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Desmoplastic Small Round Cell Tumor

National Cancer Institute

Source

National Cancer Institute. *Desmoplastic Small Round Cell Tumor*. NCI Thesaurus. Code C8300.

An aggressive malignant soft tissue neoplasm of uncertain differentiation. It is characterized by a recurrent chromosomal translocation t(11;22)(p13;q12) and the presence of small round cells in a desmoplastic stroma. It usually affects children and young adults. The most common site of involvement is the abdomen. Patients usually present with abdominal distention, pain, ascites, and a palpable abdominal mass. The prognosis is usually poor.