Case Report

Cytologic features of microcystic adnexal carcinoma

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Abstract

Microcystic adnexal carcinoma (MAC) is an uncommon skin neoplasm with a predilection location around the lips. It is characterized by cords and nests of neoplastic cells forming ductular or glandular structures that are embedded in dense collagenous stroma. An eighty-seven year old Caucasian female patient presented with a painless, slowly enlarging mass measuring 3.3 x 2.7 x 1.0 cm on the lower lip for approximately 6 months. The patient underwent 2 fine needle aspiration biopsies (FNAs). Smears made from both FNAs demonstrated similar features, including low cellular smears, three dimensional cell clusters forming a glandular structure, round to oval cells with high N:C ratio, occasional cytoplasmic lumens, without distinct hyperchromasia, focal inconspicuous nucleoli, smooth regular nuclear membranes, abundant naked nuclei, occasional squamoid cells and focal acellular stromal fragments in the background. The cytologic differential diagnosis included skin adnexal carcinoma and low grade mucoepidermoid carcinoma arising in the minor salivary gland. The mass was subsequently excised. The diagnosis of microcystic adnexal carcinoma was made. We report cytologic features of MAC and also suggest that MAC can possibly be diagnosed by FNA with the appropriate clinical vignette and immunohistochemical profile..

Key words: Dermatology, microcystic adnexal carcinoma, skin neoplasm

INTRODUCTION

Microcystic adnexal carcinoma (MAC) is an unusual skin neoplasm. It was first described as a distinct entity by Goldstein et al. in 1982.[1] The authors reported six cases which demonstrated similar features, including islands of basaloid keratinocytes with occasional horn cysts and abortive hair follicles in a desmoplastic stroma. Similar cases might have been previously reported as malignant syringoma. A total of 223 cases of MAC were identified by Surveillance, Epidemiology, and End results registry 1973–2004. They were able to identify the predilection site of this entity for the head and neck area (74%).[2]

CASE REPORT

An 87-year-old Caucasian female patient presented to the ear nose and throat (ENT) clinic with a painless, slowly enlarging mass on the lower lip for approximately 6 months. She denied a history of previous malignancies. Physical examination revealed a firm, flesh-colored, indurated thick plaque measuring 3.3 x 2.7 x 1.0 cm, located 0.5 cm away from the lower vermilion border. There were no epidermal changes. No lymphadenopathy was noted. The patient underwent two fine-needle aspiration (FNA) biopsies, each one month apart. Both FNAs revealed similar findings, including paucicellular smears, round tight three dimensional cell clusters, round to oval cells

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with high N: C ratio, occasional cytoplasmic lumens, without distinct hyperchromasia, focal inconspicuous nucleoli, smooth regular nuclear membranes, abundant naked nuclei, and focal acellular stromal fragments in the background. Occasional squamoid cells were identified [Figure 1a-c]. The diagnoses were reported as "Epithelial neoplasm, favor adnexal tumor." The differential diagnosis included salivary gland neoplasm, especially a low-grade mucoepidermoid carcinoma. The patient received a wide excision. On histopathological examination, the tumor consisted of islands and cords of cells with mild atypia, as well as occasional tadpole-like ductular formation and microcysts embedded in a dense desmoplastic stroma. The neoplasm infiltrated the underlying skeletal muscle. Focal horn cysts were present, as well as perineural infiltration. The tumor extended close to the deep margin [Figure 2a-c]. Periodic acid-Schiff diastase (PAS-D) and mucin stain failed to demonstrate mucin deposit in the lumen of the ductular structure. The tumor cells were immunohistochemically reactive to AE1/AE3, but nonreactive to S-100 and BerEP4 [Figure 2d]. The diagnosis of MAC was made.

DISCUSSION

MAC is an uncommon, locally aggressive adnexal neoplasm. The tumor occurs mostly on the face, especially the lips.【3】MAC has also been reported to occur on perianal areas.【4】

After conducting an extensive literature search, we found that the cytologic features of MAC by FNA have been described once by Orell et al. The authors reviewed the case that had been misdiagnosed as infiltrating basal cell carcinoma on both FNA and surgical biopsy. They described the tight clusters of basaloid cells with microtubular structure and squamous differentiation.【5】Also, only a handful of FNA diagnoses of benign adnexal neoplasms

Figure 1: a) A three dimensional cluster of epithelial cells with low-grade atypia form a glandular structure with intraluminal content. The cells demonstrate high N : C ratio (Pap x400); b) Occasional basophilic stromal fragments are identified (Diff Quik x400); c) A small cluster of neoplastic cells with cytoplasmic vacuoles with several stripped nuclei in the background are identified (Diff Quik x400)
squamous carcinoma of the breast (LASC) which is a rare entity. The histologic characteristics of LASC include "tadpole"-shaped ductal formation composed of epithelial cells with low-grade atypia, bland stroma, and epidermoid cells. And these findings are very similar to those of MAC. Theoretically, the FNA findings of LASC should be similar to those of MAC as well. Ferrara et al. described a moderately cellular smear with prevalent small clusters of overlapping ductal cells. The epithelial cells are small and monotonous. They exhibit inconspicuous nucleoli. Additionally, they identified a number of fibroblast-like spindle cells singly and in clusters.

We noticed some similarities among the cytologic features of MAC in this current study, the case that was reviewed and described by Orell et al., and those of LASC described by Ferrara et al. These include dual population of cell, which include both epithelial and stromal cells.
Ductular formation is observed with round cells showing low-grade atypia. We believe that there may possibly be enough morphologic evidence to establish diagnostic criteria for MAC. We suggest that with the appropriate clinical vignette, the diagnosis of MAC could be made. Table 1 describes the diagnostic features of MAC that we would like to propose.

There have been a number of studies of immunohistochemistry on MAC and its mimickers, including DT and MBCC. Cytokeratin (CK) 15 can be helpful in distinguishing MAC from BCC. Hoang et al. demonstrated that 92% of MAC and 100% of trichoepithelioma were immunohistochemically reactive to CK15, whereas 0% of BCC showed immunoreactivity to CK15. They also reported that 38% of MAC, 57% of DT, and 100% of MBCC were positive for BerEP4. Another similar study by Smith et al. reported that all of their cases of MAC, MBCC, and DT were all negative for BerEP4. Our current case is also negative for BerEP4. Smith et al. also demonstrated that a small percentage of tumor cells in two of ten MAC cases were positive for P53, whereas 0% of DT were positive for P53. Statistically, none of the immunohistochemical markers performed in this study was proven to be useful for distinguishing between MAC and DT.

Although it is difficult to exclude the possibility of DT when facing the cytologic challenge of MAC despite the aid of immunohistochemical studies, it is still possible to distinguish MAC from DT by using the clinical manifestations. DT commonly presents on the cheek and forehead, in contrast to MAC which mainly occurs around the lips. The clinical appearance of DT is also different from MAC. DT typically manifests as annular dermal papules or plaques. MAC usually presents as ill-defined round papules or plaques without central depression; therefore, the clinical context including the location and appearance of the lesion is essential for making the diagnosis.

The treatment options for MAC include an excision, Mohs surgery, and radiotherapy. Although Mohs surgery has been reported to be the preferred method, standard treatment is still unclear.

In conclusion, we report the cytologic features of MAC that have been described in the literature only once. We also suggest that MAC can be diagnosed by FNA with the appropriate clinical vignette and immunohistochemical profile. But the diagnosis should be confirmed by a conventional biopsy due to limited studies on the cytologic diagnosis of MAC.

**COMPETING INTEREST STATEMENT BY ALL AUTHORS**

The authors declare that they have no competing interests

**AUTHORSHIP STATEMENT BY ALL AUTHORS**

Each author acknowledges that this final version was read and approved. All authors of this article declare that we qualify for authorship as defined by ICMJE http://www.icmje.org/#author. Each author has participated sufficiently in the work and take public responsibility for appropriate portions of the content of this article.

**ETHICS STATEMENT BY ALL AUTHORS**

Our institution does not require approval from the Institutional Review Board for a case report without identifiers.

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