Emphasizing the pivotal role of fine-needle aspiration cytology in a case of recurrent malignant chondroid syringoma

ABSTRACT
Fine-needle aspiration cytology (FNAC) features of malignant chondroid syringoma (MCS) are rarely documented. Here, we report a case of recurrent MCS, highlighting its interesting clinicopathologic features. Initially, we received cytology and histopathology slides (for review) of a 57-year-old woman who had undergone resection for an occipital MCS and later presented with recurrence. On reviewing the slides, cytology was consistent with recurrent MCS. However, tissue sections showed features of a malignant epithelial tumor with comedonecrosis and sebaceous differentiation owing to which a diagnosis of metastatic adenocarcinoma/malignant adnexal tumor was suggested. Due to an ambiguous histology, a repeat FNA was performed to perform immunocytochemistry (ICC) and oil-red O stains, which confirmed the diagnosis of MCS with sebaceous differentiation. Thus, in a situation where histology was inconclusive due to lack of representative sections, FNAC played a major role in resolving the diagnostic dilemma and facilitating an appropriate clinical management.

Key words: Chondroid syringoma; cytology; fine-needle aspiration; malignant; metastasis; satellite nodules

Introduction
The role of fine-needle aspiration cytology (FNAC) in the diagnostic workup of cutaneous adnexal tumors is limited. However, tumors such as chondroid syringomas (CSs) are often diagnosed accurately on cytology. Most CSs are benign (BCS) although rare malignant chondroid syringomas (MCSs) are known to occur chiefly in the extremities of young women. Distinction between BCS and MCS is often difficult on cytology although, rare cases of preoperatively diagnosed MCSs are on record. Here, we report an interesting case of recurrent MCS, highlighting the conclusive role of FNAC in an inconclusive clinical situation.

Case Report
We received May-Grünwald-Giemsa (MGG) and Papanicolaou (Pap)-stained cytologic smears of a 57-year-old female presenting with progressively increasing recurrent swellings in the upper cervical and occipital regions for review. The swelling in the occipital region was the recurrence of a previously excised tumor diagnosed to be a “malignant chondroid syringoma” while the upper cervical swelling was suspected to be metastatic. They were firm-to-hard, fixed to the underlying structures, and measured 3 × 3 cm in their maximum dimension. Surrounding these swellings were a few tiny satellite nodules.
nodules of 2-3 mm diameter. Smears were highly cellular with a striking mucoid and chondromyxoid/fibromyxoid background, within which enmeshed were clusters as well as tissue fragments of malignant-appearing round to polygonal cells [Figure 1a] and a few discrete plasmacytoid cells. We agreed with the prior diagnosis of MCS after excluding the possibilities of metastatic adenocarcinoma, extraskeletal chondrosarcoma, and chordoma. Subsequently, histologic sections from a single tissue block were also received, which showed unremarkable epidermis and a well-delineated neoplasm in the dermis. Tumor cells were arranged in nests and cords with large areas of comedonecrosis and prominent lymphovascular emboli in the superficial papillary dermis. Also seen was an occasional focus of epithelial cells reminiscent of sebaceous differentiation. Owing to the lack of representative sections, a differential (histologic) diagnosis of metastatic adenocarcinoma and malignant skin adnexal tumor was suggested [Figure 1b]. In this inconclusive situation, clinicians rereferred the case for ancillary tests on additional FNAC material.

Gel-like material was obtained on repeat FNA. Apart from MGG and Pap staining, immunocytochemistry (ICC) was performed on smears for pan cytokeratin (CK), epithelial membrane antigen (EMA), S-100, calponin, and α-smooth muscle actin (α-SMA). In view of suspected sebaceous differentiation on histology, an oil-red O stain was also performed. Smears were markedly cellular with abundant chondromyxoid/fibromyxoid matrix, and easily appreciable plasmacytoid cells. Occasional cell clusters revealed sebaceous differentiation with fine cytoplasmic, as well as background vacuolation, which was confirmed by oil-red O stain [Figures 2b and c]. Most neoplastic cells were round to polygonal with moderate to abundant cytoplasm and indistinct cell borders. There was a moderate nuclear pleomorphism with a high nucleus to cytoplasmic (N:C) ratio, coarse chromatin, irregular nuclear membrane, and two to three prominent nucleoli. The nuclear features were better appreciated with Pap-staining. A few bizarre cells, occasional tumor giant cells, and the cells mimicking adenocarcinoma [Figures 2d and e] were also noted. Tumor cells showed strong positive expression of EMA, S-100 [Figures 2f and g], calponin, pan CK, and α-SMA, establishing the diagnosis of MCS.

Discussion

Chondroid syringomas (CSs) are uncommon tumors with only rare case reports available in the cytology literature. The cytologic features described are the prominent mesenchymal elements with chondroid appearance, clusters of monomorphic epithelial cells with moderate to abundant cytoplasm, and plasmacytoid myoepithelial cells embedded in a metachromatic ground substance. Also described are the clusters and sheets of small cells with relatively scant cytoplasm, well-defined nuclear membrane, evenly distributed granular chromatin, and occasional small chromocenters. Their histochemical and immunohistochemical (IHC) features such as stromal positivity with alcian blue and mucicarmine stains, epithelial

![Figure 1](image1.png)  ![Figure 2](image2.png)
cell expression of CK and EMA, and S-100 positivity of myoepithelial cells have also been documented both on FNAC and histology.[5,9] According to Masood and Hardy,[3] BCSs are not reliably distinguishable on cytology while Mishra and Agarwal[6] described features such as markedcellularity, hemorrhagic background, and highly pleomorphic, discohesive, epithelial cells arranged in ill-formed cords as evidence of malignancy. The size and location of CS are also considered important in distinguishing BCS versus MCS.[9]

Some of the aspects of the present case are interesting. Clinically, the lesion involved the soft tissue of the head and neck region in an elderly female in contrast to its usual location of extremities in young female patients. As for the tissue diagnosis, although we agreed with the outside diagnosis of MCS, in view of the tumor location and cytologic features such as chondromyxoid and mucoid stroma, we had to exclude the possibilities of chordoma, extraskeletal chondrosarcoma, and metastatic adenocarcinoma. The absence of characteristic “physaliphorous cells” argued against a diagnosis of chordoma. Myxoid stroma with spindle cells and absence of chondrocytes embedded within a classic chondroid matrix excluded chondrosarcoma. Despite the presence of mucinous material and vacuolated cells, the prominent myxoid stroma excluded metastatic adenocarcinoma. Other myxoid sarcomas were excluded by the presence of distinct epithelial and myoepithelial cells. However, in view of the misleading histology, we proceeded with ICC on repeat FNA material that established the diagnosis. Notably, ICC findings assisted in excluding all possible differential diagnoses including a remote possibility of sarcomatoid carcinoma. The sebaceous differentiation was well-demonstrated with oil-red O stain on cytologic smears. Although not essential for diagnosing CS, pathologists should be aware of this uncommon and interesting finding.

Most cases of MCS present with local recurrence; some cases show regional lymph node or osseous metastasis. Clinically, the presence of satellite nodules and involvement of deep structures are indicative of malignancy; this was the situation in our case as well. Often, the histology of MCS is similar to that of BCS[4] although anaplasia in CS is always associated with an aggressive course, distant metastasis, and a decreased 5-year survival rate.[10]

**Conclusion**

To conclude, we have documented a rare case of recurrent MCS of the head and neck region in an elderly woman. A cytopathologist familiar with its classic cytomorphology and differential diagnosis can assist in its management, even in tricky situations. ICC performed on cytologic smears assists in its accurate diagnosis. Sebaceous differentiation in MCS is an uncommon event, which can be demonstrated on cytologic smears.

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**Conflicts of interest**

There are no conflicts of interest.

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