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Epithelioid angiosarcoma of bone and soft tissue: a report of seven cases with emphasis on morphologic diversity, immunohistochemical features and clinical outcome

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ABSTRACT

Aims and background. Epithelioid angiosarcoma is a rare histopathologic variant of angiosarcoma characterized by an epithelioid morphology. This subset can histologically mimic non-vascular neoplasms and impose serious challenges in reaching a correct diagnosis, especially in the context of limited tissue sampling (e.g., needle core biopsy). To improve recognition of epithelioid angiosarcoma – and the spectrum of morphologic diversity associated with this rare variant – and to avoid a misdiagnosis, we describe the clinical, histopathologic, and immunohistochemical findings of cases of epithelioid angiosarcoma diagnosed at our institution.

Methods and study design. Seven cases of epithelioid angiosarcoma with appropriate pathologic material were identified from our archives. Immunohistochemistry was used to detect the expression of CD31, CD34, Factor VIII, cytokeratin, epithelial membrane antigen, vimentin, HMB45, CD1a, CD68, lysozyme, CD45, desmin, and smooth muscle actin in all cases. Follow-up information was obtained by reviewing medical records or by direct communication with family members.

Results. The lesions involved the bone (n = 4) and soft tissues (n = 3). Microscopically, all tumors had a predominantly diffuse growth pattern, with a focal nested architecture in 6 cases, which closely mimicked metastatic carcinoma. The initial biopsy was performed in 2 of 6 patients and revealed the presence of a malignant neoplasm suggestive of metastatic carcinoma. Immunohistochemically, the epithelioid endothelial cells usually showed strong reactivity for CD31 (7/7), variable or focal positive staining for CD34 (5/7), Factor VIII (4/7), cytokeratin (6/7), epithelial membrane antigen (2/7), vimentin (7/7), and CD68 (3/7). In contrast, they were negative for CD1a, HMB45, lysozyme, CD45, desmin, and smooth muscle actin. Three patients died of disease within one year of the diagnosis, 2 patients developed local recurrence or metastases, and another 2 were disease-free at this writing.

Conclusions. With any unusual epithelioid neoplasm displaying some or all of the morphologic features described above, epithelioid angiosarcoma should be included in the differential diagnosis. In such an instance, endothelial markers should be incorporated in the immunohistochemical analysis to avoid misdiagnosis, particularly with limited sampling.

Key words: differential diagnosis, epithelioid angiosarcoma, histopathology, immunohistochemistry, prognosis.

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