

Clear Cell Sarcomas: Genomic landscape and functional validation

Clear Cell Sarcoma (CCS) are rare sarcomas characterized by an aggressive phenotype with high metastatic potential, manifesting through frequent in-transit, lymphatic, and early distant metastases. The hallmark molecular aberration in CCS is a translocation event $t(12;22)(q13;q12)$. The resulting oncogenic protein consists of EWSR1, a transcription factor fused to one of the CREB-family of transcription factors, in most cases ATF1, and less frequently CREB1 or CREM. Presently, surgical resection represents the only effective treatment option for CCS, leaving clinical (basket) trials as the standard of care for patients with metastatic disease. The identification of therapeutic vulnerabilities necessitates comprehensive bioinformatics and their validation requires informative disease models.



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Publications:

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