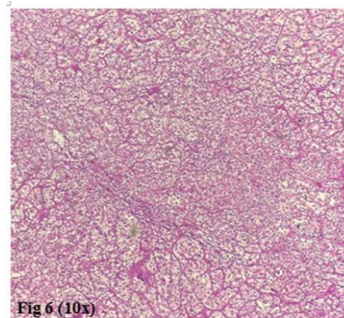
Microscopic examination



Revealed a well circumscribed tumor with a thin, fibrous capsule (Fig 2) (blue arrow). It is a biphasic tumor (Fig 3) showing a combination of epithelial and myoepithelial elements.



The epithelial element was seen arranged in nesting, glandular, tubular and solid growth patterns and lined at places by basement membrane. (Fig 4 & 5)

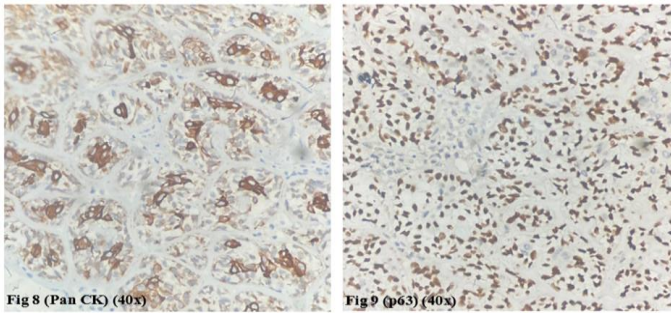


The myoepithelial element shows spindled to clear myoepithelial cells with a few intermixed myxoid areas. The individual tumor cells were mild to moderately pleomorphic, having round to oval nucleus, inconspicuous nucleoli and moderate eosinophilic cytoplasm.

Some of the glands show intraluminal eosinophilic material. (Fig 4) (blue arrow)

The tumor showed no evidence of necrosis or highly pleomorphic cells and only occasional mitosis was noted. The surrounding salivary gland tissue was unremarkable. On histopathology the diagnosis of Epithelial myoepithelial carcinoma was made.

Immunohistochemistry: To further confirm our diagnosis, immunohistochemistry was done.



Pan CK AE1/AE3 was done as epithelial marker and showed strong diffuse cytoplasmic positive in tumor cells. (Fig 8)

p63 done as myoepithelial marker and showed strong diffuse nuclear positive in tumor cells. (Fig 9)

Discussion

Epithelial-myoepithelial carcinoma of the salivary glands was described by Donath et al in 1972.[1]

It is a rare tumor with an incidence rate of 1 – 2 % and has a slight female predilection (M:F – 1:1.6). It has a wide age range from the 6th decade on. [2] (Mean age 64 years)

The most common site is the parotid gland (70%) but incidences of it have also been reported of tumor found in the minor salivary glands, maxillary sinus, trachea, larynx and hypo pharynx.[3]

Epithelial myoepithelial carcinoma has a biphasic arranged in inner luminal ductal cells with dense eosinophilic cytoplasm and outer polygonal myoepithelial cells with clear cytoplasm.[4]

With regard to recurrence, it is a low-grade tumor with a recurrence rate of 35-50% and a metastatic rate of 8.1-25%. [5]

The tumor typically has a 5-, 10-, and 20-year overall survival of 72.7%, 59.5%, and 38.3% respectively and a mean survival time of 165.5 months.[6]

Surgical treatment appears to be the mainstay of treatment, with radiation reserved for positive or close

margins or patients who are not surgical candidates or who refuse surgery.[7]

Conclusion

Epithelial-myoepithelial carcinoma is a rare carcinoma that can often be misdiagnosed as benign tumors like Pleomorphic adenoma. It has a low metastatic rate and good survival rate post - surgical resection. Radiation is needed only in rare cases. Histopathological examination stays gold standard in diagnosis.

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