in some cases, have lent credibility to the notion that EMC has neural-neuroendocrine differentiation and is not strictly a cartilaginous tumor[9]. The fact that tumor cells are positive for vimentin in virtually all cases, but only positive for S-100 protein focally in a minority of cases, again points to a non-hyaline cartilage phenotype. Epithelial markers, GFAP, HMB-45, desmin, and myoglobin are all typically negative. The overall prevalence of gene fusion in EMC is estimated to be approximately 75% as reported in the literature[14]. EMCs harbor a balanced translocation involving the NR4A3 (TEC) gene on chromosome 9, that in approximately 75% of case results in a t(9;22)(q22;q12) translocation[15]. Although less common, occasionally the translocations involving the NR4A3 gene may result in a balanced translocation with other chromosomes, resulting in t(9;17)(q22;q11), (9;15)(q22;q21) or t(9;3)(q22;q12) gene fusions[15-17]. Although incompletely understood, translocation of NR4A3 is believed to play a critical role in the pathogenesis of EMC. Translocations involving EWSR1 in EMC have been identified using numerous methods including conventional karyotyping, reverse-transcription polymerase chain reaction (RT-PCR) and FISH[14-18]. Detection rates in archival tissue using RT-PCR range from 50% to 83%. A recent study has found FISH for EWSR1 to be highly informative (93% of cases)[8]. A recent study investigated the expression of SMARCB1/INI1 protein that is known to be involved in malignant rhabdoid tumor, another sarcomas with EWSR1 gene rearrangement. Other myxoid lesions may enter into the differential diagnosis of EMC and need to be clearly separated due to unique therapeutic and prognostic implications. The low cellularity and abundant myxoid matrix associated with EMC may cause diagnostic confusion with certain benign lesions such as myxoma and nodular fasciitis. The presence of nuclear hyperchromasia and the typical septated, nodular growth pattern help in distinguishing EMC from the above-mentioned entities. Recognition of variants of EMC, including predominantly cellular to solid tumors with minimal myxoid matrix (cellular variant), rhabdoid differentiation, or spindle cells resembling fibrosarcoma or pleomorphic sarcoma, is also extremely important. In cases composed predominantly of solid areas, the tumor may resemble other tumors with small round cells, including Ewing's sarcoma and small cell synovial sarcoma. However, careful sampling of the tumor usually reveals areas with histologic features of conventional EMC. In cases where histologic distinction of EMC from the above mentioned entities is difficult on morphologic grounds alone, studies to detect the presence of the characteristic gene fusion involving EWSR1, and the fusion partner in Ewing' sarcoma, are invaluable in aiding in the accurate classification of this tumor.

EMC is a low-grade sarcoma in which the prognosis is determined predominantly by tumor cellularity. Despite being a slow-growing tumor, it has a high rate of local recurrences and history of metastases to uncommon sites like the mandible, liver, retroperitoneum, right ventricle, pancreas, and central nervous system. EMC is typically associated with a protracted clinical course, even when metastases develop, and is hence thought to be best classified as a low-grade sarcoma. In the largest series analyzing the prognostic parameters in soft tissue EMC, the authors studied the morphologic, clinical, immunohistochemical, and ultrastructural features in 117 cases[5]. The estimated median survival time was 18 years and median intervals to metastases and local recurrences were 12 years and 8 years, respectively. Metastases occurred in 46% of cases and most frequently were to the lungs, followed by soft tissues, lymph nodes, and bones. The estimated 5-, 10-, and 15-year