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Molecular analyses could improve treatment options for a rare soft tissue tumor

Desmoplastic small- and round-cell tumor is rare, aggressive, and difficult to treat. Researchers at the National Center for Tumor Diseases (NCT) Heidelberg and the German Cancer Research Center (DKFZ) have comprehensively analyzed 30 tumors and identified potential personalized treatment options for nearly all patients. In some patients, new targeted approaches were able to halt the disease for a surprisingly long time.
The National Center for Tumor Diseases (NCT) Heidelberg is a joint institution of the German Cancer Research Center (DKFZ), the Heidelberg University Hospital (UKHD), the Heidelberg Medical Faculty of the Heidelberg University and the Thoraxklinik Heidelberg.

Desmoplastic small and round cell tumor (DSRCT) is a very rare, aggressive soft tissue tumor that primarily affects young men. Because it is so rare, it is very difficult to conduct clinical trials for this type of cancer. Treatment options are correspondingly limited. This problem is exacerbated by the fact that cancer cells in DSRCT usually exhibit only a few genetic alterations. On the one hand, this reduces the likelihood that the tumors will respond to standard immunotherapies. On the other hand, it means that only a small number of patients have tumors with classic genetic alterations that are responsive to targeted therapies. Furthermore, diagnosis is challenging, so DSRCT is often initially misclassified as other types of cancer. The prognosis for patients with the disease is poor: most patients die within three years of diagnosis.

To improve treatment options, researchers at NCT Heidelberg and the DKFZ conducted a comprehensive molecular analysis of 30 patients with DSRCT who participated in the DKFZ/NCT/DKTK MASTER program. The broad molecular analysis made it possible to recommend at least one therapy tailored to their disease for 28 of the 30 participants, which was also implemented in 13 of them. Importantly, data on the response to the individually recommended therapies were also collected and evaluated.

It is noteworthy that the researchers were frequently able to detect proteins in the tumor tissue that can be targeted by novel, targeted therapies, particularly antibody-drug conjugates (ADCs) or therapeutic immune cells, known as CAR-T cells. Using a latest-generation ADC, trastuzumab deruxtecan, unusually long-lasting tumor control was achieved in two patients, in one case for over two years. Additionally, the study found that in eight out of 30 cases, the tumors could only be correctly diagnosed through molecular analysis.

Małgorzata Oleś, a bioinformatician in the Computational Oncology Group at NCT Heidelberg, is one of the study's first authors. She says: "Our work underscores the potential of broad molecular diagnostics to identify recurring treatment targets, especially for patients with rare tumor types."

Stefan Fröhling, Executive Director at NCT Heidelberg and Head of the Department of Translational Medical Oncology at the DKFZ, adds: "Progress in this type of cancer is particularly important because those affected are often very young, and we urgently need better treatment options to offer them."

The study also shows that broad international collaboration between institutions, particularly for rare tumors, is important and can provide valuable insights for the benefit of patients. As part of this study, two of the particularly successful treatments were carried out at the Centre Hospitalier de Luxembourg. Stefan Fröhling says: "We are seeing that our collaboration and sustainable strategic partnership with other centers in Germany and beyond are particularly important for creating new treatment options for patients with rare cancers."

Publication:

Marcus Renner, Małgorzata Oleś, Nagarajan Paramasivam, Christoph E. Heilig, Annika Schneider, Caroline Modugno, Catherine Herremans, Jennifer Hüllein, Barbara Hutter, Cihan Erkut, Andreas Mock, Eva Krieghoff-Henning, Cecilia B. Jensen, Amirhossein Sakhteman, Matthew The, Tony Prinz, Panna Lajer, Annika Baude-Müller, Katja Beck, Bettina Beuthien-Baumann, Leonidas Apostolidis, Sebastian Bauer, Melanie Boerries, Christian H. Brandts, Damian T. Rieke, Thomas Kindler, Frederick Klauschen, Klaus Schulze-Osthoff, Richard F. Schlenk, Guy Berchem, Michael Allgäuer, Gunhild Mechtersheimer, Albrecht Stenzinger, Daniel B. Lipka, Matthias Schlesner, Bernhard Kuster, Arne Jahn, Evelin Schröck, Christoph Heining, Maria-Veronica Teleanu, Peter Horak, Simon Kreutzfeldt, Daniel Hübschmann, Wolfgang Hartmann, Hanno Glimm, Stefan Fröhling:
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