A Once Thought Lipoma Turned Malignant Chondroid Syringoma

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INTRODUCTION
A chondroid syringoma is a benign skin appendageal tumor with an incidence of <0.098%. Its malignant counterpart, malignant chondroid syringoma (MCS), is a tumor that arises from cutaneous sweat glands and occurs on the extremities or trunk, more commonly in women, with only 51 previously reported cases. We report a case of a 65-year-old female with an elbow mass previously thought to be a lipoma but was MCS.

CASE
A 65-year-old female with a history of rheumatoid arthritis presented with a 10-year history of a right elbow mass that over the last four months had progressively become larger and painful with a developing purple/red color overlaying the skin. The mass was previously thought to be a lipoma. An MRI joint revealed a 4.1x2.1x4.3 cm solid superficial soft tissue mass over the posterior lateral aspect of the proximal forearm, with few areas of internal hemorrhage, bilobed in appearance extending to the skin surface, and abutting the superficial fascia distally. There was no deep compartment extension or intra-articular involvement. The patient then underwent a biopsy of the mass which revealed a relatively large tumor with several populations of cells including large aggregates of basloid cells that show zonal and single cell necrosis, mitotic activity, and nuclear atypia. Additionally, other parts of the tumor showed proliferation of epithelial cells that form ductal spaces. The biopsy revealed a malignant adnexal tumor that favored the diagnosis of a malignant mixed tumor consistent with MCS with multifocal lymphovascular invasion. The lesion involved the surgical margin and she ultimately had to undergo a re-excision that then showed clear margins. A follow up whole body bone scan showed focal increased uptake in the midshaft of the right radius which could represent metastasis, however, this was not at her surgical site. An MRI of the right upper extremity did not show any mass in the area of uptake on the bone scan. A CT scan post excision revealed no findings of metastatic disease or lymphadenopathy. The patient was initiated on treatment with intensity modulated radiotherapy.

CONCLUSIONS
MCS are rare skin tumors that can arise from benign lesions and be aggressive. They can recur locally in up to about 50% of patients and metastasize in up to 60% of patients in areas such as lymph nodes, lung, bone, and brain. Histological features suggestive of malignancy include cytologic atypia, infiltrative margins, satellite tumor nodules, tumor necrosis, and involvement of deep structures. This is an exceedingly rare tumor with case reports in the literature, but no standardized treatment. Definitive treatment requires surgical removal, but adjuvant treatment is not clearly defined. Given the rarity of the tumors there is no data for potential benefits of radiation or chemotherapy after surgery. However, radiotherapy has been used both to the primary site, as with other adnexal tumors, and for lymphadenopathy. Although rare, it is an important diagnosis to consider in evolving skin masses. This is the 52nd case of MCS described in literature.

REFERENCES
2. Zufall A, Mark E, Gin A. Malignant chondroid syringoma: A systematic review. Skin health and disease. 2022 Jul

Figure 1: H&E stain biopsy demonstrating variable growth patterns
Figure 2: H&E stain biopsy demonstrating high mitotic count
Figure 3: H&E stain biopsy demonstrating skin and tumor nests with comedonecrosis