Sinonasal Epithelial - Myoepithelial Carcinoma - A Rare Entity

Sultan A Pradhan • Rajan Khannan • Biswajyoti Hazarika • Meena Desai

Abstract

Epithelial- myoepithelial carcinoma is a rare salivary gland tumor. It comprises less than 1% of all salivary gland tumors. It generally arises from the parotid gland. Unusual sites of occurrence include sinonasal tract, lung, trachea, lacrimal gland and breast. Histopathologically epithelial-myoepithelial carcinoma comprises a dual population of ductal and myoepithelial cells. We report an extremely rare case of epithelial- myoepithelial carcinoma occurring in the sinonasal tract of young man.

Keywords Epithelial-myoepithelial carcinoma • Salivary gland • Sinonasal tract • Tubular structures • Cuboidal cells.

Introduction

Epithelial - myoepithelial carcinoma (EMC) is a rare, low-grade malignant neoplasm of the salivary gland, predominantly arising in the parotid gland^{1,2}. Very few cases of EMC arising in unusual sites like breast, lacrimal glands, nose, paranasal sinuses, trachea, bronchus and lung have been reported till date^{3,4,5}. We here present an extremely rare case of sinonasal EMC in a young male patient.

Sultan A Pradhan
Rajan Khannan
Biswajyoti Hazarika (✉)
Fellow, Dept of Oncology
Prince Aly Khan Hospital
Nesbit Road, Mazagaon
Mumbai - 400 010
e-mail: biswa_dr@yahoo.co.in
Dept. of Surgical Oncology

Meena Desai
Dept. of Pathology

Prince Aly Khan Hospital
Nesbit Road, Mazagaon
Mumbai - 400 010

Case report

An otherwise healthy 29 years old man reported to us in November 2005 with left sided facial swelling and left nasal block of six months duration. During this period he also suffered from occasional bouts of epistaxis. On examination a reddish polypoidal mass was seen occupying the left nasal cavity, almost occluding it. A smooth swelling of firm consistency was present in the left malar region obliterating the ipsilateral naso-labial crease but free from the infra- orbital ridge. No paraesthesia or anaesthesia was evident over the overlying skin. On posterior rhinoscopy no mass was seen in the nasopharynx. There was no palpable cervical and/or parotid lymph node.

Two years earlier the patient had undergone a lateral rhinotomy and excision of left naso-ethmoidal mass elsewhere. No details of the previous surgery were available with the patient. Following that the patient was asymptomatic for almost eighteen months after which he gradually developed the present mass and nasal blockage. The histology of biopsy from the nasal mass showed an epithelial - myoepithelial carcinoma.

A computerised tomography (Fig. 1) of the paranasal sinuses and neck revealed a faintly enhancing mass in the left nasal cavity extending to the left maxillary sinus and pushing the nasal septum towards the right. The left lateral nasal wall showed a large defect, which was due to the previous surgery. Superiorly the mass was seen extending to the ethmoidal sinuses. While the nasopharynx was free of tumor there was also no orbital and/ or intracranial extension. There was no significant cervical lymphadenopathy.

The patient underwent excision of the mass via an approach made through the previous surgical scar of lateral rhinotomy. The mass, an otherwise encapsulated one with some haemorrhage and necrosis was seen occupying the left nasal cavity and maxillary antrum. No bony destruction was noted. The mass was abutting the anterior ethmoidal sinuses but

Fig. 1 {Hematoxyline-eosine stain (40x)}

Microscopic picture of Epithelial - Myoepithelial Carcinoma,

The myo-epithelial component is represented by the cells having clear cytoplasm

Fig. 2 CT Scan PNS

Faintly enhancing mass in left nasal cavity and maxillary Antrum.

without any discrete involvement of it. The convalescence period of the patient was uneventful.

The post- operative histopathology (Fig. 2) of the mass showed a cellular tumor composed of compactly arranged small tubules lined by an inner single layer of eosinophilic cuboidal cells surrounded by layers of clear cells with hyaline-like material in between the tubules. No nuclear atypia or increased mitotic activity was noted. An intact respiratory membrane was seen overlying the tumor. Glycogen was demonstrated in the cytoplasm of the clear cells with PAS positivity digested by diastase. Based on the above findings a histopathological diagnosis of “epithelial - myoepithelial carcinoma” was made. The diagnosis was confirmed by immunohistochemistry which showed CK (cytokeratin) and SMA (smooth muscle actin) positivity highlighting the epithelial and myoepithelial component respectively while the positive S-100 protein highlighted both the epithelial and myoepithelial component

Discussion

Epithelial - myoepithelial carcinoma is a rare, low-grade carcinoma of the salivary gland, comprising about 1% of all epithelial salivary gland neoplasms^{1,2,6,7}. They are also known as clear cell adenoma, clear cell carcinoma, tubular carcinoma, glycogen rich adenoma, glycogen rich adenocarcinoma and salivary duct carcinoma. It is predominantly a tumor of the parotid gland, while rare sites of occurrence includes sub-mandibular gland, sinonasal tract, trachea, bronchus, lung, breast and lacrimal gland^{3,4,5}.

Clinical features

Though the patient was a twenty-nine years old man, in general, the mean age of occurrence of EMC is 60 years and has a slight female preponderance (60% females)⁶. In the

sinonasal region they usually present with a smooth polypoidal nasal mass and produces a malar swelling. Haemorrhage from the mass is not a common feature.

The imaging appearance of EMC is not specific (faintly enhancing lesion on CT scan).

Macroscopy

Macroscoically, EMC are well-circumscribed, gray- white to tan- white tumors. This was evident in our case. Though hemorrhage and necrosis are not common in EMCs, minimal hemorrhage and necrosis were seen intra-operatively.

Histopathologically EMC comprises a dual population of ductal and myoepithelial cells. The tubules/duct structures are lined by cuboidal cells surrounded by large polygonal clear cells containing large amount of glycogen^{2,3,4,8}. This was very much evident in the case presented here. Nuclear atypia and aneuploidy, which indicate a poor prognosis⁹, were not seen in the postoperative histopathology.

Though we had a clear- cut histological picture of EMC in both pre- and post- operative microscopy with histochemistry (PAS and PAS with diastase), the diagnosis was reconfirmed with immunohistochemistry (CK, SMA, S-100 positive). Immunohistochemistry shows cytokeratin, S-100 protein positive in neoplastic myoepithelial cells and in some cases vimentin, actin and myosin^{4,6}.

The differential diagnosis of EMC includes pleomorphic adenoma, oncocytoma, myoepithelioma, leiomyoma and metastatic renal cell carcinoma³.

Treatment

The treatment of choice of epithelial- myoepithelial carcinoma is basically wide surgical excision.

Prognosis

Local recurrence in EMC is not uncommon (30- 50%)^{2,3,9}, with our patient presenting with a recurrence within a period of two years. Metastases to local lymph nodes are seen in 8-25% cases and distal metastasis to lungs, kidney and brain in 8% cases, resulting in a similar mortality rate^{3,7,8}. In EMC recurrences and metastasis can occur a long time (> 20 years) after the first appearance, necessitating a lifetime observation of these patients.

Conclusion

Though sinonasal epithelial- myoepithelial carcinoma is extremely rare, its occurrence is still possible and both the head and neck surgeon and the surgical pathologist should be aware of this entity.

References

1. National Cancer Institute, US National institute of health, www. cancer.gov, Salivary Gland Cancer (PDQ), last modified on 12/20/2005 .
2. Brocheriou C, Auriol M, et al. Epithelial - myoepithelial carcinoma of the salivary glands. Study of 15 cases and review of literature. *Ann Pathol*, 1991, 11(5-6):316-25
3. Jin XL, Ding CN, Chu Q. Epithelial - myoepithelial carcinoma arising in the nasal cavity: a case report and review of literature. *Pathology*, 1999 May, 31(2): 148-51
4. Harada H, Kashiwagi SI, Fujiura H, et al. Epithelial - myoepithelial carcinoma: a report of case arising in the nasal cavity. *J Laryngol. Otol* 1996 April; 110(4): 397-400
5. Ryska A, Kerekas Z, et al. Epithelial - myoepithelial carcinoma of the bronchus. *Pathol Res Pract*, 1998, 194(6): 431-38
6. Ellis GL, Auclair PL. Malignant epithelial tumors in: tumors of the salivary gland, Armed Forces Institute of Pathology, Washington DC, Atlas of tumor; 3rd Edition, 1996: 15-373
7. Batsakis JG, el-Naggar AK, Luna ML. Epithelial - myoepithelial carcinoma of the salivary glands. *Ann Otol Rhinol Laryngol*, 1992; 101(6): 540-42
8. Simpson RH, Clarke TJ, Sarsfield PT, Gluckman PG. Epithelial - myoepithelial carcinoma of salivary glands. *J Clin Pathol*. 1991 May; 44(5): 419-23
9. Fonseca I, Soares J. Epithelial - myoepithelial carcinoma of salivary glands. A study of 22 cases. *Virchows Arch A Pathol Anat Histopathol*. 1993; 422(5): 389-96.