Unusual EMA and pan-keratin negative epithelioid sarcoma

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Objectives & methods

Epithelioid sarcoma is a rare malignant tumor that is positive for EMA and pan-keratin, with frequent loss of SMARCB1 expression. We present the case of a 65-year-old female patient with lower back pain and an ultrasound scan that showed an expansive formation in the abdomen with a diameter of 5.2 cm. Cytopuncture and core biopsy were performed under CT control.

Cytological findings

Cytologically, the smear showed a monomorphic population of poorly differentiated malignant cells. The cells formed clusters within which a homogeneous substance could be seen (**Figure 1**). Immunocytochemistry showed weak S100 positivity, while CK7 and CK20 were negative. It was concluded most likely to be a tumor of mesenchymal origin.

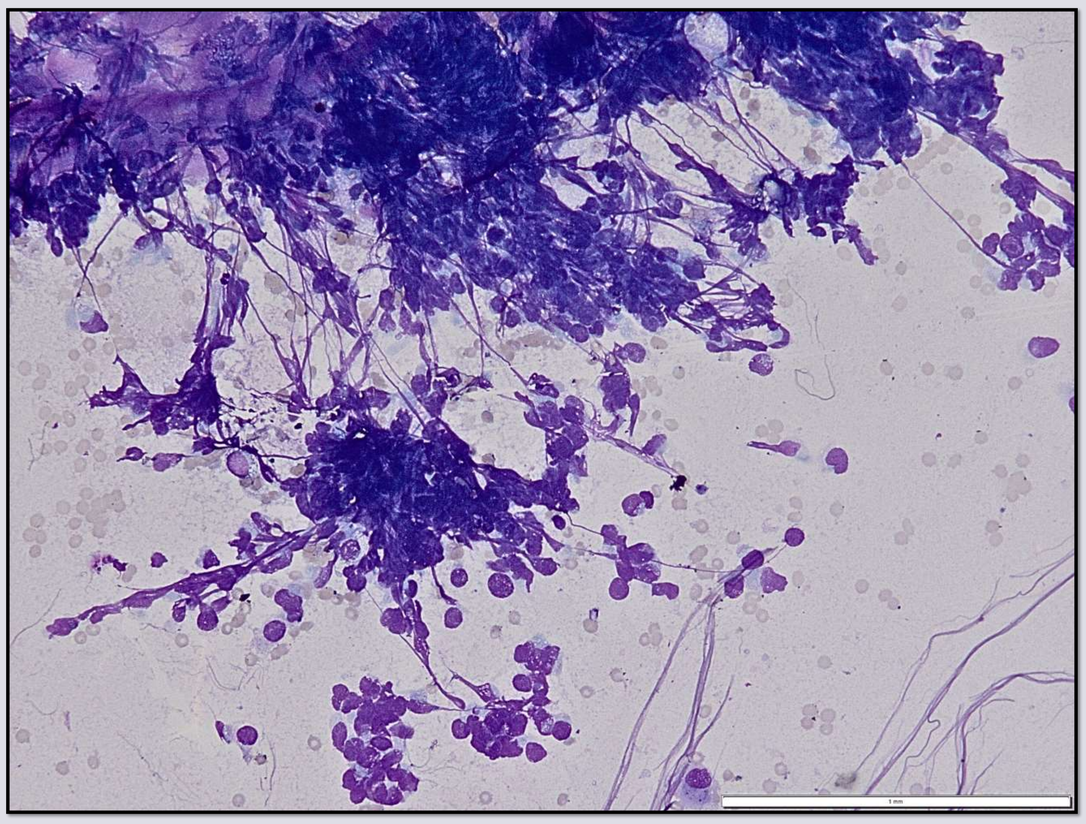


Figure 1: Poorly differentiated malignant cells within a homogenous substance (MGG, 200x)

Pathological findings

Histology showed clusters of uniform cells with scant cytoplasm (**Figure 2**), which were immunohistochemically negative for S100, melan A, CD45LCA, CKAE1/AE3, EMA, CD 138, factor VIII, desmin, SMA, MSA, chromogranin and synaptophysin, while CD99 was positive. For additional processing, prof. Abbas Agaimy, MD, PhD, (University Hospital Erlangen, Germany) was consulted, who was of the opinion that „this is a SMARCB1-deficient malignant neoplasm with rhabdoid features and an unusual complete absence of EMA and pan-keratin expression.“ He was also of the opinion that „primary tumor site in other organs should be excluded, after which the diagnosis of keratin-poor epithelioid sarcoma can be accepted.“

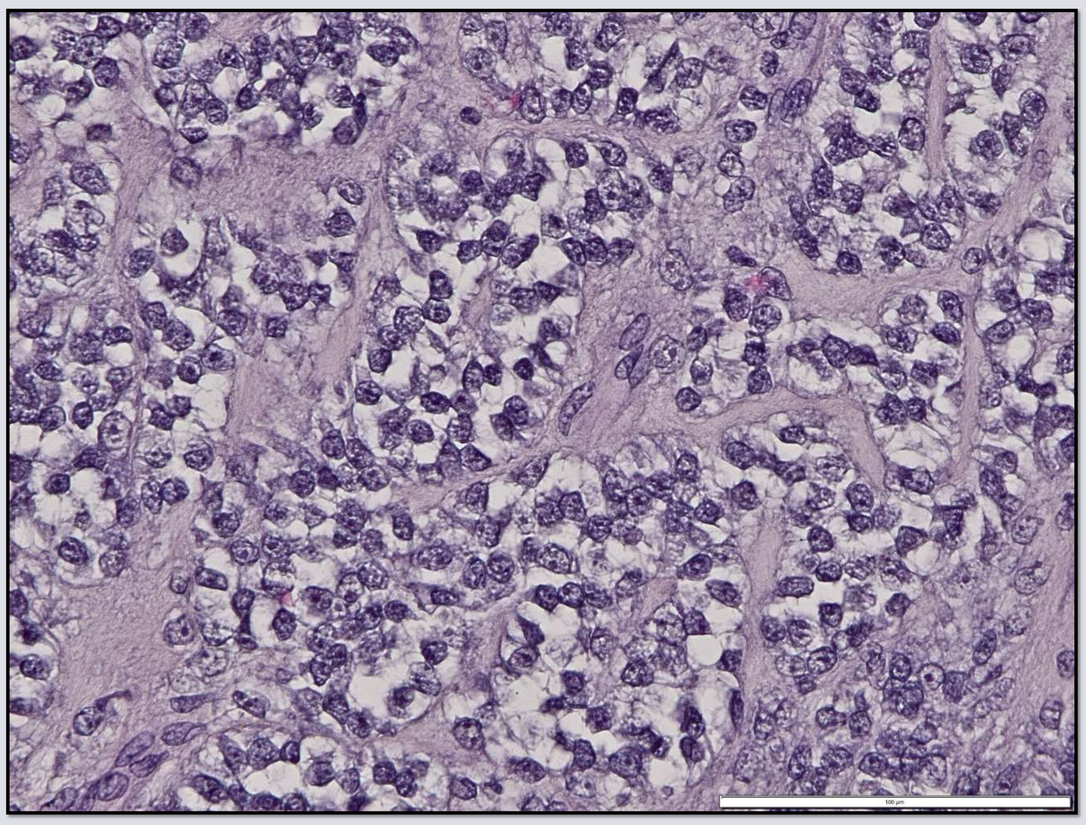


Figure 2: Clusters of uniform cells with variable clear cell morphology (H&E, 400x)

Conclusion

Further research is needed to better understand the role of SMARCB1 loss of expression in tumor immunogenicity, aiming to develop optimal immunotherapeutics and new treatment strategies for these aggressive tumors.

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