Mucoepidermoid Carcinoma Ex-Pleormorphic Adenoma of Parotid Gland: A Rare Variant with Cytodiagnostic Handicap

Parotis Bezi Mukopidermoid Karsinomu Eski Pleormorfik Adenomu: Sitodiagnostik Handikaplı Nadir Bir Varyant

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ABSTRACT

Carcinoma ex pleomorphic adenoma is a rare malignant tumor predominantly arising from salivary glands. Mucoepidermoid carcinoma ex pleormorphic adenoma (MECxPA) is a rare subtype with only ten cases reported in literature. This is a case report of a 53- year-old female with three years history of parotid swelling mimicking clinical features of a benign tumor and even eluding fine needle cytology as pleormorphic adenoma. Histopathological examination reported malignancy of MECxPA variant following superficial parotidectomy. This cytodiagnostic pitfall has been widely reported in literature hence warranting a high index of suspicion and aggressive management in addressing parotid tumors to ensure prompt and adequate treatment.

Key Words: Carcinoma ex pleomorphic adenoma, mucoepidermoid carcinoma ex pleormorphic adenoma, fine needle aspiration cytology

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ÖZET

Pleomofik adenoma karsinomu özellikle tükürük bezlerinden kaynaklanan nadir görülen malign bir tümördür. Mukoepidermoid karsinom ex pleormorfik adenom (MECxPA) literatürde bildirilen sadece on vaka ile nadir görülen bir alt tiptir. Bu, iyi huylu bir tümörün klinik özelliklerini taklit eden ve hatta pleormorfik adenom olarak ince iğne sitolojisinin atladığı üç yıllık parotis şişmesi öyküsü olan 53 yaşında bir kadına ait bir bildiridir. Histopatolojik inceleme, yüzeyel parotidektomiyi takiben MECxPA varyantında malignite olduğunu bildirdi. Bu sitodiagnostik tuzaklar literatürde geniş bir şekilde bildirilmiştir, bu nedenle hızlı ve yeterli tedaviyi sağlamak için parotis tümörlerine yönelik yüksek bir şüphe ve agresif yönetim indeksi gerektirir.

Anahtar Sözcükler: Eski pleormorfik adenoma kanseri, eski pleormorfik adenoma mukopidermoid karsinomuince iğne aspirasyon sitolojisi

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INTRODUCTION

Carcinoma ex pleormophic adenoma (CxPA) is a rare malignant tumor predominantly arising from salivary glands. It develops in 6% of all pleomophic adenomas and is defined as a carcinoma that arises in the epithelial and/or myoepithelial component of the tumor(1). This entity accounts for about 11% of all salivary gland malignancy and often involves patients in their sixth to seventh decades of life and exhibits variable gender predilection (2). The rarity of this malignancy coupled with a subtle presentation mimicking the benign counterpart poses a challenge in terms of diagnosis and timely management. We report a case of a rare variant of this entity which was a high grade mucoepidermoid carcinoma ex pleomorphic adenoma (MECxPA) of the parotid gland which was confirmed histopathologically, having missed the initial preoperative cytology assessment (3).

CASE REPORT

A 53-year-old Asian Malay female, presented with a left swelling below her left ear which was first noticed three years ago. The swelling was initially small and painless which gradually enlarged prompting her to seek medical attention. There were no episodes of redness, pain or discharge from the swelling although she did experience occasional left ear earache which was not associated with any discharge, tinnitus or dizziness. There were no constitutional symptoms. Apart from hypertension, she has been generally well with no other significant medical or surgical events. Her family history was unremarkable especially relating to malignancies. There was no history of indulgence in tobacco or alcohol consumption.

Clinical examination showed a 4x5cm swelling over left infra-auricular region extending to postauricular region. It was firm, nontender, smooth surfaced and not fixed to underlying structures.

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There were no sinuses, pits or ulcers on the overlying skin. There were no neck nodes palpable. General ENT examination otherwise was normal including rigid and flexible nasoendoscopy. Her facial nerve was intact together with all her other cranial nerves. In addition, her ear examination was also normal with normal hearing level.

Fine needle aspiration cytology (Fig. 1) was reported as pleormorphic adenoma and subsequently a contrasted CT showed left parotid tumor with local infiltration. A superficial parotidectomy was done with intra-operative findings of 4x5 cm multilobulated, well defined superficial parotid mass not involving the deep lobe. Gross examination of the specimen showed a circumscribed unencapsulated lobulated mass with focal cystic area. Areas of infiltration into the surrounding soft tissue was seen. The histopathological assessment revealed carcinoma ex pleomorphic with high grade mucoepidermoid carcinoma component. The superior margin was involved by malignant squamous epithelium (Fig. 2).

A repeated post-operative contrasted CT for deep lobe assessment and staging was done. It showed an ill defined heterogenously enhancing lesion associated with coarse calcification mainly at inferior aspect of the lesion along with adjacent fat streakiness. In addition, the lesion had clear fat plane demarcation with adjacent muscles. Multiple subcentimeter left cervical lymph nodes at level II, the largest measuring 0.8cm in short axis seen. No adjacent bone involvement or erosion. She underwent completion parotidectomy and subjected to radiotherapy.

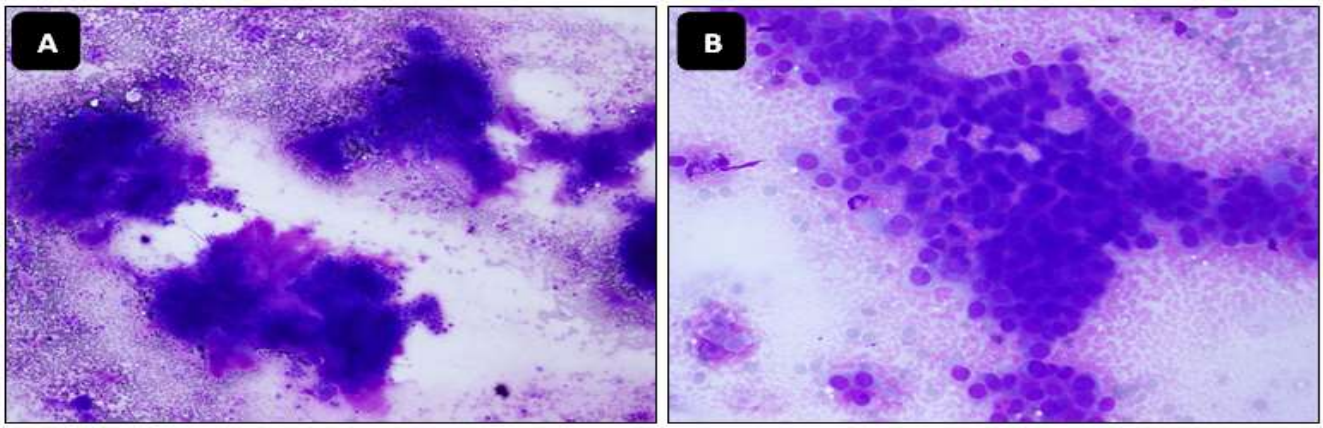

Figure 1: Photomicrograph of the fine needle cytology; A) Smears show clusters of epithelial and myoepithelial cells in chondromyxoid background (100x magnification). B) Higher magnification showing epithelial cells in small acini and cohesive clusters with bland finely granular nuclear chromatin and smooth nuclear membrane. The myoepithelial cells are plasmacytoid with well defined cytoplasm. No malignant cells seen (400x magnification).

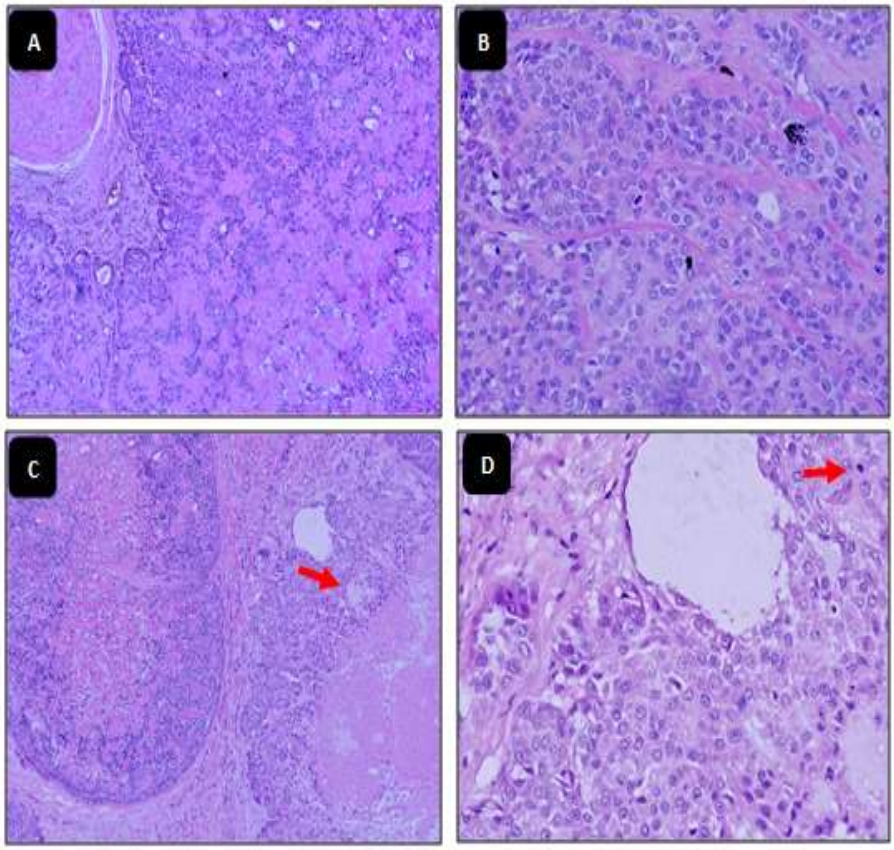

Figure 2: Histopathological examination showing features of Carcinoma ex pleomorphic adenoma with high grade mucoepidermoid carcinoma component. A) The tumour show presence of Pleomorphic Adenoma component composed of luminal epithelium and myoepithelium arranged in clusters and strands dispersed within hyalinized stroma (H&E,100x magnification). B) Higher magnification showing the luminal epithelium in clusters with myoepithelial cells seen at the basal layer (H&E, 400x magnification). C) In other areas, invasive malignant component is seen with abundant tumour necrosis (arrow) (H&E, 100x magnification). D) Higher magnification of the malignant component showing invasive epidermoid cells exhibiting pleomorphism, irregular nuclei, prominent nucleoli with eosinophilic cytoplasm. Mucous cells having vacuolated cytoplasm and mitoses are also seen (arrow) (H&E, 400x magnification).

DISCUSSION

Carcinoma ex pleormophic adenoma (CxPA) is a rare and aggressive tumor most commonly arising in the salivary glands that may present in recurrent pleormorphic adenoma post surgery or as de novo as encountered in this case(4,5). Amongst salivary gland malignancies, incidence of this subtype is low and are grouped as malignant mixed tumors comprising pleormorphic ex carcinoma, carcinosarcoma and metastasizing pleormorphic adenoma. Of these variants, pleormorphic ex carcinoma is the most common having first been described in 1957 by Beahrs et al(6).

Many different types of carcinomatous components of CxPA has been reported of which adenocarcinoma not otherwise specified is the most commonly encountered. Other types are undifferentiated carcioma, squamous cell carcinoma, salivary duct carcinoma, adenoid cystic carcinoma, mucoepidermoid carcinoma, myoepithelial and epithelial myoepithelial carcinoma(5). Of these many types, only ten cases of mucoepidermoid carcinoma variant in carcinoma ex pleormorphic adenoma has been reported thus making it a rare presentation despite mucoepidermoid carcinoma being the most common parotid malignancy in adults(2).

Pertaining to this case, the initial diagnosis was pleomorphic adenoma by FNAC as no malignant cells were seen. Based on this diagnosis, superficial parotidectomy was performed. The histopathological assessment of the surgically removed tumour showed MECxPA. The diagnosis *requires* the presence of pleomorphic adenoma component as seen in this case. The malignant cells that arose from this pleormorphic adenoma were mucoepidermoid carcinoma characterized by various population of mucuos, epidermoid and intermediate-type cells. These mucous cells were found to be interspersed between the epidermoid cells. There was no capsule seen in this tumour. In addition, the malignant cells were seen infiltrating into the adjacent soft tissue along with necrosis and haemorrhage.

Literature review done on the ten reported cases of MECxPA by Polack showed a high percentage of inaccurate fine needle aspiration cytology findings. The FNAC as a diagnostic tool proved sensitive to detect malignancy however falls short in specificity(2). The diagnostic difficulty was also caused by the malignant component being too small and elusive in gross specimen even in histopathological examination(5). Another retrospective study by Verma further underscores this pitfall concerning fine needle cytology among malignant parotid tumors(7-10).

The reliability of fine needle aspiration in parotid tumors was also stressed in another retrospective study by Altin in which the sensitivity, specificity and accuracy correlates with the size of tumor and deep lobe involvement. This study reported a false negative rate of 15% among parotid malignancies in post operative histopathological diagnosis however there was no variant of MECxPA in this study. Role of core biopsies in parotid neoplasm are dealt with caution as the high risk of tumor seeding or tumor spillage renders grave prognosis hence the advocacy for parotidectomy with adequate margins in suspected malignancies(9).

Textbook description of clinical presentation of malignant parotid tumors such as hard fixed tumors presenting with facial nerve palsy are often absent in most cases. CxPA adds to the diagnostic dilemma as de-novo cases are easily missed as they mimic benign adenomas. Hence a high clinical suspicion must be kept in all parotid tumors. Awareness of certain etiologic factors and clinical features such as age predisposition, recurrent disease, tumor size, exposure to radiation and a strong family history of malignancy will certainly add to the suspicion hence warranting careful screening for accurate diagnosis(4,5).

Treatment of parotid tumors are generally directed towards surgery though the indication and extent of surgery varies according to the cytologic or histopathologic findings(5). Most benign tumors are surgically removed especially in pleormorphic adenoma as the plausible risk of malignant transformation exist albeit small in percentage and slow to occur(4).

As for malignant tumors, the same principle applies in which early radical surgery with adequate surgical clearance along with adjuvant radiotherapy if indicated. Perineural invasion, extra-capsular spread and lymph node metastases are poor prognostic factors(1,10). The role of radiotherapy in advanced stage salivary gland malignancies are strongly advocated however the benefit among early staged malignancies are less clear(10).

CONCLUSION

It is important to be aware of such a rare variant of CxPA and more importantly to the diagnostic pitfall in regards to limitation of cytodiagnosis via fine needle aspiration. Insidious parotid swelling should be addressed with high index of suspicion hence is advisable to always offer the option of parotidectomy to patients to avoid missed malignancies.

Conflict of interest

No conflict of interest was declared by the authors.

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