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Clear cell sarcoma of Soft Tissue with eccrine differentiation: A case report and review of the literature

Clear cell sarcoma of soft tissue (CCSST) is a deep soft tissue tumor presenting in the extremities of young adults. Histopathologically, nests and sheets of polygonal cells with clear to eosinophilic cytoplasm separated by fibrous septae as well as occasional "wreath-like" giant cells are visualized. However, CCSST has been noted to have atypical histopathological features, such as epidermotropism, myxoid differentiation and presentation at atypical sites. Here, we present the first reported case of eccrine ductal differentiation in CCSST. The patient, a 21-year-old woman, presented with a lump of 10-year duration sized 3x5 cm on the plantar surface of the fourth and fifth interdigital spaces. There was an increase in size as well as pain and redness over 6 years. Besides the characteristic findings, there were ductal structures in continuity with the upper dermis indicative of ductal differentiation. The tumor stained positively with S100, HMB45 and succinic dehydrogenase; ducts stained positively with EMA and CEA. CCSST was confirmed with cytogenetic analysis showing the translocation associated with EWSR1-ATF1 fusion gene. Therefore, ductal differentiation is a unique finding that should be considered when evaluating for CCSST.

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