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CORRESPONDENCE
Dr Andrea Alpeza

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POSTER COMMUNICATION · ONCOLOGY

Unusual EMA and pan-keratin negative epithelioid sarcoma

Dr Andrea Alpeza[1]*, Dr Katarina Gregorović[1], Dr Darija Alpeza[2,3], Dr Zlatka Radičević[2,3], Dr Nikolina Kurtović[2], Dr Luka Klobučarić[2], Dr Branka Lončar[1,3]

[1] Clinical Institute of Clinical Cytology, Clinical Hospital Center Osijek, Croatia
[2] Clinical Institute of Pathology and Forensic Medicine, Clinical Hospital Center Osijek, Croatia
[3] J.J. Strossmayer University in Osijek, Faculty of Medicine Osijek, Croatia
* Corresponding author: Dr Andrea Alpeza

KEY MESSAGES

1. Epithelioid sarcoma can present with unusual absence of EMA and pan-keratin expression.
2. SMARCB1 loss is a key feature in this rare malignant neoplasm with rhabdoid characteristics.
3. Accurate diagnosis requires exclusion of other primary tumor sites and expert pathological consultation.

ABSTRACT

Introduction. Epithelioid sarcoma is a rare malignant tumor typically positive for EMA and pan-keratin, often showing loss of SMARCB1 expression. This case report presents a 65-year-old female patient with a 5.2 cm abdominal mass detected by ultrasound.

Materials and methods. Cytopuncture and core biopsy were performed under CT control. Cytological and histological analyses were conducted, including immunocytochemistry and immunohistochemistry to characterize the tumor cells.

Results. Cytology revealed monomorphic poorly differentiated malignant cells forming clusters with a homogeneous substance. Immunocytochemistry showed weak S100 positivity and negative CK7 and CK20. Histology showed uniform cells with scant cytoplasm, negative for multiple markers including EMA and pan-keratin, but positive for CD99. Expert consultation diagnosed a SMARCB1-deficient malignant neoplasm with rhabdoid features and absence of EMA and pan-keratin expression, consistent with keratin-poor epithelioid sarcoma after excluding other primary tumor sites.

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

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POSTER

Unusual EMA and pan-keratin negative epithelioid sarcoma

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[1]Clinical Institute of Clinical Cytology, Clinical Hospital Center Osijek, Croatia
[2]Clinical Institute of Pathology and Forensic Medicine, Clinical Hospital Center Osijek, Croatia
[3]J.J. Strossmayer University in Osijek, Faculty of Medicine Osijek, Croatia

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.

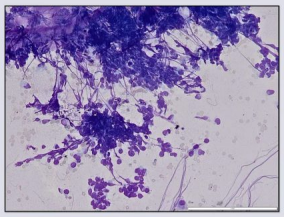


Figure 1: Poorly differentiated malignant cells within a homogenous substance (MGG, 200x)

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."

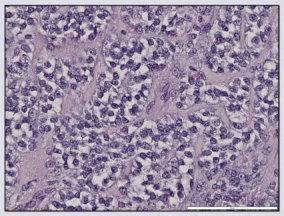


Figure 2: Clusters of uniform cells with variable clear cell morphology (H&E, 400x)

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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